

PEDIATRIC NURSE
PRACTITIONER
CERTIFICATION
REVIEW GUIDE

Fifth Edition

Primary Care

Edited by
JoAnne Silbert-Flagg and Elizabeth Sloand

THE PERFECT
STUDY GUIDE
FOR THE ANCC
EXAM!

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Fifth Edition

Primary Care

Edited by

JoAnne Silbert-Flagg, DNP, CPNP, IBCLC

Assistant Professor

Johns Hopkins University School of Nursing

Baltimore, Maryland

Elizabeth Sloand, PhD, CRNP

Assistant Professor

Coordinator, Pediatric Nurse Practitioner Track

Johns Hopkins University School of Nursing

Baltimore, Maryland



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Contents

Preface viii

Acknowledgments ix

Author Note x

Contributors xi

Chapter 1

Test Taking Strategies and Techniques 1

Janice J. Hoffman

Self-Assessment Regarding Seeking
Certification 1

Know the Test Plan 2

Develop a Specific Plan 2

Approaches to Test Taking 2

Maximizing Study Time 5

Test Day 6

References 6

Chapter 2

Human Growth and Development 7

M. Elizabeth M. Younger

Human Growth and Development:
Underlying Theory and Science of
Child Health 7

Infant Growth and Development 10

Toddler and Preschooler Growth and
Development 13

School-Aged Development 15

Adolescent Development 16

The Changing American Family as Context
for Growth and Development 17

Deviations in Physical Growth and
Behavioral Development 18

Health Supervision as Ongoing
Surveillance, Screening, and Assessment
of Physical Growth and Behavioral
Development 31

Questions 32

Answers 35

Bibliography 35

Chapter 3

Health Promotion and Well Child Care for Infants, Children, and Adolescents 37

JoAnne Silbert-Flagg

Overview: Health Maintenance and Health
Promotion 37

Child Health Supervision 37

The Pediatric History 38

The Pediatric Physical Examination 42
 Specific Normal Findings and Common Variations 44
 Selected Laboratory Tests and Values 54
 Selected Screening Tests 57
 Special Examinations 59
 Problem-Oriented Health Record 62
 Child Health Supervision Schedule of Visits with Key Issues (Nutrition, Development, Screening, Immunizations, Anticipatory Guidance, and Health Education) 62
 Questions 77
 Answers 80
 Bibliography 80

Chapter 4

Eye, Ear, Nose, Mouth, and Throat Disorders 81

Karen K. Buch

Head 81
 Eye 84
 Ear 96
 Nose 103
 Mouth 106
 Throat 110
 Questions 114
 Answers 117
 Bibliography 117

Chapter 5

Cardiovascular Disorders 119

Peggy Dorr

Congenital Heart Disease/Defects 119
 Congestive Heart Failure (CHF) 120
 Murmurs 122
 Disturbances of Rate and Rhythm 123
 Hypertension 125
 Hypercholesterolemia 126
 Kawasaki Disease (Mucocutaneous Lymph Node Syndrome) 127
 Acute Myocardial Inflammatory Disease 129
 Rheumatic Fever/Heart Disease 130
 Questions 132

Answers 133
 Bibliography 134

Chapter 6

Lower Respiratory Disorders 135

Elizabeth Sloand

Croup (Laryngotracheobronchitis) 135
 Foreign Body Aspiration 136
 Bronchitis 137
 Bronchiolitis 137
 Pneumonia 138
 Cystic Fibrosis (CF) 140
 Asthma 142
 Bronchopulmonary Dysplasia (BPD) 144
 Apnea 146
 Tuberculosis (TB) 147
 Questions 149
 Answers 151
 Bibliography 151

Chapter 7

Dermatologic Conditions 153

Jeanne Steman Findlay

Newborn Exanthema 153
 Vascular Lesions 154
 Melanocyte Cell and Pigmentation Conditions 156
 Papulosquamous Conditions 159
 Dermatitis Conditions 161
 Burn Conditions 164
 Bacterial Conditions 166
 Bacterial Conditions Involving Pilosebaceous Unit 168
 Viral Conditions 170
 Fungal Infections 172
 Insect Conditions 176
 Insect Infestations 179
 Miscellaneous Conditions of Hypersensitivity 181
 Questions 184
 Answers 190
 Reference Materials 191
 Bibliography 192

Chapter 8**Gastrointestinal Disorders 193***Lindsay Wilson*

- Gastroesophageal Reflux in Infancy (GER) 193
- Pyloric Stenosis 194
- Acute Infectious Gastroenteritis 195
- Pinworms (*Enterobiasis vermicularis*) 199
- Inflammatory Bowel Disease (IBD) 199
- Chronic Diarrhea 201
- Malabsorption 201
- Acute Abdominal Pain 202
- Functional Gastrointestinal Disorders 203
- Constipation 205
- Hepatitis 206
- Hernia 208
- Questions 208
- Answers 211
- Bibliography 211

Chapter 9**Infectious Diseases 213***Jody K. Roblyer*

- Septicemia (Sepsis) 213
- Diphtheria 214
- Pertussis (Whooping Cough) 215
- Influenza 216
- Rubella (German Measles) 217
- Rubeola (Red measles) 218
- Roseola (Exanthem Subitum) 219
- Fifth Disease (*Erythema Infectiosum*–EI) 219
- Varicella-Zoster Virus (VZV) (Chickenpox) 220
- Mumps 221
- Cat Scratch Disease (CSD) 222
- Rocky Mountain Spotted Fever (RMSF) 223
- Lyme Disease 223
- Infectious Mononucleosis (IM) 225
- Infant Botulism 226
- Poliomyelitis 226
- Tetanus (Lockjaw) 227
- Malaria 228

- Questions 229
- Answers 231
- Bibliography 231

Chapter 10**Musculoskeletal Disorders 233***Rachel Lyons*

- Torticollis (Wry Neck) 233
- Developmental Dysplasia of the Hip (DDH) 234
- Talipes Equinovarus Congenita (Clubfoot) 235
- Metatarsus Adductus/Metatarsus Varus 235
- Tibial Torsion (Internal) 236
- Genu Varum (Bowleg) 236
- Genu Valgum (Knock-Knee) 237
- Transient (Toxic) Synovitis of the Hip 237
- Legg-Calvé-Perthes Disease (LCPD) 238
- Growing Pains 238
- Osgood-Schlatter Disease 239
- Scoliosis (Idiopathic) 239
- Sports Injuries 240
- Slipped Capital Femoral Epiphysis 241
- Juvenile Idiopathic Arthritis 242
- Systemic Lupus Erythematosus 244
- Osteomyelitis 245
- Duchenne Muscular Dystrophy (DMD) 245
- Questions 246
- Answers 249
- Bibliography 249

Chapter 11**Neurological Disorders 251***Maureen C. Maguire*

- Seizure Disorders/Epilepsy 251
- Febrile Seizures 255
- Headaches 255
- Neurofibromatosis (NF1) (von Recklinghausen Disease) 257
- Tuberous Sclerosis (TS) 258
- Tic Disorders 258
- Head Injuries 260

Meningitis 262
 Brain Tumors 263
 Questions 264
 Answers 267
 Bibliography 267

Chapter 12

Genitourinary/Gynecological Disorders/ Adolescent Pregnancy 269

*Marlo A. Eldridge and MaryLou C.
 Rosenblatt*

Urinary Tract Infection (UTI) 269
 Enuresis 271
 Cryptorchidism (Undescended
 Testes) 272
 Hydrocele 273
 Hypospadias 273
 Phimosis and Paraphimosis 274
 Meatal Stenosis 274
 Testicular Torsion 275
 Labial Adhesions (Labial Fusion, Synechia
 Vulvae, Labial Agglutination) 275
 Vulvovaginitis 276
 Dysmenorrhea 277
 Premenstrual Syndrome 278
 Genitourinary Trauma 279
 Glomerulonephritis (GN) 279
 Hydronephrosis 280
 Renal Tubular Acidosis (RTA) 280
 Sexually Transmitted Infections 281
 Contraception 288
 Issues of Pregnancy and Birth for the
 Adolescent 290
 Questions 293
 Answers 298
 Bibliography 298

Chapter 13

Hematologic/Oncologic/Immunologic Disorders 299

M. Elizabeth M. Younger and Kathy Ruble

ABO Incompatibility 299
 Rh Incompatibility 299
 Neonatal Hyperbilirubinemia 300
 Hemoglobinopathies 301

Iron Deficiency Anemia 302
 Sickle Cell Disease 303
 Lead Poisoning (Plumbism) 305
 Glucose-6-Phosphate Dehydrogenase
 (G-6-PD) deficiency 306
 Bleeding Disorders 307
 Cancers 310
 Immune Deficiencies 314
 Questions 316
 Answers 318
 Bibliography 318

Chapter 14

Endocrine Disorders 321

Malinda D. Duke

Thyroid Disorders 321
 Pituitary Disorder—Diabetes Insipidus
 (DI) 326
 Growth Disturbances 328
 Adrenal Gland Disorders 332
 Miscellaneous 335
 Questions 344
 Answers 347
 Bibliography 347
 Educational Web Sites 348

Chapter 15

Multisystem and Genetic Disorders 349

Rita Marie John

Introduction 349
 Congenital Diseases 350
 Genetic Syndromes 359
 Multisystem Disorders 366
 Questions 369
 Answers 371
 Bibliography 372

Chapter 16

Advanced Practice, Role Development, Current Trends, and Health Policy 373

Janet S. Selway

Introduction 373
 Advanced Practice Nursing 373

Practice Issues	377	Questions	387
Professional Organizations	382	Answers	390
Health Policy	382	Bibliography	390
Types of Healthcare Delivery Systems	384		
Health Insurance Portability and Accountability Act of 1996 (HIPPA)	386	Index	393

Preface

We are pleased to present the fifth edition of the *Pediatric Nurse Practitioner Certification Review Guide* from Jones and Bartlett Publishers. The book is a revision of the 2004 text published by Health Leadership Associates, edited by Virginia Layng Millonig and Caryl Mobley. It has a dual purpose. The book serves pediatric nurse practitioner (PNP) and family nurse practitioner (FNP) graduates who are doing self-study preparation for the PNP primary care certification exam. Students and graduates can be overwhelmed when faced with the vast body of knowledge required for certification. This book brings together the most current and essential information in an organized, concise, and comprehensive manner. This book also serves as a valuable clinical resource for practicing PNPs and FNPs in pediatric primary care.

The first chapter of the book reviews test-taking strategies to help students plan their certification preparation, considers various approaches to test taking, and helps students organize their study time. The second chapter reviews growth and development; the third chapter, which covers health promotion and well child care for infants, children and adolescents, includes expanded information on the promotion of breastfeeding in the first year of life. The subsequent chapters of the book review the following systems: HEENT, cardiovascular, lower respiratory, dermatology, gastrointestinal, infectious diseases, musculoskeletal, neurological, GU and GYN (including adolescent pregnancy), hematologic/oncologic and immunologic

disorders, endocrine, and multisystem and genetic disorders. The book concludes with a chapter on advanced practice nursing, role development, current trends, and health policy.

Pediatric Nurse Practitioner Certification Review Guide presents the most updated information and current standards of practice for the NP in the pediatric primary care setting. It examines all the content areas required for the certification examinations for PNPs in primary care. Each system chapter presents common pediatric disorders followed by a definition of the disorder and its etiology/incidence, signs and symptoms, differential diagnosis, physical findings, diagnostic tests, and management/treatment. Practice test questions follow content in each chapter. This format enables the reader to review essential information and to explore areas that may require further study. Bibliographies and relevant web sites give the reader resources for further discussion and study.

All chapter authors are specialists in their areas. Clinical chapter authors are certified PNPs and expert clinicians who have practiced extensively in their specialty area. The editors are faculty members at the Johns Hopkins University School of Nursing in the PNP primary care master's degree program.

We hope that this review guide assists the PNP graduate to become certified and to provide competent and compassionate health care throughout their career to children, adolescents, and their families.

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We are pleased to have had the opportunity to revise the work of Virginia Layng Millonig and Caryl Erhardt Mobley. We owe additional thanks to each chapter author for her diligence, knowledge, and expertise.

We are grateful to our professional colleagues who have mentored, supported, and taught us over the years, particularly those at the Johns Hopkins University School of Nursing.

JoAnne Silbert-Flagg and Elizabeth Sloand

As a practicing PNP for over 20 years at Columbia Medical Practice in Columbia, Maryland, I have had the opportunity to provide pediatric primary care to a generation of patients. I hope I have touched their lives as much as they have touched mine. Over the years, the pediatricians and PNs in the practice have been a source of support and inspiration for me. I am especially indebted to R. Scott Strahlman, MD, a pediatrician

in the practice, who agreed to be my preceptor during the residency portion of my DNP program at Columbia University.

I have been fortunate to have the support of my parents, Herbert and Betty Silbert, and my brother Brett Silbert. They have provided encouragement as I pursued my educational, professional, and personal goals. Lastly, and most importantly, I dedicate this book to my children, Christopher and Jennifer Flagg.

JoAnne Silbert-Flagg

I am personally indebted to my husband, Bob, who makes everything possible; my children Daniel, Christine, Rachel, and Rosie, who are a continued source of joy and pride; and to my parents Dorothy and Joseph for their ever-present encouragement and love.

Elizabeth Sloand

Author Note

The authors would like to thank and acknowledge the chapter authors from the fourth edition of this text.

Chapter 1. Test Taking Strategies: Nancy A. Dickenson Hazard, MSN, RN, CPNP, FAAN and Virginia Layne Millonig, PhD, RN, CPNP

Chapter 2. Human Growth and Development: Mary A. Baroni, PhD, RN, CPNP

Chapter 3. Health Promotion and Well Child Care for Infants, Children, and Adolescents: Bernadette Mazurek Melnyk, PhD, RN, CS, PNP and Leigh Small, PhD, RN, CPNP

Chapter 4. Head, Eye, Ear, Nose, Mouth, and Throat Disorders: Connie Paczkowski, MSN, RN, CS, PNP

Chapter 5. Cardiovascular Disorders: Karen Uzark, PhD, RN, CPNP

Chapter 6. Lower Respiratory Disorders: Janet Johnston, MSN, RN, CPNP

Chapter 7. Dermatologic Conditions: Beverly Bigler, EdD, RN, CPNP

Chapter 8. Gastrointestinal Disorders: Victoria A. Weil, MSN, RN, CS, CPNP

Chapter 9. Infectious Diseases: Nancy E. Kline, PhD, RN, CPNP, FAAN

Chapter 10. Musculoskeletal Disorders: Susan Heighway, MS, RN, CS, PNP

Chapter 11. Neurological Disorders: Susan Heighway, MS, RN, CS, PNP

Chapter 12. Genitourinary and Gynecological Disorders Adolescent Pregnancy: Sandra Elvik, MS, RN, CPNP and Mary A. Baroni, PhD, RN, CPNP

Chapter 13. Hematological/Oncologic/Immunologic Disorders: Juanita Conkin Dale, PhD, RN, CPNP

Chapter 14. Endocrine Disorders: Jacalyn Peck Dougherty, PhD, MA, MS, RN, CPNP

Chapter 15. Multisystem and Genetic Disorders: Alison W. Schultz, EdD, RN, CPNP and Virginia Richardson, DNS, RN, CPNP

Chapter 16. Advanced Practice, Role Development, Current Trends, and Health Policy: LeeAnne C. Busby, DSN, RN, CS, FAANP and Mary A. Baroni, PhD, RN, CPNP

Contributors

Karen K. Buch, RN, MSN, CPNP

Clinical Instructor
Johns Hopkins University
School of Nursing
Baltimore, Maryland

Peggy Dorr, MS, CRNP

Pediatric Nurse Practitioner
Division of Pediatric Cardiology
University of Maryland Medical Center
Baltimore, Maryland

Malinda D. Duke, CPNP, CDE

Pediatric Endocrinology
Joslin Diabetes Center
University of Maryland
Baltimore, Maryland

Marlo A. Eldridge, MSN, CPNP

Co-Director
Voiding Improvement Program of the Brady Urological
Institute
Johns Hopkins Hospital
Baltimore, Maryland

Jeanne Steman Findlay, CRNP, CCRP

Instructor
Department of Dermatology
Johns Hopkins University
School of Medicine
Baltimore, Maryland

Janice J. Hoffman, PhD, RN, CCRN

Assistant Professor and Vice Chair
Organizational Systems and Adult Health
University of Maryland
School of Nursing
Baltimore, Maryland

Rita Marie John, EdD, DNP, CPNP

Pediatric Nurse Practitioner/Neonatal Nurse
Practitioner Program Director
Columbia University
School of Nursing
New York, New York

Rachel Lyons, DNP, CPNP-AC/PC

Assistant Professor of Clinical Nursing
Pediatric Orthopaedic Surgery
Morgan Stanley Children's Hospital of New York
Presbyterian
Columbia University
School of Nursing
New York, New York

Maureen C. Maguire, MSN, RN, PNP

Assistant Professor
Johns Hopkins University
School of Nursing
Baltimore, Maryland

Jody K. Roblyer, MS, CRNP-P

Clinical Instructor
Johns Hopkins University
School of Nursing
Baltimore, Maryland

MaryLou C. Rosenblatt, RN, MS, CPNP
Lead Nurse Practitioner, Adolescent Medicine Clinic
Faculty LEAH (Leadership Education in Adolescent
Health)
Johns Hopkins Hospital
Baltimore, Maryland

Kathy Ruble, PhD, RN, CPNP, AOCN
Director, Survivorship Program
Johns Hopkins University
Department of Oncology
School of Medicine
Baltimore, Maryland

Janet S. Selway, DNSc, CRNP
Adult/Pediatric Nurse Practitioner
Assistant Professor
University of Delaware
School of Nursing
Newark, Delaware

Lindsay Wilson, RN, MSN, CPNP
Pediatric Nurse Practitioner
Pediatric Gastroenterology and Nutrition
Johns Hopkins Children's Center
Johns Hopkins Hospital
Baltimore, Maryland

M. Elizabeth M. Younger, PhD, CPNP
Pediatric Immunology
Johns Hopkins Hospital
Baltimore, Maryland

Test Taking Strategies and Techniques

Janice J. Hoffman

The decision has been made to take the certification examination, so it is now time to make plans. Based upon prior experiences with standardized testing situations, there may be some anxiety and questions about the certification process. This chapter includes an overview of the test plans for primary care pediatric nurse practitioner certification, followed by study and test-taking strategies to assist in preparation for the examination.

■ SELF-ASSESSMENT REGARDING SEEKING CERTIFICATION

The first question that only the nurse contemplating the certification process can answer is: “Why do I want to become certified as a pediatric nurse practitioner?” There are many benefits of professional certification, including demonstration of specialized knowledge and skills, recognition as an expert in the field, possible monetary benefits, possible career advancement, personal growth, and greater satisfaction as a professional nurse (Lamonte, 2007). Personal confidence in caring for these patients is another benefit of professional certification.

Once the decision is made to seek certification, questions may arise about how best to study, who and what can assist in the preparation, as well as feelings about past testing experiences. In the following sections, specific content is provided about preparing for the examination and about ways to deal with the emotions associated with taking standardized examinations.

Based upon past examination performance, most test takers are considered either strong or weak test takers.

For those nurses who have experienced success with formal and/or standardized tests in the past, examine approaches used and attempt to adapt these practices in preparing for this certification exam. For those who have not performed well on past tests, it is important to determine whether this past performance was based upon poor test-taking skills or lack of preparation, which may be related to lack of knowledge or insufficient review of the test plan. The area of pediatrics that the nurse is currently working will have an impact on performance as well as preparation. For nurses working in a very specialized area, such as pediatric cardiac surgery, while they are experts in cardiac surgery and care of the critically ill child, they may need to focus on primary care, normal growth and development, and other areas that are tested on the exam.

Becoming familiar with areas of content to be reviewed is usually straightforward; examining and dealing with test anxiety is another issue. For those individuals with significant test anxiety, one approach is to become very familiar with the content and process of the certification examinations. Another strategy is to complete practice tests, even simulating testing circumstances like sitting in a quiet monitored place and completing all questions prior to review. There are on-line testing resources available to further familiarize the candidates with the examination process in an effort to decrease anxiety. Test taking is a skill, and as with any skill, test taking should improve with consistent practice. Finally, the candidate needs to use personal strategies that have been successful in other high anxiety circumstances, such as deep breathing, visualization, exercise, etc.

■ KNOW THE TEST PLAN

Certification as a pediatric nurse practitioner (PNP) can be gained through the American Nurses Credentialing Center (ANCC) or the Pediatric Nursing Certification Board (PNCB). One of the first priorities in certification examination preparation is to review the requirement for each organization. This information can be found at the following Web sites:

<http://www.nursecredentialing.org/NurseSpecialties/PediatricNP.aspx> (ANCC)

<http://www.pncb.org/ptistore/control/exams/pnp/admin> (PNCB)

Once verification of eligibility for taking the examination has been determined, specific planning should begin. In preparing for the exam, make sure that sufficient time is planned for review of content and practice questions. On both of the above listed Web sites, there are specific resources listed to assist in the preparation for the certification examination. Other things to consider are work schedule and any other commitments or conflicts that might potentially compete with study time.

The next step is to review the test plan for each examination; the detailed plans for both the ANCC and PNCB examinations are available from their respective Web sites. Reviewing the content is important in guiding the individual plan of study and will assist in estimating the time each individual candidate needs to review and study. In the next section, suggested guidelines for developing a specific study plan are described.

■ DEVELOP A SPECIFIC PLAN

In reviewing the test plans, it is important to begin to prioritize the content areas that review is most needed. One strategy is to develop a rubric for self-assessment such as the following:

- 4 Very knowledgeable (little review needed)
- 3 Knowledgeable (some review needed)
- 2 Familiar (significant time needed for study and review)
- 1 Unknown (priority area for study and review)

In using this four-choice rubric there is no middle or average choice which forces a decision between an area needing significant study and review and one that requires less time.

Now that a self-assessment rubric has been determined, a review of the test plan is needed. **Table 1-1** provides an overview of the PNCB examination and **Table 1-2** provides the outline of the ANCC test. It is important to schedule study time, with details about length of time allotted and content to be reviewed. One approach is to review your calendar for the next three months and to schedule specific study days and times,

just as work and other appointments are scheduled. Based upon the self-assessment of baseline knowledge compared with the content outline and timeframe until test day, an individualized plan of study can be developed. In addition to a review of content headings, **Tables 1 and 2** provide percentages and numbers of test questions for the respective examinations based upon the most recent practice analyses for pediatric nurse practitioners. **Table 1-3** is a sample study plan for one week, based upon the PNCB test plan. While there are similarities in content, it is best to select specific study guides and review resources specific to the examination that is being taken.

A strategy to address potential content issues is to conscientiously review any new medication, term, diagnosis, and so forth that is encountered in practice or in a journal during the preparation time. Consider a small (2 × 3 inch) notebook or an electronic device to record this “new” data. Review this content to broaden your knowledge base. Both ANCC and PNCB suggest taking advantage of the review resources available, including books, audiovisual materials, and workshops.

This one-week calendar is used as an example in planning study time. It is important to plan study time in relation to work schedule. If working 12-hour shifts, it is probably not feasible to study on these work days. To be more efficient with study time, it is beneficial to “schedule” the study time and the specific topic for study. Valuable time can be wasted “trying to decide what to study.” With a plan, as in **Table 1-3**, not only are the times detailed, but also the specific content is identified. The amount of time for each category of the test plan should be based upon the individual self-assessment results.

■ APPROACHES TO TEST TAKING

All questions on the pediatric NP certification examinations are multiple-choice questions. Each question includes the stem and four to five choices. The stem contains the content being tested and is sometimes stated as a question. The key to correctly answering these types of questions is to accurately determine what the question or stem is asking. Because questions may have distracting information that is not needed to answer the question correctly, one strategy is to reword the question into a short phrase that you can clearly understand and consider. For example, the question “A benign (Still’s) murmur is most accurately described as ___” may be reworded to “Describe a Still’s murmur.”

Cognitive Levels of Questions

The pediatric NP certification exam questions, as with most other professional nursing organization certification examinations (AACN, AORN, CEN, etc.), are based upon Bloom’s taxonomy of cognition. The first levels,

■ **Table 1-1** Pediatric Nursing Certification Board Test Plan

Domains of Practice	Number of Questions	Knowledge Level	Percent of Test	Hours for Study
Health Promotion/Prevention/Anticipatory Guidance	34		23%	
Assessment	53		35%	
History and Physical Exam	14		9%	
Diagnostic Testing and Screening	11		7%	
Growth and Development	12		8%	
Diagnosis	16		11%	
Management	53		35%	
Therapeutic Interventions	29		20%	
Procedures	3		2%	
Counseling and Education	8		5%	
Referral	3		2%	
Collaboration and Case Management	5		3%	
Evaluation and Follow-up	5		3%	
Professional Issues	10		7%	

Adapted from *PNCB Primary Care Pediatric Practitioner Detailed Content Outline* (2009). **Bolded/shaded headings represent major categories of the test.** Note those content areas that have high percentages on the examination, and ensure sufficient study time dedicated to these areas based upon self-assessment. A more detailed study plan can be developed by using the detailed test plans.

knowledge, comprehension, application and analysis, are used in the development of multiple-choice examinations. As with other nursing examinations, the majority of questions are at the application and analysis level. A short review of the different types of questions and examples are presented in this section.

Based upon Bloom's taxonomy, knowledge-based questions are at the lowest cognitive level and test the ability to recognize and recall information. An example of a knowledge-based question is:

What is a common complication of a blood transfusion?

- A. Fever
- B. Elevated blood pressure
- C. Hematuria
- D. Change in level of consciousness

The correct answer is A. This question requires the test taker to remember only a fact in order to answer the question correctly. There are not large numbers of this level of question on the exam, but basic knowledge is important to review.

The next level of question is **comprehension**, which requires the nurse to understand the meaning of the material. An example of this level of question is:

The nurse understands that fever is a complication of a blood transfusion because antibodies in the patient's blood cause:

- A. changes in the temperature set point in the hypothalamus
- B. collapse of blood vessels

■ **Table 1-2** ANCC Pediatric Nurse Practitioner Board Certification

Domains of Practice	Number of Questions	Knowledge Level	Percent of Test	Hours for Study
Health Promotion and Disease Prevention	29		19.00%	
Assessment of Acute and Chronic Illness	35		23.00%	
Clinical Management of Acute and Chronic Illness	41		27.50%	
Nurse Practitioner and Patient Relationship	31		21.00%	
Professional Role and Policy	12		8.00%	
Research Utilization	2		1.5%	

Adapted from ANCC Web site, <http://www.nursecredentialing.org/>

■ Table 1-3 Sample One Week Study Plan

	Sunday	Monday	Tuesday	Wednesday	Thursday	Friday	Saturday	
6 a.m.	Work	Work				Work		
7 a.m.								
8 a.m.			Study Respiratory	Study Respiratory	Study GI Disorders			Study
9 a.m.								
10 a.m.								
11 a.m.								
12 a.m.								
1 p.m.			Study Respiratory	Study Respiratory				
2 p.m.								
3 p.m.								
4 p.m.					Study			
5 p.m.								
6 p.m.								
7 p.m.								
8 p.m.					Study Infectious Disease			
9 p.m.								
10 p.m.								

- C. an increase in the metabolic rate
- D. a reaction against donor proteins

The correct answer is D. The question is asking why does fever occur after a blood transfusion and requires the nurse to understand the physiologic etiology of the fever.

Application is the next level of question in Bloom’s taxonomy and evaluates the nurse’s ability to take information and apply it to a given situation. The following question is an example of an application question:

The nurse is caring for a 15-year-old patient receiving a blood transfusion. Which findings would require the nurse to immediately stop the transfusion?

- A. Pain at the insertion site
- B. Coolness at the insertion site
- C. Fever and chills
- D. Thirst

The correct answer is C. This question requires the nurse to know the side effects of a blood transfusion and which would require an immediate intervention.

The highest cognitive level tested on the certification examination is at the **analysis** level. At this level, the nurse is often expected to take a familiar piece of information and apply it to an unfamiliar setting. An example of an analysis question is:

Which dietary selections would be MOST effective for the patient with pernicious anemia?

- A. Peaches and cottage cheese
- B. Liver and onions

- C. Pasta with clam sauce
- D. Chicken breast and corn

The correct answer is B. This question requires the nurse to know that pernicious anemia is related to iron deficiency and also have knowledge of foods high in iron. While some of the other selections have some iron, liver and kidneys have large amounts of iron. In review, the certification exam has questions from all four cognitive levels, but the majority of the questions are at the application and analysis levels.

Prioritization Issues

Prioritizing issues is a competency that is relevant for success on certification examinations. In many questions, the nurse practitioner will be asked which intervention to complete first. There are two major approaches to prioritizing care: using the “ABC’s” (Airway, Breathing, and Circulation) and Maslow’s hierarchy of needs.

Consider the following question:

Four patients present to the Emergency Room. Which patient should the pediatric nurse practitioner see first?

- A. 2-year-old with chest burns
- B. 3-year-old with 2-day history of diarrhea
- C. 4-year-old with suspected pneumonia
- D. 5-year-old with cut to left thigh

The correct answer is A. While all of these patients require an assessment, based upon the ABCs, the child with chest burns is the highest priority. All patients with burns to the face, chest, and abdomen require

immediate evaluation for potential airway swelling secondary to heat near the airway. If airway swelling occurs, the patient may have a significantly compromised airway and require immediate intubation.

In using Maslow's hierarchy of needs, it is important to remember that physiologic needs always have the priority over psychological needs. Another important concept is a focus on safety, food, and water as basic needs for all people.

Potential Test Item Traps

As described above, one of the key actions to increase the likelihood of correctly answering the questions is to clarify the meaning or intent of the question. Based upon the format of the question, there are several potential traps that can lead to choosing the incorrect answers. Here are suggestions to help the PNP to correctly identify the topic of the question.

Many questions will ask the PNP to prioritize or make a clinical judgment in order to select the correct answer. The stem of the question may ask the PNP to choose the "first" (indicating prioritization) or "most" or "best," which requires discrimination and clinical judgment. It is really important to recognize these words and consider them carefully in selecting the correct answer.

Another potential trap is the use of "negative" questions. When was the last time the nurse reviewed the patient assignment and asked, "what will I not assess in this patient?" There are two relatively frequently used negative formats used on standardized tests that are important to review here. Consider the following question:

The nurse practitioner is evaluating a 6-year-old admitted with a diagnosis of acute appendicitis. Which finding requires an immediate intervention?

- A. Nausea and vomiting
- B. Dry mucous membranes and scant urine output
- C. Abdominal pain and cramping
- D. Anorexia and constipation

The correct answer is B. The other options are expected clinical manifestations in patients with appendicitis. Option B indicates a complication; the patient is showing signs of dehydration secondary to the vomiting, anorexia, and decreased oral intake. This type of question asks the nurse to find something in the answer choices that requires an action or intervention.

Another potential trap, again involving a negatively formatted question, is in the form of a patient teaching scenario. There are two ways that teaching questions are typically asked on a standardized nursing examination:

"Which statement indicates that the patient/family member **understands** the information?"

"Which statement indicates the **need for further teaching**?"

The second question is an example of asking "what is wrong." It is important with these types of questions that the nurse clarifies whether the question is asking for a correct or incorrect response by the patient. Consider the following example:

Which statement, by the parent of a 1-day-old child, indicates the need for further teaching?

- A. "I should bring the baby back for a check-up in two weeks."
- B. "The baby should have at least four wet diapers per day."
- C. "The baby will want to nurse every 2–3 hours."
- D. "The stool probably will be dark and sticky for the first couple of days."

The correct answer is B; all the other statements are correct about a newborn infant. A newborn with less than 8 wet diapers per day is most likely not getting sufficient fluids. Consistently remembering to clarify the meaning of the stem with each question increases the likelihood of correctly answering these negatively formatted questions.

▣ MAXIMIZING STUDY TIME

As discussed previously, it is very important to develop an individualized study plan based upon work schedule, personal commitments, and self-assessment of test plan content. Because most certification books include practice questions, and there are online resources available for both pediatric NP certification exams, test-taking skills can be improved with practice. Additionally, these practice items often familiarize the nurse with the types of questions to expect on the actual exams. Another consideration is whether to study in private or with a colleague who may also be taking the examination. Study groups may be very effective for the auditory learner who learns best through hearing and discussing content.

While practicing taking questions is important, effective review of the correct answers is equally, if not more, important. After completing a practice examination, it is important to review all questions to determine content areas that are still in need of further study, as well as those areas that mastery of content is observed. When reviewing the answer to an item, while it is good that a question was answered correctly, it is **more** important that the question was answered for the correct reason. Examine the stated rationale for the correct answer, as well as the rationale for why the other choices were not correct. This strategy for reviewing exam questions familiarizes the nurse with the types of questions and reminds the nurse of relevant content that is

available in the rationales for both correct and incorrect answers.

As the day for the exam gets near, consider simulating actual testing circumstances—quiet room, no interruptions, and no use of notes! Prepare a sample test, or plan on taking a set of 50 questions in 60 minutes, as this closely reflects the approximate time needed to answer the questions. Remember, on both exams there are 175 questions (150 scored and 25 experimental), and the allotted time to complete the exam is 3 hours.

▣ TEST DAY

Just as important to sufficient preparation for the examination is being attentive to getting ready for the test day. Because last minute cramming has not been shown to be effective, and because it may increase test anxiety, this is not a recommended strategy. In the schedule for the day before the exam, the “stop” time for studying needs to be clearly identified. Getting a good night’s sleep and being well rested is more important at this stage of preparation than last minute cram sessions. Also, at this late date, an unfamiliar concept, terminology, or diagnosis may undermine the confidence that has been growing over the weeks or months of preparation.

The day of the exam, make sure you eat a healthy meal and take several healthy snacks and beverages with you to the testing site. Ensure that the location of the testing center is known, and make plans to arrive there 15–20 minutes early; many testing centers do not allow test takers to start the examination late. No study guides or books should be taken to the testing site, as again, coming across an unfamiliar concept may increase test anxiety. If you **MUST** have something with you, have flash cards or index cards with specific data to review; for example development tasks that are expected at specific ages, normal laboratory values, or recommended immunizations schedules and common side effects.

After completing the exam, consider some type of personal reward! A lot of time and effort was expended in preparing for the exam. Remember, while the ultimate goal is certification, a great deal of what was reviewed and learned in preparation for the examination contributed to professional growth and will benefit

your patients. Once you receive your certification, make an effort to encourage a colleague to pursue this goal, and use your newly developed study habits and competence to mentor this potential candidate.

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Human Growth and Development

M. Elizabeth Younger

■ HUMAN GROWTH AND DEVELOPMENT: UNDERLYING THEORY AND SCIENCE OF CHILD HEALTH

- Definition: How and why a person changes and/or stays the same over time
 1. Nature (heredity and genetic predisposition) vs. nurture (environmental influence)
 2. Continuity (continual process) vs. discontinuity (distinct stages)
- Three Fundamental Domains: Conceptual distinctions but overlapping processes in reality
 1. Physical domain
 - a. Genetic factors
 - b. Physical stature and appearance
 - c. Nutritional status
 - d. Physical health and well-being
 - e. Fine and gross motor abilities
 2. Cognitive domain
 - a. Perception, thinking, information processing, and memory
 - b. Communication—receptive and expressive language
 3. Psychological and social domain
 - a. Temperament and personality
 - b. Interpersonal relationships
 - c. Moral development
 - d. Home environment and other social contexts

Classic Developmental Theories and Theorists

- Nature Positions: Emphasis on heredity and maturational process
 1. Jacque Rousseau (1712–1778)
 - a. *Emile—Treatise on Education*
 - b. Child as an “untamed savage”
 2. Charles Darwin (1809–1882)
 - a. *The Origin of Species*—human evolution (phylogeny) in order to adapt; “survival of the fittest”
 - b. First use of “baby journal” as systematic method to document observed behavioral development
 3. Alfred Binet (1857–1911) and Theophile Simon (1873–1961)
 - a. First standardized measurement of intelligence
 - b. Intention was to identify “mentally defective” children needing specialized education
 4. G. Stanley Hall (1844–1924)
 - a. *Adolescence* (1904)—classic work that first described adolescence as a critical developmental period of “Sturm und Drang” or “Storm and Stress.”
 - b. Development of questionnaires as method for the study of child development via adult retrospective recall
 5. Arnold Gesell (1880–1961)
 - a. Maturational-organismic theory—biological basis of development; organizing principle of the theory is

- structure (closed system of transformational rules governing thought)
 - b. *The Atlas of Infant Behavior* (1934)—developmental milestones
 - c. Developed one of the earliest infant tests
- 6. Sigmund Freud (1856–1939)
 - a. *Three Contributions to the Sexual Theory* (1905)
 - b. Stage theory of psychosexual development
 - (1) Infancy: Oral stage
 - (2) Toddler: Anal stage
 - (3) Preschool: Phallic stage
 - (4) School age: Latency stage
 - (5) Adolescence: Genital stage
 - c. Key principles
 - (1) Id: Principle of pleasure
 - (2) Ego: Principle of reality and/or self-interest
 - (3) Superego: Principle of morality or conscience
- 7. Neo-Freudian: Erik Erikson (1902–1994)—stage theory of life span psychosocial development
 - a. Infancy: Trust vs. mistrust
 - b. Toddler: Autonomy vs. shame and doubt
 - c. Preschool: Initiative vs. guilt
 - d. School age: Industry vs. inferiority
 - e. Adolescence: Identity vs. role confusion
 - f. Young Adult: Intimacy vs. isolation
 - g. Middle-age: Generativity vs. stagnation
 - h. Older Adulthood: Integrity vs. despair
- 8. Neo-Freudian: Margaret Mahler (1897–1985)—psychological birth of the infant
 - a. Psychological birth of the child with emerging sense of self as separate from mother
 - b. Inadequate early mothering and “psychological birth” results in mental illness
 - c. Phases of “psychological birth”
 - (1) Autism from 0 to 2 months—no real social awareness or concept of self
 - (2) Symbiosis from 2 to 5 months—mother-infant dependency
 - (3) Separation-individuation from 6 to 36 months
 - (a) Differentiation and practicing (6 to 12 months)—beginning awareness of self as separate from mother
 - (b) Rapprochement (12 to 24 months)—exploration, emotional refueling, and ability to sustain brief separations
 - (c) Consolidation (24 to 36 months)—increased ability to cope with separations through symbolic play
- 9. Jean Piaget (1896–1980)
 - a. Interactionist—structuralist stage theory of cognitive development
 - b. Sensorimotor stage: Birth to 2 years
 - c. Preoperational thinking: 2 to 7 years
 - d. Concrete operational thinking: 7 to 12 years
 - e. Formal operational thinking: 12 years +
- Nurture Positions: Emphasis on learning and environment
 - 1. John Locke (1632–1704)
 - a. *Essay Concerning Human Understanding and Some Thoughts Concerning Education*
 - b. Child as “tabula rasa” or “blank slate”
 - 2. John B. Watson (1878–1958)—behaviorism
 - a. Classical conditioning—neutral stimulus associated with a meaningful one over time leading to a “conditioned” response that can be elicited by neutral stimulus alone as though it were the meaningful one
 - b. *Psychological Care of the Infant and Child* (1928)
 - 3. B. F. Skinner (1904–1990)—student of Watson
 - a. Mechanistic-learning theory
 - b. Operant or instrumental conditioning—behavior is reinforced or extinguished by positively or negatively experienced consequences such as use of “time-out” for misbehavior
 - 4. Albert Bandura (born 1925) and Walter Mischel (born 1930)—Social learning theory
 - a. Influenced by both behaviorism and psychodynamic theories
 - b. Behavior results from interaction of individual characteristics, the environment and the behavior itself
 - c. Modeling—learning from direct observation and subsequent imitation of what is seen and done by significant others in the proximal environment
- Ethologic Theories
 - 1. Konrad Lorenz (1903–1989)—*sensitive periods* as biologically-programmed periods predisposed for particular learning, e.g., *imprinting*
 - 2. Harry Harlow (1905–1981)—Wisconsin primate laboratory’s classic rhesus monkey experiments on maternal deprivation (separation, isolation, and “terry cloth vs. wire surrogate mothers”)
 - a. Maternal separation and social isolation resulted in dramatic impairment of social-emotional development

- b. Physical contact and comfort as necessary for normal social and emotional development
- 3. John Bowlby (1907–1990): *Attachment and Loss* (1969)
 - a. Attachment defined as “an affectional tie the infant forms to another specific person that binds the two together in space and endures over time”
 - b. Importance of early mothering and consequences of “maternal deprivation” as observed in orphanages and asylums
- 4. Marshall Klaus and John Kennel—*Maternal-Infant Bonding* (1976)
 - a. Seminal studies on the impact of early contact vs. separation on maternal-infant bonding
 - b. Influential in advocating change in hospital policies regarding rooming-in and father participation in the delivery room
- 5. Mary Ainsworth (1913–1999)—*Patterns of Attachment: A Study of the Strange Situation* (1978)—developed laboratory paradigm “strange situation” to assess security or insecurity of the attachment relationship
 - a. Early maternal responsiveness to infant needs promotes secure attachment
 - b. Secure maternal-infant attachment provides “safe base” from which the child can begin to actively explore the environment and a source of comfort when distressed
 - c. Securely attached infants show more optimal cognitive gains and, later, school performance, illustrating the interconnectedness of psychosocial and cognitive domains
- Humanistic Theories
 - 1. Abraham Maslow (1908–1970)
 - a. Theory of basic needs and human potential derived from study of healthy, creative individuals; few people achieve self-actualization
 - b. Hierarchy of needs
 - (1) Physiologic
 - (2) Safety, security, and stability
 - (3) Affiliation, acceptance, and love
 - (4) Ego, self-worth, confidence, competence, and success
 - (5) Self-actualization
 - 2. Carl Rogers (1902–1987)
 - a. Client-centered approach from a phenomenologic perspective
 - b. Key strategies for intervention
 - (1) Unconditional positive regard, empathy, and genuineness
 - (2) Empathy
- (3) Congruence or the ability to be genuine
- Moral Development
 - 1. Lawrence Kohlberg (1927–1987)—Stages of moral development
 - a. Based on his original cross-sectional study of 84 school-aged boys (10 to 16 years) recruited from two suburban Chicago schools who were later followed longitudinally
 - b. Responses to hypothetical moral dilemmas
 - c. Preconventional or pre-moral level
 - (1) Stage 1: Punishment-obedience—child behaves to avoid punishment
 - (2) Stage 2: Instrumental-exchange—child behaves well for some gain or reward
 - d. Conventional level
 - (1) Stage 3: Good-boy/good-girl orientation—child behaves for approval
 - (2) Stage 4: Law and order perspective—child behaves to avoid getting caught
 - e. Postconventional level
 - (1) Stage 5: Social contract—child/adolescent behaves in accordance with generally accepted social norms
 - (2) Stage 6: Universal ethical principles—child/adolescent decides on moral standards of behavior through individual reflection and reasoning
 - 2. Carol Gilligan (1936–)—Gender differences in moral development
 - a. Male social development orients to ethic of *principles*, with moral issues decided on the basis of fairness and justice
 - b. Female social development orients to ethic of *interpersonal relationships*, with moral issues decided on the basis of compassion and caring
- Language Development: L. S. Vygotsky (1896–1934)
 - 1. Language as biologically-programmed but children learn language actively through direct experience and culture
 - 2. Zone of proximal development (ZPD): Zone between child’s opportunity to observe/participate and ability to internalize the learned behavior

Transactional and Contextual Theories of Development

- Heredity-Environment Interactions
 - 1. Growth and developmental outcomes result from “main effects” and “interaction effects”

- of and between both heredity (nature) and environment (nurture)
- 2. *Reaction Range*—range of phenotypes that may emerge from similar genotypes developing under varied environmental contexts
- Transactional and Resiliency as Models of Development
 1. Transaction model first described in 1975 as a “continuum of reproductive risk and care-taking casualties” by Arnold Sameroff and Michael Chandler
 2. Research based on transactional model emerged in 1980s; describes risk and protective factors associated with vulnerability and resilience in the face of adversity
 - a. Emmy Werner (1929–1982)—Kauai longitudinal study from birth to adulthood to explore perinatal risk and environmental factors on subsequent developmental outcomes, resiliency
 - b. Michael Rutter (1933–1987)—epidemiological studies of children of mentally ill parents to examine risk and protective factors influencing subsequent psychopathology
 - c. Alan Sroufe’s longitudinal study of competence as a developmental construct
 3. Examples of clinical problems with multifactorial etiology better understood through a transactional perspective include failure-to-thrive, child abuse, attention deficit, hyperactivity, conduct, and eating disorders
- Ecological Model or “Development in Context”
 1. Developed and described by Urie Bronfenbrenner in 1979 publication—*Ecology of Human Development: Experiments by Nature and Design*
 2. Person-place-process model
 - a. Microsystems—immediate settings within which a child spends time during development (e.g., home, school, hospital)
 - b. Mesosystem—relationship or linkages between microsystems (e.g., service coordination)
 - c. Exosystem—settings that may indirectly influence development (e.g., parent’s work place, school boards)
 - d. Macrosystem—broad-based historical, cultural, demographic, and institutional context (e.g., managed care and welfare reform initiatives)
 3. Understanding how and why a child changes or stays the same over time requires examination, not only of the child’s emerging capabilities but the quality of the settings

- (places and processes) where children spend time; communication/linkages between these settings; influence of policy decisions; and overall social, cultural, and political context
- 4. Examples of clinical interventions better understood through an ecological perspective include early intervention services, home visiting services, and care coordination/case management strategies

- Collaborative effort toward integration of scientific knowledge concerning childhood development and implications for policy and practice

▣ INFANT GROWTH AND DEVELOPMENT (BIRTH TO 2 YEARS)

- Physical Domain: Major tasks—physiologic regulation/motor control
 1. Definitions
 - a. Preterm—newborn with gestational age estimated as less than 37 weeks. Late preterm is newborn 34 weeks to $36\frac{6}{7}$ weeks.
 - b. Low birth weight (LBW)— < 2500 g
 - (1) Very low birth weight (VLBW)— < 1500 g
 - (2) Extremely low birth weight (ELBW)— < 1000 g
 - c. Assessment of weight for gestational age
 - (1) AGA—appropriate for gestational age
 - (2) LGA—large for gestational age; weight > 90 th percentile; often associated with diabetic pregnancies
 - (3) SGA—small for gestational age
 - (a) Symmetric intrauterine growth retardation (IUGR)—weight, length and head circumference are SGA; reflects long-standing compromise and/or factors that are intrinsic to the infant such as a syndrome complex
 - (b) Asymmetric intrauterine growth retardation (IUGR)—underweight for length and head circumference; reflects acute compromise extrinsic to fetus such as placental insufficiency
 2. Average U.S. newborn is 19 to 21 inches (48–53 cm), weighs 6 pounds, 2 ounces (2,812 grams) to 9 pounds, 2 ounces (4,173 grams), with head circumference of 13 to 14 inches (33.0 to 35.6 cm) and with chest circumference measuring approximately 2 cm less than head circumference
 3. Initial 8–10% weight loss in average newborn in first 3 to 4 days of life is usually regained by 7 days if formula fed, 14 days if breastfed

4. Weight doubles by 6 months, triples by one year, and quadruples by 2 years
 - a. 5–7 oz (150–210 grams) weight gain per week during first 6 months
 - b. 1.5 pounds (680 grams) during 6 to 12 months
 - c. Average weight gain in second year is 8 to 9 oz (240–270 grams) per month
 - d. By one year, birth weight should be tripled
 5. Length usually increases by 50% by one year, doubles by 4 years and triples by 13 years—increases by 1 inch (2.54 cm) per month during first 6 months then $\frac{1}{2}$ inch (1.3 cm) per month through first year
 6. Head circumference increases $\frac{1}{2}$ inch (1.3 cm) per month during first 6 months then $\frac{1}{4}$ inch (0.65 cm) per month through first year
 7. Serial measurements and observation over time with use of standardized growth charts—<http://cdc.gov/growthcharts/>
 8. Cranial sutures/Fontanelles
 - a. Cranial sutures close during first year of life
 - b. Posterior fontanel closes by 6–8 weeks of age
 - c. Anterior fontanel closes by 12–18 months of age
 9. Dental development
 - a. Formation of teeth begins during the third fetal month and continues through adolescence
 - b. Primary or deciduous teeth are first set of teeth that are later replaced by permanent teeth
 - c. Eruption timing can vary greatly but eruption sequence of deciduous teeth is generally consistent—see **Table 2-1**
 10. Motor development
 - a. Early reflexive responses—involuntary responses to stimuli that may be viewed as precursors to late motor skills
 - (1) Survival reflexes
 - (a) Breathing, hiccups, sneezes, spitting up as infant tries to regulate breathing, sucking, and swallowing
 - (b) Temperature control reflexes—cry, shivering, tucking legs close to body
 - (c) Feeding reflexes—sucking, rooting, crying, and swallowing
 - (2) Nonsurvival reflexes—Babinski, stepping, swimming, grasping, Moro or startle
 - (3) Ontogeny of Infant reflexes
 - (a) Palmar grasp: strongest by 1–2 months, disappears by 3 months
 - (b) Plantar grasp: disappears after 8 months
 - (c) Moro reflex: disappears by 6 months
 - (d) Stepping: disappears before voluntary walking
 - (e) Asymmetric tonic neck (fencing) reflex: diminished by 3–4 months, disappears by 6 months
 - b. Gross and fine motor milestones—age of attainment varies but sequence is generally consistent (see **Tables 2-2 and 2-3**)
- Cognitive Development: Major tasks—sensorimotor and early language development
 1. Vision
 - a. Presence of blink reflex and pupil constriction to light are indications of newborn vision
 - b. Newborns can focus on objects between 4 to 30 inches including mother’s face during feeding
 - c. Binocular vision develops between 4 to 6 months
 - d. Visual acuity is difficult to measure during infancy; distance acuity has been estimated between 20/150 to 20/400 in newborns, improves to 20/70 by 2 years, and 20/30 by 5 years
 2. Hearing
 - a. Infants have greater auditory acuity for high rather than low frequency sounds

■ **Table 2-1** Timetable: Primary Teeth Eruption

Primary Teeth	Maxillary Eruption	Mandibular Eruption
(1) Central incisors	6–8 months	5–7 months
(2) Lateral incisors	8–11 months	7–10 months
(3) Cuspids (Canines)	16–20 months	16–20 months
(4) First molars	10–16 months	10–16 months
(5) Second molars	20–30 months	20–30 months

■ **Table 2-2** Gross Motor Milestones

Milestone	Age
Good head control	2–3 months
Rolls—front to back	4–5 months
Rolls—back to front	5–6 months
Sits alone	5–6 months
Creeps or crawls	7–8 months
Pulls to standing, cruises	9–10 months
Stands alone	11–12 months
Walks—forward	12–14 months
Walks—backwards	14–16 months
Walks—up steps with assistance	16–18 months
Walks—up and down steps alone	22–24 months
Jumps with both feet	24–28 months

Baroni, M. (2004). *Pediatric nurse practitioner certification review guide* (4th ed.). (Virginia Layne Millonig and Caryl Mobley, Eds.). Sudbury, MA: Health Leadership Associates, Inc.

- b. Responsiveness to “motherese” as high-pitched “baby talk”
 3. Sensorimotor development (Piaget)
 - a. Reflexes (birth to 1 month)
 - b. Primary circular reactions (1 to 4 months)—adaptation of reflexes to the environment through coordination of 2 actions such as seeing and grasping
 - c. Secondary circular reactions (4 to 8 months)—increased awareness of objects, persons, and expected responses
 - d. Coordination of means and ends (8–12 months)—object permanence introduces this substage with awareness that people and objects continue to exist when out of sight
 - e. Tertiary circular reactions—active exploration and trial-and-error learning (12 to 18 months)
 - f. Mental combinations—ability to problem-solve simple situations without trial-and-error (18 to 24 months)

■ **Table 2-3** Fine Motor Milestones

Milestone	Age
Grasp and shakes rattle	2–3 months
Reaches for object or person	3–4 months
Brings hands to mid-line	4–5 months
Hand-to-hand object transfer	5–6 months
Unilateral reaching	6–7 months
Raking grasp	6–7 months
Finger feeding	7–9 months
Pincer grasp	8–10 months
Simple games—pat-a-cake	9–10 months
Makes marks on paper with pencil/crayon	10–12 months
Opens book, turns pages	12–13 months
Stacks 2 blocks	12–15 months
Uses eating utensils	15–17 months
Stacks 3 blocks	17–18 months
Simple puzzles—circles shapes first	18–20 months
Stacks 6–7 blocks	22–24 months

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4. Language development—receptive language precedes expressive abilities (see **Table 2-4**)
- Psychosocial Development: Major tasks—development of good fit between temperament and environment; development of secure attachment relationship
 1. Temperament—the “how” of behavior rather than “what” or “why”
 - a. New York Longitudinal Study (NYLS) defined 9 dimensions of temperament—activity, rhythmicity, approachability, adaptability, intensity, threshold of arousal, mood, distractibility, and attention
 - b. Categories of temperamental profiles
 - (1) Easy—rhythmic, approachable, adaptable, positive moods, and low intensity (40%)
 - (2) Slow-to-warm-up—less active, more avoidant, less adaptable, more negative moods, and low intensity (15%)
 - (3) Difficult—arrhythmic, more avoidant, less adaptable, more negative moods, and high intensity (10%)
 - (4) Intermediate high and intermediate low (35%)
 2. Parent-infant interaction and the attachment relationship
 - a. Synchrony—sensitive, coordinated, mutually regulated, and reciprocal style of social interactions that evolve between parent and infant during the first year of life leading to a secure attachment by one year
 - b. Emotional development begins with physiologic experience and expression of “comfort/discomfort” which later differentiates into more fine-tuned emotions
 - (1) Social smile in response to persons begins around 6 weeks
 - (2) Emergence of fears and anxiety accompanies the cognitive milestone of object permanency as the infant distinguishes “mother” from “stranger”; fear of strangers (stranger anxiety) begins around 6 months and peaks about 12 months; separation anxiety begins between 8 to 9 months and peaks around 14 months
 - c. Attachment (definition)—the enduring and specific affective bond that develops over the first year of life
 - (1) Secure attachment—underlying emotion is love
 - (2) Insecure-avoidant—underlying emotion is anger
 - (3) Insecure-anxious—underlying emotion is anxiety/ambivalence
 - (4) Insecure-disorganized—underlying process is confusion/dysfunction

■ TODDLER AND PRESCHOOLER GROWTH AND DEVELOPMENT

- Physical Domain: Major tasks—locomotion and continued motor development
 1. Physical growth

■ Table 2-4 Language Milestones

Milestone	Age
Responds to sounds	Birth
Smiles and coos	2–3 months
Laughs, expresses delight	4–5 months
Babbles	5–6 months
“Dada” & “mama”	8–9 months
Waves “bye-bye”	8–9 months
Understands “no”	9–10 months
2 words other than “mama/dada”	11–12 months
Jabbering	12–13 months
Begins to point to body parts	15–18 months
2-word phrases & sentences	18–22 months
30–50 word vocabulary	22–24 months

- a. Reduced rate of growth between 2 to 6 years resulting in fewer caloric needs and decreased appetite
 - b. Growth in length is approximately 3 inches (7 cm) per year and weight gain approximately 4 $\frac{1}{2}$ pounds (2 kg) per year
 - c. Average U.S. 6-year-old weighs about 46 pounds (21 kg) and is 46 inches (117 cm) tall
2. Motor development
 - a. CNS maturation during toddler and pre-school years allows for better control and coordination of both gross motor and fine motor skills
 - b. Motor development milestones—see **Table 2-5**
 3. Cognitive development—symbolic thinking and increased language development
 - a. Preoperational thinking (Piaget)
 - (1) Preconceptual (2 to 4 years)
 - (2) Intuitive (2 to 7 years)
 - b. Centration—preschoolers’ tendency to focus on one idea or characteristic feature of an object or situation at one time
 - (1) Egocentrism—tendency to focus thinking and understanding about the world from their own perspective only
 - (2) Animism—everything animate or inanimate thinks and feels the way the preschooler does
 - c. Difficulty distinguishing fact from fantasy—normative “lying,” nightmares, imaginary friends, and potential to feel responsible for bad or good things happening based on their own thoughts, feelings, or behaviors
 - d. Language development
 - (1) Expressive language
 - (a) Up to 425 word vocabulary with 75% speech understandable—2 $\frac{1}{2}$ years
 - (b) Increased complexity of sentences including 4 to 5 words—3 $\frac{1}{2}$ years
 - (2) Receptive language
 - (a) Carries out 2 to 3 item commands—3 years
 - (b) Understands opposite analogies—4 years
 - (c) Understands “if,” “because,” and “when”—5 years
 4. Psychosocial development: Major tasks—autonomy, impulse control/discipline, and gender identity
 - a. Parenting styles and discipline
 - (1) Authoritarian parent—strict parenting with firm or harsh discipline and without questioning; high expectations, low support, low parent-child communication
 - (2) Permissive parenting—few demands and low expectations
 - (a) Democratic-indulgent—low expectations with high support and high parent-child communication
 - (b) Permissive-neglectful—low expectations with low support and low parent-child communication
 - (3) Authoritative parenting—firm limits but opportunity for dialogue; high expectations, high support, and high parent-child communication; associated with best-child outcomes
 - b. Aggression and impulse control

■ **Table 2-5** Gross and Fine Motor Milestones

Gross Motor Milestones	Mean Age
(1) Balance on each foot 2 seconds	2 $\frac{1}{2}$ years
(2) Hops	3 $\frac{1}{2}$ years
(3) Walks upstairs alternating feet	3 $\frac{1}{2}$ years
(4) Walks on tip toes	3 $\frac{1}{2}$ years
(5) Heel-to-toe walking	4 $\frac{1}{2}$ years
Fine Motor Milestones	Mean Age
(1) Tower of 8 cubes	2 $\frac{1}{2}$ years
(2) Thumb wiggle	2 $\frac{1}{2}$ year
(3) Copies circle	3 to 3 $\frac{1}{2}$ years
(4) Draws a person with 6 parts	4 $\frac{1}{2}$ years
(5) Copies square	5 years

- (1) Instrumental aggression—common form of aggression among preschoolers focused on retrieving an object, space, or special privilege; frequency decreases with increased understanding of sharing and impulse control
- (2) Hostile aggression—person-oriented aggression that is not common among preschoolers but may emerge with school entry if impulse control remains problematic
- c. Play as the major medium for early mastery of a variety of physical, cognitive, and social skills has been appropriately described as the “work of children”
 - (1) Infancy—solitary play as the earliest level of sensorimotor or skill mastery play; associated with little awareness of other children
 - (2) Toddlers
 - (a) Onlooker play is common at this age with curious watching of other children playing
 - (b) Parallel play—becomes predominant style of play with toddlers engaged in similar play activity but with minimal interaction
 - (3) Preschoolers play—more social in character including:
 - (a) Associative play—some interaction and sharing of toys may occur but not organized and consistent enough to be called a game
 - (b) Cooperative play—children are taking turns and actively playing together
 - (c) Dramatic or pretend play—make believe play during which the children create and act out a scene such as “playing house”
 - (d) Rough-and-tumble play—physical play involving gross motor activities like running, jumping, chasing, and wrestling that appears aggressive but is actually playful
- d. Fears
 - (1) Nightmares
 - (2) Night terrors—partial arousal from deep non-REM sleep with minimal recollection of screaming/thrashing
- e. Gender identity—emerging sense of self as a male or female person
 - (1) 2-year-olds can distinguish gender and will identify themselves as boy or girl

- (2) 3- to 4-year-olds tend to show sex-typed preferences; gender identity is usually firmly established and unlikely to change
- (3) 5- to 6-year-olds begin to express notions about how males and females should dress, behave, and feel

■ SCHOOL-AGED DEVELOPMENT

- Physical Domain: Motor coordination and skill development
 - 1. Physical growth—relatively stable, smooth, and uneventful
 - a. Average school-aged child gains about 5 pounds and $2\frac{1}{2}$ inches per year
 - b. By age 10, the average school-aged child weighs about 70 pounds and is 54 inches tall
 - 2. Motor development—few gender differences except for stronger forearm strength in males and increased flexibility in females
 - 3. Permanent teeth timing and eruption sequence—see **Table 2-6**
- Cognitive Development: Major task—concrete thinking and adaptation to school
 - 1. Concrete operational thought (Piaget)
 - a. “5 to 7 shift”—transition period between preoperational and concrete operational thinking
 - b. Logical operations include:
 - (1) Reversibility—ability to reverse a process or action such as understanding that if $2 + 3 = 5$ then $5 - 3 = 2$
 - (2) Conservation (number, mass, and volume)—understanding that certain aspect or quality of an object can change in appearance without changing the object itself
 - (3) Classification—ability to group objects on the basis of similar characteristics such as color or shape
 - (4) Seriation—arrangement of items in a series, such as by increasing size
 - 2. School readiness
 - a. School entry is a major transition for children
 - b. School refusal may reflect separation anxiety masked as vague somatic complaints
- Psychosocial Development: Major tasks—self-esteem, peers, and after school activities
 - 1. Self-esteem—competence to think, learn, and make decisions as well as believe that one is worthy of love and respectful treatment from others

■ **Table 2-6** Timetable: Permanent Teeth Eruption

Permanent Teeth	Maxillary Eruption	Mandibular Eruption
a. Central incisors	7–8 years	6–7 years
b. Lateral incisors	8–9 years	7–8 years
c. Cuspids (Canines)	11–12 years	9–11 years
First premolar (Bicuspid)	10–11 years	10–12 years
Second premolar (Bicuspid)	10–12 years	11–13 years
First molars	6–7 years	6–7 years
Second molars	12–13 years	12–13 years
Third molars	17–22 years	17–22 years

Baroni, M. *Pediatric Nurse Practitioner Certification Review Guide*, 4th edition. Eds. Virginia Layne Millonig and Caryl Mobley. Health Leadership Associates, Inc., 2004.

2. Peers and development of prosocial behavior through cooperative games, sports, and activities
3. The need for organized after-school activities has become critical with the increasing number of children who are regularly left unsupervised after school because of parental work schedules and variations in family structure in the U.S.

■ ADOLESCENT DEVELOPMENT

- Physical Domain: Major tasks—puberty, sexual maturation
 1. Physical growth
 - a. Average U.S. female gains about 38 pounds (17 kg) and 9 $\frac{1}{2}$ inches (24 cm) between 10 to 14 years.
 - b. Average U.S. male gains about 42 pounds (19 kg) and 9 $\frac{1}{2}$ inches (24 cm) between 12 to 16 years.
 2. Puberty—period of rapid physical growth and sexual maturation resulting in adult size, shape, and reproductive potential
 - a. Sequence of puberty—individual variation in onset but sequence of somatic and physiologic changes is relatively set
 - (1) Females
 - (a) Puberty onset—9 to 10 years
 - (b) Precocious puberty < 8 years
 - (c) Delayed puberty > 13 years
 - (d) First physical sign—breast buds
 - (e) Peak height velocity—12.4 years
 - (f) Menarche—12.5 years
 - (g) Fertility—15 years
 - (2) Males
 - (a) Puberty onset—11 to 12 years
 - (b) Precocious puberty < 9 years
 - (c) Delayed puberty > 14 years
 - (d) First physical sign—testicular growth
 - (e) Peak height velocity—14.4 years
 - (f) Spermatarche—13 to 14 years
 - (g) Fertility—15 years
 - b. Anovulatory cycles are common during the first two years after menarche (50% of cycles vs. 20% after 5 years)
 - c. Stages of genital maturity in males—takes approximately 4 years to move from stage 2 to 5 (Tanner staging)
 - (1) Stage 1: Preadolescent testes, scrotum, and penis
 - (2) Stage 2: Enlargement of scrotum and testes; scrotum reddens and roughens
 - (3) Stage 3: Penis enlarges primarily in length
 - (4) Stage 4: Penis enlarges in breadth and development of glans
 - (5) Stage 5: Adult size and shape
 - d. Stages of breast development in females (Tanner staging)
 - (1) Stage 1: Preadolescent breast with nipple elevation
 - (2) Stage 2: Breast buds with areolar enlargement
 - (3) Stage 3: Breast enlargement without separate contour with nipple
 - (4) Stage 4: Projection of areola and nipple as secondary mound to breast
 - (5) Stage 5: Adult breast with areola receding and nipple projecting from breast
 - e. Stages of pubic hair development in males and females (Tanner staging)
 - (1) Stage 1: Preadolescent without pubic hair
 - (2) Stage 2: Sparse, pale, fine pubic hair
 - (3) Stage 3: Darker, more curled, increased amount of pubic hair

- (4) Stage 4: Hair is adult in character but doesn't cover entire pubic area
- (5) Stage 5: Adult distribution in quantity, quality, and pattern
- Cognitive Development: Major tasks—ability to abstract and decision-making
 1. Formal operational thought—ability to abstract (Piaget)
 2. Characteristic ways of thinking
 - a. Egocentrism—difficulty an adolescent may have in thinking rationally about their own personal experiences as completely unique
 - b. Invincibility fable—sense of invincibility that can lead to risk-taking behaviors
 - c. Personal fable—variation of adolescent egocentrism whereby adolescent feels personally gifted in some way
 - d. Imaginary audience—exaggerated sense that everyone is watching and focused on the adolescent resulting in feeling self-conscious
- Psychosocial Development: Major tasks—independence, intimacy, vocation or career goals
 1. Majority of adolescents cope well with their transition to adulthood and do not experience “storm and stress” as once assumed
 2. A second phase of “separation-individuation” occurs during adolescence that requires new approaches to parenting, communication, decision-making, and independence
 3. Three psychosocial periods of adolescence
 - a. Early adolescence—middle school (11 to 14 years)
 - (1) Importance of peers and feeling “normal”
 - (2) Moodiness
 - b. Middle adolescence—high school (15 to 17 years)
 - (1) Body image, sexuality, dating
 - (2) Asserting independence
 - c. Late adolescence—vocation and career choices (18 to 21 years)
 - (1) Identity formation—achievement, moratorium, foreclosure, diffusion
 - (2) Vocation and career choices including college, military, and employment opportunities
 - (3) Intimacy in relationships

■ THE CHANGING AMERICAN FAMILY AS CONTEXT FOR GROWTH AND DEVELOPMENT

- Demographic Changes
 1. Increasing rates of divorce and remarriage
 2. Delays and declines in child-bearing
 3. Rise in female participation in work force—voluntary, dual earner families, female head of household, and welfare reform work requirements
 4. Increasing incidence of single parent families, childhood poverty, and homelessness
- Variations in Family Structure
 1. Decrease in “traditional” two-parent family
 2. Increase in single-parent households—divorce/separation, births to unmarried mothers, death of spouse, adoption
 3. Remarriage, blended families, and step-parenting
 4. Extended and/or “skip generation” families with grandparents as primary caregivers and/or with several generations living together
 5. Foster families—estimated 510,000 children in foster care at any given time; <http://www.childwelfare.gov/pubs/factsheets/foster.cfm#one>
 6. Group living including homeless shelters—estimated 43% of the homeless population include families with children
 7. Gay and lesbian families—sources vary
- Family Functioning: Critical concepts
 1. Family functioning is more directly related to healthy growth and development than is family structure
 2. Components of family functioning include provision of a stable and safe physical environment as well as financial and emotional resources necessary to provide supportive and nurturing care with appropriate supervision and guidance
 3. Screening and assessment tools
 - a. Family Inventory of Life Events (FILE)
 - b. Family Coping Strategies (F-COPES)
 - c. Adolescent-Family Inventory of Life Events and Changes (A-FILE)
 - d. Parenting Stress Index (PSI)
 4. Family-centered care—Institute for Family-Centered Care
 - a. Recognition that the family is the constant in a child's life
 - b. Family-professional collaboration at all levels

- c. Respect for family diversity in structure, race, ethnicity, culture, and socioeconomic status
 - d. Recognition of family strengths, uniqueness, and diversity in coping strategies
 - e. Communication and sharing of information on an ongoing basis with families in a supportive and nonjudgmental manner
 - f. Support and facilitation of family-to-family support
 - g. Understand and incorporate strategies supportive of growth and developmental needs of children and their families into healthcare settings/systems
 - h. Implementation of policies and programs to support emotional and financial well-being of families
 - i. Accessible health care that is flexible, culturally competent, and responsive to family-centered need
5. Resources: Institute for Family-Centered Care
<http://www.familycenteredcare.org>

■ DEVIATIONS IN PHYSICAL GROWTH AND BEHAVIORAL DEVELOPMENT

Failure-to-Thrive (FTT)

- Definition
 1. No consensus on definition
 2. Descriptive rather than diagnostic term
 3. Generally refers to infants and young children whose weight is below the 3rd percentile on National Center for Health Statistics (NCHS) growth standards and/or whose weight trajectory has decreased by two major growth percentiles
 4. Traditional categories include organic FTT, nonorganic FTT, and mixed etiology FTT
 5. Newer categories include neurodevelopmental FTT and socioemotional FTT
- Etiology/Incidence
 1. Multifactorial etiology, including underlying organic disease or predisposing medical condition, maladaptive parent-infant interaction, maternal depression, poverty, deficits in parenting information and skills, child abuse and neglect
 2. Accounts for between 3% to 5% of all pediatric admissions of infants less than one year, with as many as 50% without underlying medical condition
 3. Males and females are equally affected
- Clinical Findings
 1. Inadequate intake—inadequate milk production, mechanical problems with suck/swallow coordination, systemic disease, errors in formula preparation, misunderstanding about infant needs and feeding practices
 2. Increased losses or decreased utilization—vomiting and/or malabsorption
 3. Increased caloric requirements—underlying disease process including cardiac, respiratory, hyperthyroid, cancer, recurrent infection
 4. Altered growth potential—prenatal insult, genetic disorder, or endocrine dysfunction
- Differential Diagnosis
 1. Organic causes
 - a. Gastrointestinal—gastroesophageal reflux disease (GERD), pyloric stenosis, cleft palate, lactose intolerance, Hirschsprung disease, milk-protein intolerance, hepatitis, malabsorption
 - b. Cardiopulmonary—cardiac defects, bronchopulmonary dysplasia (BPD), asthma, cystic fibrosis (CF), tracheobronchial or tracheoesophageal malformations
 - c. Endocrine—thyroid dysfunction, diabetes, adrenal insufficiency, pituitary disorders, growth hormone deficiency
 - d. Infection from any organism
 - e. Neurologic—mental retardation, fetal alcohol syndrome, lead poisoning, prematurity, neuroregulatory difficulties
 - f. Genetic—mitochondrial diseases
 2. Social-emotional and environmental causes
 - a. Maternal depression or other mental illness, isolation, marital/relationship difficulties
 - b. Poverty/inadequate resources
 - c. Inadequate parenting knowledge and skills
 - d. Difficult temperament
 - e. Child abuse and neglect
- Diagnostic Methods/Findings
 1. History—prenatal, perinatal, neonatal; complete diet history and feeding practices; environmental, social, and family history
 2. Identification of risk factors—prematurity, LBW, difficult temperament, regulation problems, social stresses
 3. Height, weight, head circumference; review longitudinal growth data, corrected for gestational age as appropriate; vital signs including blood pressure
 4. Physical examination—signs of underlying organic disease; severity of malnutrition, evidence of abuse or neglect

5. Developmental assessment and caregiver concerns
 6. Feeding observation to assess behavioral or interactional contributing factors
 7. Home visit or public health nurse referral to assess environmental factors
 8. Laboratory assessment should be judicious based on history and clinical findings; CBC, UA if applicable; comprehensive metabolic panel; other labs as indicated by history, including blood lead levels, TB test; thyroid function; test for reflux/malabsorption; sweat test; stool specimen for parasites; bone age if height is poor
- Management/Treatment
 1. Importance of developing therapeutic alliance with caregiver
 2. Usually managed on an outpatient basis if possible with hospitalization indicated when there is evidence of abuse or severe neglect, severe malnutrition, medical instability, or when outpatient management has failed
 3. Interdisciplinary approach is optimal utilizing health care, nutritional, mental health, and social services
 4. Provide caregivers with necessary information regarding nutritional needs of child and appropriate feeding skills to promote optimal growth
 5. Close monitoring and follow-up on growth and development, social environment, and interdisciplinary/interagency communication

Stuttering

- Definition: Speech dysfluency with initial onset during the preschool years that is characterized by repetitions of sounds, syllables, and/or short words, as well as pauses in timing of speech
 1. Mild or developmental stuttering—speech dysfluency most notable when a preschool-aged child is tired or excited that usually resolves spontaneously if not given excessive attention
 2. Moderate or severe stuttering (sometimes referred to as acquired stuttering)—persistent speech dysfluency that is inappropriate for age and usually has a neurologic and/or psychogenic etiology
- Etiology/Incidence
 1. Multifactorial etiology with a 3:1 male to female ratio of occurrence

2. Prevalence estimated at 1% among prepubertal children with decrease to 0.8% estimate post-puberty
3. Mild developmental stuttering occurs in 4% to 5% of children between 2 to 5 years that spontaneously resolves; approximately 20% of early stuttering does not resolve without intervention

- Clinical Findings
 1. Repetition and/or prolongation of sounds, syllables, or short words
 2. Pauses within words or sentences
 3. Signs of physical tension and struggling with speech such as eye blinking and trembling lips
 4. Avoidance of words that cause particular problems
 5. Child and/or parents are frustrated and/or embarrassed by stuttering posing difficulty at home, in school, or with peers
- Differential Diagnosis
 1. Normal developmental dysfluency
 2. Hearing impairment
 3. Speech-motor deficit
- Management/Treatment
 1. Assess frequency, type, and duration of dysfluency
 2. Encourage parents to avoid excessive attention to dysfluency and be patient in listening to child's speech
 3. Refer to speech and language pathologist for assessment if child is showing signs of embarrassment, speech dysfluency interferes with communication, and/or parent expresses significant concern regardless of severity
 4. Stuttering Foundation of America
3100 Walnut Grove Road, Suite 603
P.O. Box 11749
Memphis, TN 38111-0749
(800) 992-9392
<http://www.stutteringhelp.org/>

Autism Spectrum Disorder

- Definition: Neurobiological disorders characterized by a spectrum of symptoms involving impairment in social interaction, impairment in interpersonal communication, and restricted and repetitive behaviors and interests. Symptoms range in severity but usually manifest themselves within the first three years of life

DSM-IV Classification:

1. Autism Spectrum Disorder (ASD)
 - a. Autistic disorder

- b. Asperger disorder
 - c. Pervasive developmental disorder, not otherwise specified
- 2. Other developmental disorders not considered ASD
 - a. Rett's disorder
 - b. Childhood disintegrative disorder
- Etiology/Incidence
 1. Unclear etiology
 2. In less than 10%, ASD may be associated with another syndrome or disease, such as fragile X, tuberous sclerosis complex, Duchenne muscular dystrophy, Down syndrome
 3. Believed to have a primarily genetic cause but environment may affect the expression of genetic material (e.g., advanced parental age, toxin exposure during early gestation)
 4. No association with vaccines
 5. Considered highly heritable (increased rate in siblings, and even higher in monozygotic twins)
 6. Affects 1 out of every 150 children in the U.S.; male:female ratio—4.3:1
- Clinical Findings
 1. Normal growth and development usually reported until 2 to 2½ years when parents notice delays in language, symbolic or imaginative play, and/or other social interactions; onset of such delay or abnormal behavioral pattern before 3 years of age is considered part of diagnostic criteria
 2. DSM-IV-TR (2000) diagnostic criteria require a total of six behavioral manifestations from three categories including:
 - a. Qualitative impairment in social interaction—poor eye contact; lack of shared enjoyment in activities with peers or family
 - b. Qualitative impairment in communication—delayed or deviant language development; lack of interest in toys, activities for symbolic or imaginative play appropriate for age
 - c. Restrictive repetitive and stereotypic patterns of behavior, interests, and/or activities—repetitive rituals or motor movements such as spinning or hand flapping
 3. Associated problems may include other cognitive delays, problems learning, unusual responses to sensory stimuli, difficulties with sleeping and eating, differences in emotional responsiveness, and seizure activity
- 4. Significant variation in clusters of symptoms and characteristics ranging from mild to severely affected
- Diagnostic Tests/Findings
 1. No specific medical diagnostic test for autism
 2. EEG indicated for associated problems including seizures and language delay
 3. Laboratory studies to assess etiological factors
 - a. Urine amino acids and organic acids; metabolic screen for metabolic disorders such as PKU
 - b. DNA probe for fragile X on blood plasma
 4. MRI or CT scan may show structural abnormalities in cerebellum but otherwise not particularly helpful in absence of clinical signs such as asymmetries, focal tremors, or paralyses
- Differential Diagnosis
 1. Mental retardation
 2. Sensory impairment (hearing or vision)
 3. Severe abuse or neglect
 4. Rett's disorder
 5. Childhood psychosis or other mental illness
 6. Gifted child
 7. Tourette's syndrome
 8. Fragile X
 9. Childhood disintegrative disorder
- Management/Treatment
 1. No specific medical interventions currently available
 2. Screening: Early screening, diagnosis, and referral to early intervention is critical. APA recommends:
 - a. Ongoing developmental surveillance
 - b. Targeted developmental screening at 9, 18, and 30 months
 - c. Autism specific screening at 18 and 24 months or at any time when there is a suspicion of problem
 3. Treatment is primarily psychoeducation requiring individualized plan
 4. Interdisciplinary team to coordinate care including parents, teachers, primary care provider, psychologist, physical therapy, speech and language, and other early intervention staff as appropriate
 5. Address associated problems through specific therapies (sensory integration), counseling (family adjustment, behavioral management), medications (seizures and behavioral problems)
 6. Community education, resource identification, and parent-to-parent support

- a. Autism Society of America
7910 Woodmont Avenue, Suite 300
Bethesda, Maryland 20814-3067
1-800-328-8476
<http://www.autism-society.org/site/PageServer>
- b. CHAT— Checklist for Autism in Toddlers (Baron-Cohen, Cox, Baird, Swettenham, Nightengale, Morgan, Drew, & Chaman, 1996)

Obesity

- Definition
 1. Excess accumulation of body fat relative to lean body mass that results from excessive caloric intake relative to energy expenditure
 2. Body Mass Index (BMI) > 95th percentile
- Etiology/Incidence
 1. Multifactorial etiology with interaction of genetic, environmental, developmental, and behavioral factors
 - a. Genetic predisposition and parental obesity
 - b. Dietary patterns
 - c. Inactivity (television, video games)
 - d. Cultural and familial food preferences
 - e. Use of food as emotional buffer
 - f. Physical disorders with decreased energy expenditure (spina bifida, Down syndrome, Prader-Willi syndrome)
 - g. Endocrinopathy
 2. Most prevalent nutritional problem in U.S.
 3. According to National Health and Nutrition Examination Survey (NHANES) 16.3 % of children and adolescents in the U.S. are obese
 4. Ethnic differences: Non-Hispanic black girls and Mexican-American girls are more likely to be obese than non-Hispanic white girls; among boys, Mexican Americans are more likely to be obese than non-Hispanic white boys
- Clinical Findings
 1. Parent and/or child concern regarding body weight
 2. Clinical observation of large size and/or excess fat on child
 3. Measurement of weight, height, and BMI
- Differential Diagnosis
 1. Endocrine dysfunction
 2. Congenital disorders/short stature
 3. Large frame
 4. Muscular hypertrophy
 5. Medication-induced obesity (including corticosteroids, psychotropic drugs)
- Diagnostic Tests/Findings
 1. History includes detailed dietary and activity level history (past and present); family history of obesity and related morbidities including hypertension, cardiovascular disease, hyperlipidemia, diabetes, depression; review of systems for associated morbidity (glucose intolerance, orthopedic difficulties); developmental milestones; psychosocial concerns about weight
 2. Physical examination, vital signs, blood pressure
 3. Anthropometric measurements
 - a. Weight for height ratio greater than 95th percentile on CDC growth charts is commonly used but doesn't account for large frame or increased muscle mass
 - b. Percent of ideal body weight greater or equal to 120%; calculated by dividing child's actual weight by ideal body weight (50th percentile for age and sex) and multiplied by 100
 - c. Skin-fold thickness per calibrated caliper measurements at or above 85th percentile for age, sex, and race; tricep measurements are most common and reference charts are available
 - d. Body Mass Index (BMI) considered most useful index
 - (1) Weight in kilograms/height in meters²
 - (2) Screening classifications using BMI in adolescents
 - (a) Obesity—BMI equal or greater than 95th percentile for age and gender
 - (b) At risk for obesity—between 85th and 95th percentile or with rapid weight gain of > 2 BMI units in one year
 3. CBC, UA, thyroid function, lipid profile
- Management/Treatment
 1. Prevention in infancy through parent education regarding nutritional needs and feeding strategies
 2. Discuss moderate modification of diet and caloric content while increasing exercise program
 3. Goal for younger child is weight maintenance rather than weight reduction while linear growth catches up; goal for adolescent may include weight reduction if treated after growth spurt
 4. Behavior modification strategies directed at alternative coping measures to deal with stress, maintain motivation, and reinforce regimen

5. Involvement of family in therapeutic program increases likelihood of success
6. Refer to and collaborate with available community resources as appropriate

Child Abuse and Neglect

- Definition: Abuse usually refers to actual “acts of commission” and neglect refers to “acts of omission” although there remains no consensus on specific definitions
 1. First described as “Battered Child Syndrome” by Dr. Henry Kempe in 1962
 2. Legal definitions and reporting requirements vary from state to state but all 50 states have mandated reporting of suspected abuse or neglect by healthcare providers
 3. Categories include physical, sexual and emotional abuse, negligent care, and Munchausen syndrome by proxy (MSP)
 - a. Soft tissue injuries most common—bruises, abrasions, and lacerations
 - b. Head injuries less frequent but cause majority of deaths—“Shaken Baby Syndrome” (altered consciousness with or without signs of head injuries)
 - c. Burns account for 10% of abuse injuries
 - d. Abdominal injuries—usually blunt injuries from hitting or kicking; liver lacerations; kidney/pancreas contusions
 - e. Fractures—rib, spiral, and multiple fractures at same or various ages should trigger suspicion
 - f. Sexual abuse—probably least reported and underdiagnosed
 - g. Munchausen syndrome by proxy (MSP)—disturbed parent-child relationship with fabrication or actual harm to produce symptoms of illness requiring medical attention
- Etiology/Incidence
 1. Etiological factors associated with abuse and neglect
 - a. Perpetrator—history of being maltreated as a child, cognitive or psychiatric impairment, socially isolated, inadequate parenting knowledge and skills, including unrealistic expectations of child
 - b. Victim—unwanted pregnancy, difficult temperament, premature and/or disabled, no significant gender differences
 - c. Social context—violence (family/community), poverty, unemployment, substance abuse
 2. National Child Abuse and Neglect Data System (NCANDS) data from 2007:
 - a. 794,000 children reported to be victims of abuse or neglect (10.6 per 1000 children)
 - b. 32% of victims were under the age of 4, 24% between 4–7 years of age
 - c. Male:female ratio 48.2:51.5
 - d. African-American, American Indian, Alaskan Native, and mixed-race children have significantly higher rates of victimization than Hispanic or white children. Asian children have the lowest rates of victimization
 - e. Neglect (59%) is most common form of maltreatment followed by physical abuse (11%), sexual abuse (7.6%), medical neglect (1%), and multiple maltreatment (13%)
 - f. Most (80%) perpetrators are parents (39% mothers, 18% fathers, 17% both parents)
 3. Child abuse/neglect is a significant cause of pediatric mortality in infants (second to SIDS) and young children (second to accidents)
 - a. 1530 deaths resulting from abuse or neglect were reported in 2006
 - b. Most deaths (84.5%) were of children 6 years or younger
- Clinical Findings
 1. History is vague, inconsistent, and/or incompatible with child’s developmental stage and severity of injury (e.g., bruises on legs if child is not yet cruising)
 2. History may change during course of interview
 3. Delay in seeking medical attention for injury
 4. History of recurrent injuries
 5. Soft tissue injuries with markings characteristic of source of abuse such as hand marks, curved mark of a belt, burn mark in shape of electric iron
 6. Bruises
 7. Burn markings characteristic of immersion
 8. Undernutrition, poor hygiene
 9. Developmental delays
 10. Inappropriate parent-child interaction
- Differential Diagnosis
 1. Unintentional injury
 2. Underlying disease process, e.g., hemophilia, leukemia, osteogenesis imperfecta
 3. Birth marks, Mongolian spots, and/or other variations in skin pigmentation
 4. Folk medicine and cultural practices, e.g., coin rubbing
 5. Sudden Infant Death Syndrome (SIDS)

- Diagnostic Tests/Findings
 1. History to determine and precisely document type of injury, alleged circumstances, and action taken by caregiver
 2. Observation of parent and child for behavioral extremes or exaggerated responses
 3. Physical examination to assess location, type, and characteristic of any lesions or burns for characteristic pattern, shape, or outline
 4. Coagulation studies for severe bruising; radiographs of long bones, ribs, and/or skull series as indicated by history and physical examination; ultrasound for suspected visceral injury
- Management/Treatment
 1. Report of suspected neglect or abuse to child protective services is mandated
 2. Calling the police may also be immediately necessary
 3. Appropriate medical care for child including immediate hospitalization if indicated by severity of injuries
 4. Ensure safety of child, utilizing foster care or relatives if necessary
 5. Assessment of siblings for maltreatment and assurance of safety
 6. Identify and make appropriate referrals to available community resources to facilitate interdisciplinary and interagency collaboration—child protective services, public health, parenting classes, child care/school programs
 7. Education and prevention
 - a. Identify families with risk factors associated with child maltreatment and make referrals to appropriate community-based preventive resources before serious abuse occurs
 - b. Close primary care supervision and acute care follow-up for at-risk families and children
 - c. Mandated reporting of suspected abuse or neglect
 - d. Support community-based child abuse prevention efforts
- Etiology/Incidence
 1. Multifactorial etiology that remains poorly understood but may be associated with:
 - a. Delayed CNS maturation
 - b. Genetic factors
 - c. Prenatal, perinatal, or postnatal trauma or illness
 - d. Dysfunction of catecholamine neurotransmitters
 - e. Male-to-female ratio ranges from 4:1 (general population) to 9:1 (clinic populations)
 2. Controlled studies have not demonstrated evidence of additives, sugar, or salicylates as associated factors
 3. Approximately 3% to 5% of school-aged children meet DSM-IV criteria
- Clinical Findings: DSM-IV-TR (2000) diagnostic criteria for ADHD include:
 1. Six (or more) symptoms of inattention
 - a. Poor attention to detail/careless mistakes
 - b. Difficulty maintaining attention during activities
 - c. Failure to listen even when directly spoken to
 - d. Problems following directions and/or with completion of assignments or tasks
 - e. Disorganized in activities and tasks
 - f. Avoids activities that require focused mental attention
 - g. Frequently loses items necessary for successful completion of task, activity or assignment
 - h. Easily distracted by external stimuli
 - i. Forgetful

or
 2. Six (or more) symptoms of hyperactivity/impulsivity
 - a. Fidgets and squirms
 - b. Difficulty staying seated when expected or appropriate
 - c. Excessive running and climbing
 - d. Difficulty with quiet play activities
 - e. Very high energy and activity level
 - f. Excessively verbal and talkative
 - g. Answers questions abruptly before question is completed
 - h. Difficulty waiting for turn
 - i. Frequently interrupts others and acts intrusively

and
 3. Symptom onset prior to 7 years of age

Attention Deficit Hyperactivity Disorder (ADHD)

- Definition
 1. A behavioral syndrome characterized by a persistent pattern of inattention, poor concentration, impulsivity, and overactivity that exceeds normal developmental variation
 - a. Onset is typically by the age of 3
 2. DSM-IV-TR (2000) defined subtypes
 - a. Attention deficit/hyperactivity disorder—predominantly inattention type

4. Symptoms identified as problem-causing in at least two settings, e.g., home and school
 5. Negative impact of symptoms on social, academic, and/or work performance
 6. Symptoms not attributed to more significant underlying psychiatric or medical disorder
- Differential Diagnosis
 1. Age appropriate for highly active child
 2. Inadequate environments (understimulating or chaotic)
 3. Learning disabilities/sensory impairment
 4. Seizures or mental retardation
 5. Situational anxiety and/or depressive reaction
 6. Oppositional behavior and/or conduct disorder
 - Diagnostic Tests/Findings
 1. History—Perinatal (maternal substance abuse); past (early health problems including ear infections, lead poisoning, iron deficiency anemia, frequent injuries due to activity); present (frequency, severity, and context of symptoms at home and school); social and developmental; parenting style; review of systems
 2. Physical examination, screen for “neurological soft signs”; affective behavior; laboratory data of limited value; CBC, lead screen
 3. Height, weight, blood pressure, vital signs
 4. Vision and hearing screen
 5. Sample behavioral assessment from multiple settings—(home, babysitter/child care, relatives, school) using rating scale of direct observation by different observers (e.g., parents, teachers, and babysitters/daycare providers)
 - a. Connor’s Abbreviated Parent-Teacher Questionnaire
 - b. ADHD Comprehensive Teacher Rating Scale
 - c. Achenbach and Edelbrach’s Child Behavior Checklist
 - d. Edelbrach Child Attention Problem Scale
 - e. DuPaul ADHD Rating Scale for Teachers
 6. Psychological evaluation and cognitive testing
 - Management/Treatment
 1. Provide structured environment—regular routine; clear and simple rules; firm limits; minimize distraction, overstimulation, and fatigue
 2. Behavioral management—formal operant conditioning techniques to reward/reinforce good behaviors; punishment strategies (time-out) or extinction techniques (systematic ignoring) to decrease unacceptable behaviors
3. Evaluate need for mental health referral
 - a. Child-based therapy for depression, anxiety, low self-esteem; cognitive-behavioral training to increase self-control
 - b. Parenting classes or family therapy for relationship difficulties
 4. Pharmacotherapy—unacceptable side effects (including insomnia and growth retardation) and noncompliance are problematic; still no consensus that this is the gold standard for care
 - a. Methylphenidate—effective in 75% to 80% with trial of at least 2 to 3 weeks; give 20 to 30 minutes before meals to maximize effectiveness; avoid p.m. doses to minimize insomnia
 - b. Dextroamphetamine—effectiveness in 70% to 75% with rapid response
 - c. Atomoxetine—non stimulant, causes less insomnia
 - d. If there is a psychiatric comorbidity such as depression or anxiety, treating the comorbidity may have a positive impact on the ADHD
 5. Close follow-up assessment and monitoring of growth and response to medication and behavioral management plan every 3 to 4 months
 6. Ongoing coordination and communication with family, school personnel, primary care provider, and mental health resources is critical to successful management
 7. Medication may be discontinued after 2 to 3 month trial if no change in behavior
 8. Non-conventional treatments (no documented evidence of effectiveness in controlled studies)
 - a. Megavitamins, mineral therapy, fish oil, acetyl-l-carnitine
 - b. Elimination of sugar, additives, coloring
 - c. Neurophysiologic interventions (patenting, sensory integration, optometric training)
 9. Community resources
 - a. Children and Adults with Attention Deficit/Hyperactivity Disorder (CHADD) International non-profit parent support organization
8181 Professional Place—Suite 150
Landover, MD 20785
301-306-7070
<http://www.chadd.org/>
 - b. Attention Deficit Disorder Association
PO Box 7557
Wilmington, DE 19803-9997
(800) 939-1019
<http://www.add.org>

Aggression, Defiance, and Disruptive Behavioral Disorders

- Normal behavior vs. dysfunctional patterns and clinical disorders
 1. Most children manifest some degree of developmentally normative aggressive, defiant, and disruptive behavior during infancy and early childhood, e.g., breath-holding, temper tantrums, lying, fighting, breaking things (i.e., some of this behavior is normal)
 2. Almost half of all parents consult with primary care providers regarding difficulty managing disruptive and defiant behaviors of their preschoolers
 3. Repetitive and persistent patterns of aggressive, defiant, and disruptive behaviors lasting over 6 months warrant detailed assessment and possible mental health referral

- Conduct Disorder: A repetitive and persistent dysfunctional pattern of aggressive behavior and/or violation of the law, social norms, and basic human rights
 1. Etiology/Incidence
 - a. Multifactorial etiology—biological-genetic component suggested from twin/adoption studies
 - b. Child onset (less than 10 years) has graver prognosis with increased risk of later substance abuse and antisocial personality disorder than adolescent-onset
 - c. Incidence greater among males with estimates of 6% to 16%; female estimates of 2% to 9%
 2. Clinical Findings—DSM-IV-TR (2000) criteria include:
 - a. Aggressive/threatening behavior to people or animals
 - b. Deliberate, intentional destruction of property
 - c. Lying, stealing
 - d. Serious rule violations such as staying out all night, running away, truancy
 3. Differential Diagnosis
 - a. Adjustment disorder
 - b. ADHD
 - c. Oppositional-defiant disorder
 - d. Manic episode
 - e. Other psychiatric diagnosis
 4. Diagnostic Tests/Findings
 - a. Separate interview with parent and child
 - b. Assessment of parental anxiety regarding dependency/control issues; impact on family functioning

- Oppositional-Defiant Disorder: A repetitive and persistent dysfunctional pattern characterized by negative, disobedient, defiant, and hostile behavior directed at authority figures
 1. Etiology/Incidence
 - a. Multifactorial etiology associated with difficult temperament; disruption in early care-giving environment; harsh, inconsistent, and/or neglectful parenting; history of psychiatric disorder in at least one parent, including maternal depression
 - b. Incidence among males is greater than females until puberty, after which rates become more equal; overall estimates range from 2% to 16% depending on population
 2. Clinical Findings
 - a. Loses temper easily
 - b. Argumentative with adult authority figures
 - c. Actively defiant of rules and adult requests
 - d. Deliberately annoys other people
 - e. Blames others for mistakes and misbehavior
 - f. Edgy and easily annoyed
 - g. Frequently resentful or angry
 - h. Spiteful and vindictive behavior
 3. Differential Diagnosis
 - a. Within normal range of oppositional behavior for age and developmental level
 - b. Conduct disorder
 - c. ADHD
 - d. Mental retardation
 4. Management/Treatment
 - a. Early identification and monitoring of defiant, aggressive, and oppositional behaviors
 - b. Parenting education, support, and effective discipline
 - c. Refer for family therapy/intervention if child manifests four or more of clinical

- Management/Treatment
 - a. Identify and communicate concern for child and family well-being
 - b. Refer for psychological/psychiatric evaluation and intervention
 - c. Refer to social service and/or other community resources for parenting education, support, and to reduce family stress and potential for violence

behavioral features that have persisted for longer than 6 months

Learning Disabilities (LD)

- Definition
 1. Generic term referring to a heterogeneous cluster of disorders manifested by significant difficulties in the acquisition and use of language, listening, reading (dyslexia), writing (dysgraphia), reasoning, or mathematical abilities (dyscalculia)
 2. School performance in deficit areas significantly below that expected for age, grade, and level of intelligence
- Etiology/Incidence
 1. Multifactorial etiology of genetic and environmental factors
 2. Possible abnormal function in parietal and/or occipital lobes of brain
 3. Conditions associated with LD
 - a. Developmental delay
 - b. Lead poisoning
 - c. Fetal alcohol syndrome
 - d. Fragile X syndrome
 - e. May be part of syndrome complex (e.g., ataxia telangiectasia, DiGeorge syndrome)
 4. General prevalence estimated at 6% to 11% of school-age children
 5. Dyslexia—most common LD, affects 2% to 10% of general population
- Clinical Findings
 1. Specific academic skill deficits
 - a. Basic reading skills, reading comprehension, spelling
 - b. Mathematical calculations and reasoning
 - c. Disorders of written expression and writing skills
 2. Perceptual-motor impairments
 - a. Distinguishing shapes and sizes
 - b. Fine motor skills, e.g., writing, coloring, cutting
 - c. May make letter and number reversals
 3. Memory and thinking impairment (integrative processing)
 - a. Haphazard, ineffective study habits and strategies for memorization
 - b. Sequencing of data
 - c. Understanding abstract concepts, e.g., time, space, parts, whole
 - d. Lacking skills for effective problem-solving and task completion
- 4. Speech and language deficits
 - a. Language delay
 - b. Difficulty with grammar (syntax), meaning (semantics), and/or social use of words (pragmatics)
- 5. Attention deficits—difficulty concentrating and staying on task
- 6. Hyperactivity—difficulty sitting still, constantly in motion
- 7. Impulsiveness
 - a. Often acting without thinking
 - b. Poor planning skills
 - c. Lack of self-regulation skills
- 8. General deficits in coordination—clumsiness
- 9. Emotional problems
 - a. Lability and moodiness
 - b. Often isolated or rejected by peers
 - c. May exhibit inappropriate attention-getting behaviors
 - d. Difficulty reading nonverbal social cues
 - e. May be passive learners
- Differential Diagnosis
 1. Undiagnosed sensory impairment—vision or hearing deficits
 2. Attention deficit hyperactivity disorder (ADHD)
 3. Seizure disorder
 4. Mental retardation
 5. Maladaptation to chronic disease
 6. Social and environmental factors
 - a. Child abuse and neglect
 - b. Situational anxiety or depressive reaction
 - c. Ethnic or cultural minority
- Physical Findings
 1. Neurological soft signs commonly present
 - a. Poor fine motor coordination and tactile discrimination
 - b. Strabismus
 - c. Poor hand-eye coordination
 - d. Balance problems
 2. Phenotypic features of associated conditions—FAS, fragile X
 3. Other abnormal physical findings likely to be related to LD
- Diagnostic Studies/Findings: Diagnosis requires multidimensional assessment
 1. Complete medical and social history
 2. Developmental and behavioral history
 3. Educational history and school functioning
 4. Physical examination—soft neurological signs
 5. Laboratory studies—associated conditions
 - a. Lead screening
 - b. EEG if seizures suspected

6. Psychoeducational testing
 7. Analysis of history, test results, and academic achievement leads to diagnosis
- Management/Treatment
 1. Developmental surveillance, early identification, and referral is critical
 2. Early intervention to optimize learning and minimize emotional sequelae
 3. Thorough psychoeducational evaluation to determine skills and deficits
 4. Interdisciplinary conference to evaluate findings (M-team)
 5. Individualized education plan (IEP) developed based on multidimensional assessment
 6. Yearly reevaluation of IEP with revisions as needed
 7. Address other associated behavioral, social, and family issues through counseling and direct instruction
 8. Inform parents of legal rights under Individuals with Disabilities Education Act (IDEA) and availability of appropriate special services

Eating Disorders: Anorexia Nervosa and Bulimia Nervosa

- Definition: Chronic and often severe disturbances in eating behavior accompanied by distorted perception of body weight, size, and shape
 1. Anorexia nervosa—eating disturbance associated with weight loss and refusal to maintain body weight at minimally normal level (85% of expected body weight for age and sex; with subsequent amenorrhea in girls)
 2. Bulimia nervosa—eating disturbance associated with episodic binge eating followed by compensatory efforts to prevent weight gain (self-induced vomiting; dieting; fasting; excessive exercise or misuse of laxatives, enemas and/or diuretics)
 3. Anorexia may occur with or without associated bingeing/purging (restricting vs. binge-eating/purging types); bulimia may occur with or without purging (purging vs. nonpurging types)
- Etiology/Incidence
 1. Family enmeshment hypothesis—rigid, overprotective families with difficulty with conflict resolution; separation-individuation
 2. Fear of sexual maturation; history of sexual abuse
 3. Social pressure to be thin
 4. Ballet dancers and gymnasts at particular risk
- Associated with psychological profile of low self-esteem in spite of outward successfulness.
- 6. Both disorders are more common among females
- 7. Anorexia affects approximately 1% of white middle/upper class females; 5% to 10% of cases are male often associated with gender identity conflict
- 8. Bimodal distribution with one peak at 14 years and second at 18 years
- 9. Bulimia is more common than anorexia with later age onset
- 10. Suicide rates of 2% to 5% of those with chronic anorexia with overall mortality rate as high as 10%
- 11. Normal to high intelligence, overachievers, perfectionists
- Clinical Findings
 1. Self-imposed weight loss
 2. Anemia, jaundice, and secondary amenorrhea
 3. Vigorous exercise regimen to increase weight loss
 4. Constipation (chronic laxatives) and reflux esophagitis (self-induced vomiting)
 5. Dry skin, brittle nails
 6. Lower body temperature, blood pressure, and heart rate
 7. Lanugo
 8. Sore throat, calluses on dorsum of fingers, loss of tooth enamel (from induced vomiting)
- Differential Diagnosis
 1. General medical condition—gastrointestinal disease, diabetes, thyroid disorder, AIDS, systemic lupus erythematosus
 2. Pregnancy
 3. Depressive disorder or substance abuse
- Diagnostic Tests/Findings
 1. History—include nutritional patterns as well as effort to lose weight (dieting, exercise, vomiting); preoccupation with food and/or “feeling fat”; past medical, family, and social history; review of systems (amenorrhea)
 2. Weight and height percentiles—degree of malnutrition determined as percentage below ideal weight (IBW)
 - a. Mild malnutrition— < 20% below IBW
 - b. Moderate malnutrition—20% to 30% below IBW
 - c. Severe malnutrition— > 30% below IBW
 3. Physical examination—signs of malnutrition; dry skin, brittle nails, muscle weakness, flat affect, decreased blood pressure, pulse and body temperature; Tanner staging delays

4. CBC, serum albumin, glucose, electrolytes, thyroid function, ECG; others as appropriate based on history and clinical findings
- Management/Treatment
 1. Interdisciplinary treatment plan including nutritional intervention, behavior modification techniques, psychotherapy (individual, family, and/or group therapy); pharmacologic management with antidepressants if appropriate
 2. Hospitalization for rehydration, refeeding, and/or psychiatric treatment if condition warrants. As the family is a contributing factor, hospitalization as a means to remove the child from the family is sometimes effective
 3. Approximately one half of patients show varying degrees of improvement, 25% show long-term improvement, 25% do poorly regardless of intervention
 4. Refer to appropriate community resources for assessment tool, support, and education: Eating Disorder Inventory-2
 - a. Psychological Assessment Resources, Inc. PAR, Inc.
16204 North Florida Avenue
1-800-331-8378
 - b. Foundation for Education about Eating Disorders
P.O. Box 1637
Baltimore, MD 21210
(410) 467-0603
<http://www3.parinc.com/>
 - c. National Eating Disorders Association
603 Stewart Street, Suite 803
Seattle, WA 98101
(800) 931-2237
<http://www.nationaleatingdisorders.org/>
 - d. National Association of Anorexia Nervosa and Associated Disorders
P.O. Box 7
Highland Park, IL 60035
(847) 831-3438
<http://www.anad.org/>

Childhood Depression

- Definition: Behavioral pattern lasting at least two weeks that is characterized by affective and behavioral symptoms including sad or tearful moods, irritability and/or social withdrawal with associated decreased interest and pleasure in developmentally appropriate activities
- Etiology/Incidence: Multifactorial etiology with associated risk factors including:
 1. History of traumatic event(s) involving significant separation(s) or loss(es) of parent, caregiver, or significant other
 2. Family history of depression, especially in mother; evidence of genetic component from twin and adoption studies
 3. Chronic neglect and lack of nurturance due to family disruptions or dysfunction including long-term effects of poverty and/or homelessness
 4. Substance abuse, physical or sexual abuse in household
 5. Chronic illness and/or disability may increase risk, especially with familial predisposition
 6. Estimated incidence of 3% for overall pediatric population and as high as 9% for adolescents; no gender differences in early childhood depression but increases among females in adolescence with ratio of 5:1
- Clinical Findings
 1. Major depressive episode—significant distress and/or interference with normal daily functioning lasting at least 2 weeks associated with:
 - a. Depressed or irritable mood and/or decreased interest and pleasure in developmentally appropriate activities and a minimum of 4 additional symptoms
 - b. Appetite changes with associated weight gain or loss
 - c. Insomnia or hypersomnia
 - d. Difficulty concentrating; decline in school performance
 - e. Feelings of worthlessness, guilt, fearfulness, isolation
 - f. Social withdrawal from friends, family; school refusal and/or truancy
 - g. General somatic complaints of aches and pains with nonspecific etiology, e.g., fatigue, headaches, stomachaches
 - h. Agitation, irritability, and/or disruptive behavior
 - i. Recurrent thoughts of death and/or suicidal ideation
 2. Dysthymic disorder—long-standing depressed or irritable mood lasting one year or more but symptoms of distress and interference with normal daily functioning not as pronounced as a major depressive episode
 3. Adjustment disorder with depressed mood—depressed or irritable mood, sadness, tearfulness, and/or feelings of hopelessness causing some degree of impairment of daily functioning that occurs within 3 months of a significant and identifiable stressful life event

- Differential Diagnosis
 1. Normal periods of sadness and/or mood swings
 2. Acute depressive reactions/adjustment disorder with depressed mood in response to identifiable life stress
 3. Masked depressive disorder—somatization and denial of feelings
 4. Underlying physical disorder
 5. Substance abuse
 6. Psychiatric depressive disorder with suicidal risk
- Diagnostic Tests/Findings
 1. Behavioral symptoms according to DSM-IV-TR diagnostic criteria
 2. History—developmental, family, social, and school; current medications; chronic disease/disability
 3. Separate interview with child or adolescent is essential
 4. Physical examination—neurological screening
 5. Height, weight, vital signs, assess for recent weight loss or gain
 6. Laboratory tests—as indicated by history (drug screen, pregnancy test)
- Management/Treatment
 1. Early screening and identification of children and adolescents at risk for depression
 - a. Center for Epidemiologic Studies Depression Scale for Children
 - b. Children's Depression Rating Scale
 - c. Children's Depression Inventory
 - d. Child Behavior Checklist
 - e. Rose Institute Adolescent Depression Scale
 - f. Mood Questionnaire for Adolescents
 2. Evaluate severity of depression including suicidal risk and make appropriate referrals
 3. Psychiatric intervention is necessary for major depressive episode or dysthymic disorder
 - a. Family and/or individual psychotherapy
 - b. Pharmacotherapy—may be included as part of multimodal treatment plan
 - (1) Selective serotonin reuptake inhibitors (SSRI)—most effective medication for major depressive episodes (fluoxetine or sertraline)
 - (2) Tricyclic antidepressants have limited clinical use with equivocal effectiveness demonstrated in controlled studies (monitor blood levels)
 4. Supportive counseling for adjustment disorder with depressed mood

5. Resources
 - a. Depression/Awareness, Recognition, and Treatment (D/ART)
National Institutes of Mental Health
Science Writing, Press, and Dissemination Branch
6001 Executive Boulevard, Room 8184,
MSC 9663
Bethesda, MD 20892-9663
1-866-615-6464
<http://www.nimh.nih.gov/index.shtml>

Suicidal Behavior

- Definition: Passive or active thoughts/wishes about death and dying (suicidal ideation); talking and/or threatening to take one's life or self-injury without intent to die (suicidal gesture); deliberate self-injury with the intent to die but not resulting in death (suicide attempt); and self-inflicted death (completed suicide)
- Etiology/Incidence
 1. Accounts for 10% of teenage deaths representing second leading cause of adolescent mortality
 2. 15% to 40% of completed suicides were preceded by one or more suicide gestures/attempts
 3. Females have higher rates of suicide attempts; males have higher rates of completed suicides
 4. Higher rates among Native American, Asian, and chronically ill adolescents
 5. Ingestion of medication is most common method; violent methods (hanging and shooting) are more frequently used by males than females and are more often fatal
- Clinical Findings
 1. Severe and/or chronic depression
 2. Hopelessness
 3. Previous suicidal gestures
 4. History of suicide in family
 5. Existence of specific plan
- Differential Diagnosis
 1. Unintentional injuries due to carelessness and/or adolescent sense of "invincibility"
 2. Suicidal gesture as desperate call for help
 3. Imminent risk/acute suicidal intent
 4. Psychotic episode
- Diagnostic Tests/Findings
 1. History—past and present health, chronic depression, chronic illness, or disability;

- school performance; family history of depression and/or suicide; current medications; substance abuse
- 2. Suicidal risk—suicidal ideation, extent of premeditation and existence of plan, likelihood of rescue, suicide notes, previous suicidal gestures
- 3. Observation and/or reports of suicidal behavior by family, teachers, or peers
- Management/Treatment
 1. Refer immediately for crisis intervention resources, e.g., 24-hour hotline
 2. Suicidal ideation and/or gestures with existence of plan requires immediate psychological evaluation
 3. Assure safe environment for child/adolescent including hospitalization if necessary
 4. Short-term hospitalization is recommended for all suicide attempts; attending to emergency treatment and/or surgical management is necessary but insufficient without additional follow-up intervention
 5. Treat underlying depression
 6. Inform child/adolescent of seriousness of concern and need to notify family and mobilize necessary community resources
 - a. National Adolescent Suicide Hotline
1-800-621-4000
<http://www.helpingteens.org/resources/suicide/national-adolescent-suicide-hotline.html>
 - b. American Association of Suicidology
5221 Wisconsin Avenue, NW
Washington, DC 20015
<http://www.suicidology.org/web/guest/home>
(202) 237-2280

Substance Abuse (Tobacco, Alcohol, and Other Drugs)

- Definition: Use of any drug or chemical for purposes of stimulation, pleasure, or in a way that interferes with normal functioning and/or threatens health; includes misuse of legal “recreational” drugs, prescribed or nonprescribed medications, as well as illegal substances; DSM-IV-TR distinguishes substance abuse from dependence
 1. Substance abuse—a maladaptive pattern of substance use associated with significant impairment or distress, including inability to meet expected daily obligations, use of substance in situations/context that may be hazardous to self or others and/or resulting in legal and/or other interpersonal conflicts
- 2. Substance dependence—a pattern of repeated substance abuse that results in tolerance, withdrawal, and/or compulsive use that can be psychological and/or physiologic-based
- 3. Categories of substances include—alcohol, marijuana (cannabis), nicotine, amphetamines, caffeine, cocaine, hallucinogens, inhalants, opioids, phencyclidine (PCP), sedatives and hypnotics, anabolic steroids
- Etiology/Incidence
 1. Multifactorial etiology including some evidence of biological predisposition along with psychosocial and environmental risks such as impulsivity, non-conformity/rebellion, peer pressure, ineffective coping with stress, undiagnosed depression, family dysfunction, history of child abuse or neglect, parental substance abuse
 2. Majority of adolescents will engage in some form of drug use at some point
 3. More frequent use of all substances among males vs. females
 4. Highest overall incidence of substance abuse among Caucasian teenagers followed by Hispanic youth; lowest incidence among African-American teenagers
 5. Most recent national drug use survey (2007) reported increasing prevalence of current illicit drug use—9.5% teenagers from 12 to 17 years of age
 6. Trend is away from street drugs toward prescription drugs used illicitly
- Clinical Findings
 1. Nicotine—decreased exercise tolerance, fatigue, muscle weakness; pallor, tachycardia, staining of teeth, tobacco odor on breath and clothes
 2. Alcohol—initial euphoria and talkativeness; grogginess; impaired short-term memory; decreased reaction time; hypoglycemia
 3. Marijuana (THC, pot, cannabis, joint, reefer, weed, hash, grass)—euphoria, drowsiness, slowed reaction time and motor coordination, time distortions, tachycardia and transient hypertension, bloodshot eyes
 4. Amphetamines—dilated pupils, tachycardia, anorexia, insomnia, weight loss, anxiety, and suicidal behavior
 5. Cocaine (coke, freebase, crack, nose, flake)—agitation, hyperactivity, euphoria followed by depression, confused thinking, occasional paranoid ideation, tachycardia, habitual “snorting” induced nasal septum scabbing or necrosis

6. Hallucinogens
 - a. LSD—dilated pupils, visual and auditory hallucinations and flashbacks, disorganized and confused thinking, increased attention to stimuli; chronic use can lead to psychosis and major personality changes
 - b. PCP—euphoria, motor incoordination, hallucinations; paranoia with aggressive/violent behavior
 7. Inhalants/volatile substances (glue, cleaning agents, hydrocarbons)—relaxation, hallucinations, light headedness, giddiness; seizures, coma, cardiac arrhythmias, and sudden death
 8. Anabolic steroids—used to increase muscle mass and strength; fluid retention, mood swings, menstrual abnormalities; male gynecomastia, female hirsutism, breast atrophy
 9. Opiates include naturally occurring (e.g., morphine, codeine), semisynthetic (e.g., heroin, dilaudid), and synthetic (e.g., fentanyl, meperidine, methadone); constricted pupils; respiratory depression; euphoria; analgesia; dermatologic lesions, “tracks”; tattoos in unusual places to conceal track marks; chronic infections (skin, HIV, scarring and cellulitis); constipation; decrease in libido; urinary retention
- Differential Diagnosis
 1. Social recreational use of legal substances
 2. Experimentation vs. abuse
 3. Chronic depression
 4. Neurological disorder
 5. Learning disabilities
 - Diagnostic Tests/Findings
 1. History—past and present, environmental, family, social, and academic history; review of systems; current medications; specific drug history including specific drugs, frequency of use, settings of use, impairment of daily functioning including suspensions or legal difficulties
 2. Interviews with and observations from child/adolescent, parents, school personnel, peers
 3. Physical examination with close assessment of skin integrity (nasal septum, skin lesions/track marks); neurological assessment; vital signs, weight, height, blood pressure
 4. Serum and urine toxicology, HIV testing, others as appropriate to specific history
 - Management/Treatment
 1. Assure appropriate privacy, confidentiality, and nonjudgmental atmosphere

2. Referral to appropriate substance abuse treatment resources
3. Identify and provide appropriate referrals for management of underlying psychosocial difficulties contributing to substance abuse
4. Educate and counsel regarding legal and physical risks of substance abuse
5. Support community-based prevention programs
6. Additional National Resources
 - National Institute on Drug Abuse (NIDA)
(800) 662-HELP
<http://www.nida.nih.gov/>
 - National Family Partnership
<http://www.nfp.org/>
(800) 705-8997
 - National Clearinghouse for Alcohol and Drug Abuse Information
<http://ncadi.samhsa.gov/>
(800) 729-6686

■ HEALTH SUPERVISION AS ONGOING SURVEILLANCE, SCREENING, AND ASSESSMENT OF PHYSICAL GROWTH AND BEHAVIORAL DEVELOPMENT

- Definitions
 1. Surveillance: A continuous process of periodic assessment and monitoring of growth and development overtime through a variety of methods including direct observation, health history, parent/child interviews, and physical examination
 2. Screening: Use of standardized or generally accepted methods with essentially well populations in order to identify individuals who may be at risk for physical, cognitive, or psychosocial abnormality and warrant further assessment; good screening tools are simple, inexpensive, acceptable, valid, and reliable
 3. Assessment: A more systematic evaluation using a standardized or generally accepted method leading to recommendations for intervention
 4. Sensitivity: Proportion of those with the abnormality who are correctly identified through screening (true positives)
 5. Specificity: Proportion of those without the abnormality who are correctly identified as negative through screening (true negatives)
 6. Positive predictive value (PPV): Proportion of those individuals correctly screened as positive of all those who actually have the abnormality

- Examples of Screening and Assessment Tests Used in Child Health Supervision
 1. Physical assessment and laboratory screening (refer to chapter 3 on Health Promotion)
 2. Developmental screening and assessment
 - a. Global development
 - (1) Newborn Behavioral Assessment Scale (NBAS)—assessment of newborn’s behavioral capacities including state control, autonomic reactivity, reflexes, habituation, and responsiveness to visual and auditory stimuli
 - (2) Bayley Infant Neurodevelopmental Screener (BINS)—screens for basic neurological, receptive, expressive, and cognitive functions in infants between 3 to 24 months
 - (3) Bayley Scales of Infant Development, Second Edition (BSID-II)—current “gold standard” for diagnosing developmental delays and recommending intervention for children birth through 42 months with separate mental, motor, and behavioral rating scales
 - (4) Ages and Stages Questionnaires (ASQ)—parent-completed child monitoring system for children 4 to 48 months
 - (5) Denver II—screens in personal-social, fine motor-adaptive, language, and gross motor domains in children birth to 6 years
 - (6) First Step—screening test for evaluating preschoolers
 - b. Cognitive development-intelligence
 - (1) McCarthy Scales of Children’s Abilities
 - (2) Weschler Preschool and Primary Scale of Intelligence Revised (WPPSI-R)
 - (3) Weschler Intelligence Scale for Children-WISC III
 - c. Language
 - (1) Early Language Milestones Scale (ELM)—(0 to 42 months)
 - (2) Receptive and Expressive Emergent Language Scale (REEL)—(0 to 36 months)
 - (3) Clinical Linguistic and Auditory Milestone Test (CLAMS)—(0 to 36 months)
 - (4) Language Development Survey—screening tool for toddlers using vocabulary checklist for enumeration of words
 - (5) The MacArthur Communicative Development Inventory—words and gestures
 - (6) The MacArthur Communicative Development Inventory—words and sentences
 - d. Behaviors
 - (1) Achenbach’s Child Behavior Checklist (ACBCL)
 - (2) Connor’s Abbreviated Parent-Teacher Questionnaire
 - e. Temperament
 - (1) Infant Temperament Questionnaire—4 to 8 months
 - (2) Toddler Temperament Scale—1 to 3 years
 - (3) Behavioral Style Questionnaire—3 to 7 years
 - (4) Middle Childhood Temperament Questionnaire—8 to 12 years
 3. Parent-child relationship and home environment
 - a. Parenting Stress Index (PSI)
 - b. Home Observation for Measurement of the Environment (HOME scale)—infant, preschool, and elementary school versions
 - c. Pediatric Review and Observation of Children’s Environmental Support and Stimulation Inventory (PROCESS)
 - d. Nursing Child Assessment Feeding (NCAF) and Teaching (NCAT) scales
 4. Mental health screening and diagnostic classifications
 - a. 0–3 Diagnostic Classification of Mental Health and Developmental Disorders of Infancy and Early Childhood (1995)
 - b. DSM-PC Classification of Child and Adolescent Mental Diagnoses in Primary Care (1996)
 - c. DSM-IV-TR Diagnostic and Statistical Manual of Mental Disorders (2000)

■ QUESTIONS

Select the best answer

1. Most stage-based theories of development focus primarily on:
 - a. The continuity of development
 - b. The discontinuity of development
 - c. Persistence of inherent personality characteristics
 - d. The influence of context on development
2. The common practice of using “time-outs” with young children is a direct application of:
 - a. Operant conditioning
 - b. Classical conditioning
 - c. Separation-individuation
 - d. Maturation reinforcement

3. Good communication between families, schools, and primary care providers is an example of which ecological concept?
 - a. Microsystem
 - b. Mesosystem
 - c. Exosystem
 - d. Macrosystem
4. Which of the following findings would be most likely associated with asymmetric intrauterine growth retardation?
 - a. Weight, length, and head circumference ranging from 3rd to 5th percentile
 - b. Heavy maternal smoking throughout pregnancy
 - c. Weight at 3rd percentile and length at 25th
 - d. Gestational diabetes
5. Early reflexive responses that are not related to survival include all but:
 - a. Babinski
 - b. Moro
 - c. Swimming
 - d. Rooting
6. The most likely weight of a one-year-old child whose weight at birth was $6\frac{1}{2}$ pounds would be:
 - a. 19–20 pounds
 - b. 13–14 pounds
 - c. 25–26 pounds
 - d. Impossible to estimate
7. One of the major psychosocial tasks of infancy is:
 - a. Development of secure attachment
 - b. Separation-individuation
 - c. Symbiosis
 - d. Regulation
8. Which developmental theory best explains the multifactorial etiology of failure-to-thrive?
 - a. Organismic-maturational theory
 - b. Social learning theory
 - c. Transactional theory
 - d. Psychoanalytic theory
9. Most healthy infants are able to reach, grasp, and hold onto a rattle or other small toy by about:
 - a. 2 months
 - b. 6 months
 - c. 8 months
 - d. 10 months
10. The pincer grasp is a fine motor skill that involves the ability to pick up a small object such as a raisin or piece of cereal with the thumb and forefinger that usually is mastered around:
 - a. 4 months
 - b. 6 months
 - c. 9 months
 - d. 16 months
11. You would be concerned about the language development of a child who:
 - a. Repeats simple phrases at 32 months
 - b. Stutters when excited or tired at 42 months
 - c. Has a vocabulary of 10 words at 12 months
 - d. Pronounces words that are not understandable at 36 months
12. The most common temperamental profile is:
 - a. Easy
 - b. Difficult
 - c. Slow-to-warm-up
 - d. Intermediate
13. The underlying emotion of an insecurely attached (avoidant) relationship is:
 - a. Ambivalence
 - b. Deprivation
 - c. Anger
 - d. Conditional love
14. The stage of cognitive development that Piaget described as characteristic of the way preschoolers think is the:
 - a. Preoperational stage
 - b. Mental combinations stage
 - c. Tertiary circular function stage
 - d. Sensorimotor stage
15. A preschool boy whose parents have separated and are beginning divorce procedures:
 - a. May think that he caused the divorce by misbehaving
 - b. Should not be told of the impending divorce until the parents are sure of their decision
 - c. Is likely to experience gender identity confusion
 - d. Should be able to make a decision about which parent he prefers living with
16. Which behavior would you expect to decrease during the preschool years?
 - a. Rough-and-tumble play
 - b. Instrumental aggression
 - c. Hostile aggression
 - d. Cooperative play

17. A preschool child who says that the sky is blue because it is his favorite color is illustrating the concept of:
 - a. Symbolic thinking
 - b. Egocentrism
 - c. Centration
 - d. Imaginary audience
18. Which of the following strategies would not be appropriate to include as part of your management of a 9-year-old boy who is obese?
 - a. Referral to nutritionist for weight reduction plan
 - b. Increase physical exercise
 - c. Behavior modification strategies to deal with stress and/or reinforce treatment plan
 - d. Involve family in management program
19. Which of the following issues or concepts is relevant to the school-aged child?
 - a. Operational thinking
 - b. Initiative
 - c. Concrete operations
 - d. Separation-individuation
20. The first physical sign indicating the onset of female puberty is:
 - a. Sparsely distributed fine, pale pubic hairs
 - b. Breast buds
 - c. Menarche
 - d. Peak height velocity
21. Which of the following findings would be helpful in distinguishing obesity vs. large body frame in an adolescent who is concerned with her weight?
 - a. Tricep skinfold measurement
 - b. Weight-for-height ratio
 - c. Body Mass Index
 - d. Percent of ideal body weight
22. The most common form of child abuse seen in pediatric primary care is:
 - a. Burns
 - b. Fractures
 - c. Soft tissue injuries
 - d. Shaken baby syndrome
23. A differential diagnosis for child abuse would include all of the following except:
 - a. Birth marks
 - b. Unintentional injury
 - c. Inadequate parenting
 - d. Prader-Willi syndrome
24. Which of the following symptoms are not typical of a child with ADHD?
 - a. Easily distracted
 - b. Difficulty playing quietly
 - c. Doesn't follow directions
 - d. Frequently angry and resentful
25. Which of the following clinical findings would not suggest an eating disorder with a purging component?
 - a. Sore throat
 - b. Brittle nails
 - c. Constipation
 - d. Finger calluses
26. Which of the following situations does not necessarily warrant immediate mental health assessment and/or referral?
 - a. 13-year-old girl who has been "down" for the last month with varied somatic complaints
 - b. 9-year-old boy whose parents recently separated and filed for a divorce and seems to be doing well
 - c. 16-year-old girl who has a history of long-standing depression but seems to be doing well in school
 - d. 15-year-old boy who expresses suicidal thoughts
27. Which adolescent would be at greatest risk for developing anorexia nervosa?
 - a. 12-year-old female who just had her first period
 - b. 14-year-old gymnast
 - c. 16-year-old male runner
 - d. 18-year-old female college student
28. Which of the following substances is associated with pupillary constriction?
 - a. Amphetamines
 - b. LSD
 - c. Heroin
 - d. Nicotine
29. A risk factor that is common to many psychosocial pediatric problems including failure to thrive, conduct or oppositional disorders, and childhood depression is:
 - a. Maternal depression or other psychiatric disorder
 - b. Substance abuse
 - c. Prematurity
 - d. History of sexual abuse

30. Which of the following diagnoses is not more common among males?
- ADHD
 - Conduct disorders
 - Suicide
 - FTT
31. The diagnostic criteria for autistic disorders includes which of the following?
- Speech delay, ataxia, mental retardation
 - Impairments in social interactions, interpersonal communication, and staring spells
 - Mental retardation, impairments in social interactions, and stereotypical restricted pattern of interests and activities
 - Impairments in social interactions, in interpersonal communication, and stereotypical restricted pattern of interests and activities
32. In addition to specific academic skill deficits, learning disabilities are commonly associated with which of the following characteristics?
- Perceptual-motor impairments, normal motor function
 - Perceptual-motor impairments, impulsiveness
 - Perceptual-motor impairments, Down syndrome
 - Lack of impulsiveness, perceptual-motor impairment

ANSWERS

- | | |
|--------------|--------------|
| 1. b | 17. b |
| 2. a | 18. a |
| 3. b | 19. c |
| 4. c | 20. b |
| 5. d | 21. c |
| 6. a | 22. c |
| 7. a | 23. d |
| 8. c | 24. d |
| 9. b | 25. b |
| 10. c | 26. b |
| 11. b | 27. b |
| 12. a | 28. c |
| 13. c | 29. a |
| 14. a | 30. d |
| 15. a | 31. d |
| 16. b | 32. b |

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Health Promotion and Well Child Care for Infants, Children, and Adolescents

JoAnne Silbert-Flagg

■ OVERVIEW: HEALTH MAINTENANCE AND HEALTH PROMOTION

Pediatric nurse practitioners (PNPs) have long been on the forefront of promoting and maintaining optimal physical and mental health for children and their families. Their understanding of the multiple factors that influence the overall health and development of children and adolescents enable PNPs to implement evidence-based care and individualize interventions, appropriately involving family members to enhance health outcomes.

One strategy for enhancing health and developmental outcomes in children, adolescents, and their families is the implementation of routine child health supervision. Health supervision is comprised of those measures that promote health, prevent morbidity and mortality, and facilitate optimal development and maturation within the context of the family and community. It involves routine well child visits, which include health promotion strategies, anticipatory guidance, as well as specific screening procedures at regular, timed intervals throughout childhood and adolescence.

■ CHILD HEALTH SUPERVISION

- Components of the Health Visit which are Age-Appropriate, Health and Developmentally Focused
 1. The parent/child interview
 2. Developmental and educational surveillance (including school performance)
 3. Observation of parent/child interaction
 4. Physical examination specific for each visit
 5. Screening and immunizations
- Assessment of strengths and vulnerabilities (concerns, problems, and stressors affecting the child and family)
- Evidence-based individualized interventions, including health promotion strategies and anticipatory guidance
- General Interviewing Approaches
 1. Determine who will be present for interview
 2. Provide privacy and empathetic environment
 3. Maintain eye contact and relaxed facial expressions
 4. State you will be taking notes during interview to enhance accuracy of recorded data
 5. Obtain history with child clothed
 6. Ask open ended questions that begin with “why,” “how,” or “what”
 7. Use direct questions to obtain specific information
 8. Avoid leading questions
 9. Use language parents and child understand
 10. Provide undivided attention; listen carefully, both through verbal and nonverbal actions
 11. Build self-esteem and confidence throughout interview
 12. Conduct interview with cultural sensitivity
- Communication with Parents (in addition to general approach)
 1. Obtain parent’s perception of any concerns or problems; if both parents present, obtain each parent’s view of concerns or problems
 2. Restate parental concerns to ensure accuracy and understanding

3. Be supportive, not judgmental, e.g., “Why didn’t you bring your child in earlier?”
- Communication with Young Children (less than six years of age)
 1. Talk to child at his/her eye level
 2. Use play to enhance comfort
 3. Use projective techniques to elicit information about how child is feeling, e.g., “Tell me how your bear is feeling today.”
 4. Use nonthreatening words, e.g., tube instead of needle, opening instead of cut, since young children engage in magical thinking
 5. Allow adequate time for responses
 6. Remember that young children have difficulty giving detailed information
 - Communication with Younger School-Age Children
 1. Communicate with parent first if child is initially shy
 2. Ask questions and give explanations using concrete terminology
 3. Use simple diagrams when asking child to describe location of symptoms
 4. Allow time for responses
 5. Give permission to express fears and concerns
 - Communication with Older School-Age Children and Adolescents
 1. If parent is present, conduct part of interview (questions dealing with personal or sensitive information) when alone with the older child or adolescent
 2. Interview while child is fully clothed
 3. Start interview with nonthreatening questions
 4. Inform older child/adolescent that all questions you are asking have to do with his/her health; that you ask all older children/adolescents these questions
 5. Acknowledge that although all of your questions are necessary, some may feel uncomfortable to answer
 6. Inform older child/adolescent that information shared is confidential unless he/she tells you about wanting to hurt him/herself or someone else has hurt him/her
 7. Encourage expression of feelings and concerns
 8. Enhance self-esteem and provide positive feedback during interview
 - Confidentiality Issues and Informed Consent
 1. Healthcare providers are required by law to keep the information gathered in the course of a child’s care confidential; in 2003, national standards to protect the privacy of personal health information were established as part of

- the Health Insurance Portability and Accountability Act (HIPAA) of 1996 (DHHS, 2003)
2. Privileged information may only be shared among healthcare professionals involved in the care of a child when parental consents are signed or in medical emergencies when release of information is in the best interest of the child and absolutely necessary for the provision of care
 3. Many states mandate reporting by healthcare professionals of special circumstances (reasonable cause to suspect child physical or sexual abuse or neglect, suicidal intent, gunshot and stab wounds)
 4. Some states require reporting of births, deaths, certain diseases, and other vital statistics
 5. Consents should be signed by child’s parent or legal guardian before information concerning child is released; emancipated minor (under the age of 18 years and married, parent of his or her own child, or self-sufficiently living away from home with parental consent) also may sign consents
 6. Minor’s informed consent laws vary across states by status (emancipated) and conditions
 - a. Individual states have statutes allowing access to contraceptives; pregnancy testing/prenatal care; as well as the diagnosis, treatment, and prevention of sexually transmitted diseases per consent of minor
 - b. Individual states have statutes allowing HIV testing and treatment per consent of minor

■ THE PEDIATRIC HISTORY

During the examination of a pediatric patient, the history is critical in the early detection of problems and prevention of long-term negative outcomes. Approximately 80% of the information used to arrive at a diagnosis is derived from the history.

- Complete Patient History
 1. Biographic information—demographic data; name and reliability of person providing the history as well as his/her relationship to child.
 2. Chief complaint (CC)—reason for visit
 3. History of Present Illness (HPI)—if there are symptoms
 - PQRST—parameters of symptoms
 - P = promoting, preventing, precipitating, palliating factors
 - Q = quality or quantity
 - R = region or radiation
 - S = severity, setting, simultaneous symptoms

T = temporal factors, onset, duration, intervals, frequency, course over time, has symptom occurred before?

OLDCARTS

O = onset

L = location

D = duration

C = characteristics

A = associated symptoms

R = relieving/aggravating factors

T = timing, treatment

S = severity, sequence, summary

4. Past Medical History (PMH)
 - General state of health—(in child's/parent's words)
 - Perinatal history—obtained based on child's age and appropriateness to care
 - a. prenatal history
 - b. planned or unplanned pregnancy
 - c. onset of prenatal care and compliance with recommended care
 - d. medications, drugs, alcohol use during pregnancy
 - e. medical problems during pregnancy
 - Perinatal history—obtained based on child's age and appropriateness to care
 - a. length of labor and delivery
 - b. type of delivery
 - c. medications or anesthesia
 - d. complications
 - e. birth weight
 - f. gestation at birth, Apgar scores
 - g. if multiple births, birth order
 - Postnatal—obtained based on child's age and appropriateness to care
 - a. maternal and infant problems
 - b. age and weight at discharge
 - c. early feeding history including breast-feeding history
5. Common childhood illnesses—list dates and type
6. Serious illness—list dates and course of illness
7. Mental health care received—list date and type
8. Hospitalizations—list dates and type
9. Injuries—list dates and course of treatment, recovery
10. Current Health Status (CHS)—based on child's age
 - a. Nutrition—24 hour recall, meal pattern, who eats with whom, how meal time conducted, fluid type and volume consumed, cultural expectations, special diet, caffeine, dietary supplements, herbal supplements, artificial sweeteners, carbonated beverages
- b. Elimination—toilet training, urinary characteristics, enuresis or encopresis, day and night variations, bowel pattern
- c. Sleep—hours, location, naps, snoring, enuresis, night bottle usage
- d. Development—including school performance, daily activities, recreation and hobbies, social adjustment, behavior, and temperament
- e. Discipline/behavioral concerns—of parent, teacher, childcare provider, relationship with siblings/friends, approaches to discipline
- f. Safety—specific for age (see preventive health care and anticipatory guidance)
- g. Immunizations and screening—specific for age (see preventive health care)
- h. Allergies—specify nature of allergic reaction, medications, food, airborne, transfusion
- i. Current medications—prescription, over the counter, alternative

11. Environmental History

Assessment of environmental hazards

- a. Arsenic—source is drinking water; used to preserve wood; added to poultry feed
- b. Mercury—source is contaminated fish (swordfish, tuna, and shark), industries that burn fossil fuels
- c. Lead—source is indoor paint, water, soil, or foreign bodies
- d. Polychlorinated biphenyls (PCBs), Dioxins, and Furans—source is exposure through dietary fat including fish, meat, and dairy products
- e. Asbestos—source is through housing construction
- f. Water pollutants—source is drinking water or crop irrigation
- g. Indoor air pollutants—source is carbon monoxide, environmental tobacco smoke (ETS), radon, molds, solvents, and pesticides
- h. Outdoor air pollutants—source is ozone, particulate matter, sulfur dioxide, nitrogen oxides, diesel exhaust, and polycyclic aromatic hydrocarbons
- i. Sun exposure—source is ultraviolet radiation from sun
- j. Solvents—source is gasoline, degreasers, art and craft supplies, nail products, paint, glues, varnishes, newly installed carpeting, dry cleaning products, indoor and outdoor air, and drinking water contaminants

- k. Pesticides—source is household products used in gardens, lawns, fruits and vegetables, some lice removal shampoos, and drinking/bathing water
Source for data <http://www.psr.org/resources/pediatric-toolkit.html>
- Assessment of risk prevention—appropriate for age
- smoke detector/radon detector, carbon monoxide detector
 - hot water heater setting
 - use of car seat/seat belt
 - pet safety
 - sports safety equipment
 - gun safety
 - childproofing of household appropriate to age
 - use of sunscreen with SPF 15 or higher
Source for data <http://www.psr.org/resources/pediatric-toolkit.html>
12. Growth and Development—based on child's age and appropriateness to care
- Physical growth—pattern of height, weight, head circumference, and body mass index
 - Developmental milestones—language, fine motor, gross motor, social, achievement of milestones, early intervention provided
 - Mental health/social-emotional development—temperament, relationships, mood state, coping abilities
 - School—performance, attendance, individual educational plan (IEP)
13. Family History (FH)
- Family profile and medical history (genogram)—include serious, chronic, inherited, and congenital problems in three generational family (blood relatives); drug and alcohol abuse; mental health problems
 - Family social history—include household composition/type of dwelling/family support systems
 - members of household and their relationship to the child, cultural influences, religious affiliation
 - physical environment of household
 - employment of parents and work schedule
 - socioeconomic factors of parents or legal guardian
 - healthcare coverage
 - childcare arrangements/after-school care and activities
- family stressors—current, recent, or chronic
 - family travel to high-risk areas
14. Review of Systems—(ROS) not addressed in HPI and appropriate for age; Ask “does this child now, or has this child ever, had problems with any of the following” systems?
- General—recent weight change, fever, fatigue, weakness
 - Head—injury
 - Eye—last eye examination, visual problems, use of glasses/contacts
 - Ears—last hearing screen, otitis media
 - Nose/sinus—frequent upper respiratory infections, nasal discharge, nosebleeds, sinus pain
 - Throat—frequent tonsillitis/pharyngitis
 - Dentition—last dental examination, problems with teeth, bleeding gums
 - Neck—stiffness, adenopathy, goiter
 - Respiratory—cough, pneumonia, bronchiolitis, wheezing, tuberculosis, chest x-ray, shortness of breath
 - Cardiovascular—murmur, rheumatic fever, palpitations, chest pain, hypertension
 - Gastrointestinal—abdominal pain, vomiting, gastroesophageal reflux, diarrhea, constipation, flatus, hepatitis
 - Genitourinary/Reproductive—
Female—menarche, dysmenorrhea, premenstrual syndrome, last menstrual period, sexual activity (age of onset, number of partners, dyspareunia, contraception, condom use), pregnancy, vaginal discharge, history of gynecological examinations, history of screening/treatment for sexually transmitted infections, breast self examinations, and breast abnormalities.
Male—hernia, testicular pain/self examination, penile discharge, sexual activity (age of onset, number of partners, condom use)
 - Musculoskeletal—muscle or joint pain, decreased range of motion
 - Neurological—fainting, seizures, weakness, paralysis, numbness, tremors, dizziness, headache
 - Hematological—anemia, bruising easily
 - Psychiatric—anxiety, depression, mood swing, suicide ideation, anorexia, bulimia, violence, abuse
 - Endocrine—heat or cold intolerance, endocrine disease (thyroid, diabetes, adrenal), polyuria, polydipsia, polyphagia

- Adolescent History Based on Adolescent's Age and Appropriateness to Care
HEADS—Home, Education/Employment, Activities and Peers, Drugs
 1. Home—household relationships, family dynamics and relationships, living arrangements
 2. Education/Employment—school attendance, grades, attitude about school relationships, best and worst subjects, homework, goals, type of employment and hours worked
 3. Activities and Peers—spare time, physical activity, screen time, friends
 4. Disabilities/drugs—tobacco, alcohol, substance use (by self and friends), ability to carry out activities of daily living, sleep, safety, self image, sexuality, suicide, self mutilation, history of harm to animals, history of harm to others, Internet use (time, sites, chat rooms, personal profiles, e-mail), communication with strangers, exposure to violence, conflict management, firearm exposure and use, gang membership (self and friends)
- Tools for Screening for Alcohol or Substance Use in Adolescents
 1. RAFFT Tool for Adolescents (alcohol screening)—A single “yes” answer indicates further investigation is needed. Two or more positive responses strongly suggest the probability that an alcohol dependence problem exists. Four “yes” responses indicates alcohol dependence.
Do you use alcohol to Relax?
Do you use alcohol Alone? Do you use alcohol with Family?
Do you use alcohol with Friends?
Have you experienced any Trouble (problems) as a result of your alcohol use?
 2. CRAFFT Tool for Adolescents (alcohol and/or substance use screening)
C—Have you ever ridden in a Car driven by someone (including yourself) who was “high” or had been using alcohol or drugs?
R—Do you ever use alcohol or drugs to Relax, feel better about yourself, or fit in?
A—Do you ever use alcohol or drugs while you are by yourself, Alone?
F—Do you ever Forget things you did while using alcohol or drugs?
F—Do your Family or Friends ever tell you that you should cut down on your drinking or drug use?
T—Have you ever gotten into Trouble while you were using alcohol or drugs?
- Sexual/Reproductive and Substance Use History Guidelines
 1. Inform child that these questions are asked of all older school-age children and adolescents
 2. Reinforce that although these questions are very personal or sensitive, they are necessary to gain a complete picture of that child or adolescent's health
 3. Reassure child or adolescent that the information he or she shares is confidential unless information about harm to self or others is revealed
 4. Progress from least to most sensitive questions
 5. It is best to phrase questions—“When was the first time you had intercourse?” instead of “Have you ever had intercourse?”
 6. Make sure older child or adolescent understands meaning of terms used
 7. Essential elements of sexual/reproductive history
 - a. Date of menarche (first menses)
 - b. Frequency, length, and quantity of menses with associated symptoms; e.g., cramping, headache, or backache
 - c. Date of last menses
 - d. Use of tampons or pads
 - e. Age of first intercourse, date of last intercourse
 - f. Sexual preference, e.g., males, females, or both; same sex exploration is common in teenagers; number of sexual partners
 - g. Types of sexual practices, e.g., male, female; oral sex, intercourse
 - h. Reasons for sexual activity, e.g., increases self-esteem; enjoyment; peer pressure
 - i. Pregnancies and outcomes
 - j. Current contraception
 - k. History of any sexually transmitted infections (STI); naming each disease, e.g., gonorrhea, chlamydia, etc.
 - l. Vaginal or penile discharge
 - m. Date of last pelvic examination
 - n. Contraceptive history, current contraception, use of condoms
 - o. Knowledge of STIs, AIDS, pregnancy, and prevention measures
 - p. Date of prior test or desire for HIV testing
 - q. Performance of self-breast or testicular exam
 - r. History of sexual abuse
- Interval History
 1. Chief complaint (CC)
 2. Interim health—since last visit
 3. Current health—nutrition, elimination, sleep, development, allergies, immunizations

4. Update any changes in history since last visit
 5. Review of systems since last history
- Telephone History (telephone triage)
 1. Requires triage decision
 - a. Telephone management
 - b. Office visit
 - c. Refer to emergency department or other healthcare provider
 2. Telephone protocol books are helpful in assessment and management of common illnesses and problems encountered by phone
 3. Critical elements
 - a. Identify yourself
 - b. Identify caller, his or her relationship to child, and caller's telephone number
 - c. Obtain child's name, age, and approximate weight
 - d. Ascertain thorough history of present illness or problem
 - e. If not medically necessary to see child, explain rationale and evaluate comfort of caller in home management
 - f. Tell caller when to call back, which includes advising on signs of worsening status
 - g. Ask caller to telephone again to give progress report if there are any concerns about the child; if caller does not telephone as requested, IT IS CRITICAL TO MAKE THE FOLLOW-UP CALL
 - h. Ask caller to write down information that has been given or at least to repeat the information
 - i. Offer simple, understandable explanations
 - j. Advise caller to contact you again with any questions
 - k. Convey warmth, empathy, and support
 - l. Document all telephone conversations, including history, diagnosis, and management plan

■ THE PEDIATRIC PHYSICAL EXAMINATION

- General Information
 1. Examination should be comprehensive and systematic
 2. Observation is first critical component of the examination beginning as soon as the child is seen
 3. Use examination to teach child about his or her body
- Age-Related Issues (Infants)
 1. Developmental considerations
 - a. Stage of trust versus mistrust
 - b. Stranger anxiety develops at 6 to 7 months
 - c. Separation anxiety develops at 8 to 9 months
 - d. Major fears—separation from parents, and pain
 2. Approaches to physical examination
 - a. Approach slowly
 - b. Conduct as much of examination with infant on parent's lap
 - c. Provide infant with security objects, e.g., special blanket or toy
 - d. Use distraction and engaging facial expressions during the examination
 - e. Conduct examination using noninvasive to invasive sequence, e.g., auscultate heart and lung sounds first, examine ears and throat last
 - f. Allow for brief break if infant is hungry or stressed
- Age-Related Issues (Toddlers)
 1. Developmental considerations
 - a. Stage of autonomy versus shame/doubt
 - b. Striving for independence
 - c. Negativism and temper tantrums (common)
 - d. Beginning of magical thinking
 - e. Major fears—separation from parents, intrusion of body orifices, loss of control, pain
 2. Approaches to physical examination
 - a. Use of distraction is helpful
 - b. Allow child to touch and hold equipment before examination
 - c. Demonstrate examination on doll, toy, or parent before conducting examination on child
 - d. Give child choices when possible
 - e. When necessary, tell child what you are going to do instead of gaining permission, e.g., "I am going to check your tummy" versus "Is it OK with you if I check your tummy?"
 - f. Conduct as much of examination as possible on parent's lap
 - g. Conduct examination using noninvasive to invasive sequence
- Age-Related Issues (Preschool Children)
 1. Developmental considerations
 - a. Stage of initiative versus guilt
 - b. Magical thinking
 - c. Egocentrism
 - d. Major fears—separation from parents, loss of control, body mutilation, pain
 2. Approaches to physical examination
 - a. Inform child what you are going to do and what he/she can do to help

- b. Role play with equipment, e.g., let child examine ears of doll first
 - c. Head to toe examination sequence can usually be implemented
 - d. Choose words carefully due to magical thinking
 - e. Allow choices whenever possible
 - f. Teach child about his/her body during course of examination
 - g. Praise child for helping and attempting to cooperate
- Age-Related Issues (School-Age Children)
 1. Developmental considerations
 - a. Industry versus inferiority
 - b. Concrete thinking
 - c. Desires to act brave
 - d. Enjoys gathering scientific information
 - e. Modesty emerges with older school-age child
 - f. Major fears—separation from peers, loss of control, pain, death; beginning at age 9 years
 2. Approaches to physical examination
 - a. Head to toe sequence
 - b. Scientific terminology with concrete explanations
 - c. Answer questions factually with age-appropriate vocabulary
 - d. Explain use of equipment, e.g., otoscope
 - Age-Related Issues (Adolescents)
 1. Developmental considerations
 - a. Stage of identity versus role diffusion
 - b. Striving for independence and control
 - c. Formal operational thinking
 - d. Bodily concerns
 - e. Concerns about being different
 - f. Major fears—change in body image, separation from peers, loss of control, death
 2. Approaches to physical examination
 - a. Assure privacy
 - b. Examine without parent unless adolescent prefers parent remain in room
 - c. Inform adolescent of each step of examination
 - d. Give choices whenever possible
 - e. Cover parts of body not currently being examined
 - f. Assure privacy
 - g. Teach adolescent about his/her body during course of examination
 - h. Provide reassurance of “normalcy” during course of examination
 - i. Recognize and discuss apprehension about breast, pelvic, and testicular examinations

- Measurement of Vital Signs
 1. Temperature
 - a. Rectal temperature is an accurate method, but proper technique must be used to avoid injury. A temporal artery thermometer, which is more expensive, is increasingly becoming available.
 - b. Tympanic membrane and axillary temperature is quick and noninvasive measurement, reliability may be a problem
 - c. Temperature of 38°C and above is considered a fever
 - d. A normal newborn’s temperature ranges from 36.5 to 37.5°C rectally

2. Pulse Norms

a.

	Age	Beats/ minute	Mean
(1)	birth to 7 days	95–160	(125)
(2)	1 to 3 weeks	105–180	(145)
(3)	1 to 6 months	110–180	(145)
(4)	6 to 12 months	110–170	(135)
(5)	1 to 3 years	90–150	(120)
(6)	4 to 5 years	65–135	(110)
(7)	6 to 8 years	60–130	(100)
(8)	9 to 16 years	60–110	(85)
(9)	over 16 years	60–100	(80)

- b. Conditions that commonly elevate pulse
- (1) Temperature—for every 1 degree of temperature elevation in Fahrenheit, pulse increases by 10 beats per minute
 - (2) Anxiety/stress, excitement
 - (3) Exercise
 - (4) Severe anemia
 - (5) Hyperthyroidism
 - (6) Hypoxia
 - (7) Heart disease

3. Respirations Norms

a.

	Age	Breaths per minute
(1)	Neonate	40–60
(2)	up to 1 year	24–38
(3)	1–3 years	22–30
(4)	4–6 years	20–24
(5)	7–9 years	18–24
(6)	10–14 years	16–22
(7)	14–18 years	14–20

- b. Conditions that commonly elevate respirations
- (1) Temperature—for every 1 degree of temperature elevation in Fahrenheit,

- respirations increase by 4 breaths per minute
- (2) Anxiety/stress, excitement
 - (3) Pain
 - (4) Respiratory conditions, e.g., pneumonia
 - (5) Heart disease
4. Blood Pressure
- a. Appropriate cuff size required for accurate reading
 - (1) Bladder width should be approximately 40% of the circumference of the arm measured at a point midway between the olecranon and acromion
 - (2) Bladder length should cover 80% to 100% of the circumference of the arm
 - (3) Blood pressure should be measured with cubital fossa at the heart level; the arm should be supported
 - (4) The stethoscope bell is placed over the brachial artery pulse, proximal and medial to the cubital fossa and below the bottom edge of the cuff
 - b. Use Korotkoff sound IV (muffling sound) as diastolic blood pressure in children under 13 years of age; use Korotkoff sound V (disappearance of sound) as the diastolic blood pressure in children 13 years of age and older
 - c. Plot blood pressure on standard blood pressure graphs for boys or girls
 - d. Begin to measure blood pressure at well-child visits, starting at 3 years of age
 - e. A single elevated blood pressure measurement in an apparently healthy child does not necessarily reflect disease
 - f. Hypertension—average systolic and/or diastolic blood pressure 95th percentile for age and sex on at least 3 separate occasions using the same arm, same cuff, and same position

■ Blood Pressure Readings from 50th Percentile Ages Birth to 12 Months

Age	Systolic (mm Hg)	Diastolic (mm Hg)
Birth to 6 months	76–106	68–66
6 to 12 months	68–65	65–67

Four limb blood pressure measurements can be used to assess for coarctation of the aorta. Mean difference between upper and lower extremities should be 10 mm Hg or less.

Data from *The Harriet Lane Handbook* (2009).

■ Blood Pressure Readings from 5th to 95th Percentile Ages 1 to 17 Years

Age	Systolic (mm Hg)	Diastolic (mm Hg)
1 to 3 years	98–109	54–67
4 to 6 years	106–117	66–76
7 to 10 years	110–119	74–78
11 to 14 years	117–128	78–84
15 to 17 years	126–136	81–89

Data from National Institutes of Health, National Heart, Lung and Blood Institute. *Fourth Report on the Diagnosis, Evaluation, and Treatment of High Blood Pressure in Children and Adolescents* (2005)
http://www.nhlbi.nih.gov/guidelines/hypertension/child_tbl.htm

- g. Taller, heavier children have higher blood pressure than smaller children of same age
- h. Pulse pressure—difference between systolic and diastolic blood pressures (normal is 20 to 50 mm Hg)
 - (1) Wide pulse pressure from high systolic pressure is usually due to fever, exercise, or excitement
 - (2) Wide pulse pressure from low diastolic pressure is usually due to patent ductus arteriosus, aortic regurgitation, or other serious heart disease

■ SPECIFIC NORMAL FINDINGS AND COMMON VARIATIONS

- Head and Neck
 1. History indicating possible abnormalities
 - a. Difficult birth; use of forceps, vacuum
 - b. Unusual head shape or preferred position at rest
 - c. Poor head control for age
 2. Selected physical examination findings
 - a. Head circumference is approximately 2 cm larger than chest during first year of life; head and chest circumferences should be equal at 1 year of age; during childhood, chest is usually 5 to 7 cm larger than head
 - b. Fontanels—best to assess while infant is sitting up and not crying
 - (1) Posterior fontanel rarely palpable at birth; closes by 2 months of age
 - (2) Size of anterior fontanel should be no larger than 4 to 5 cm in diameter
 - (3) Anterior fontanel closes by 18 months of age
 - (a) Early closure usually leads to synostosis
 - (b) Late closure is commonly seen with increased intracranial

- pressure, hypothyroidism, rickets, syphilis, Down syndrome, osteogenesis imperfecta
- (4) Large anterior fontanel may indicate:
 - (a) Chronically increased intracranial pressure
 - (b) Subdural hematoma
 - (c) Rickets
 - (d) Hypothyroidism
 - (e) Osteogenesis imperfecta
 - (5) Bulging anterior fontanel is usually seen with conditions that cause increased intracranial pressure, e.g., meningitis/encephalitis, fluid overload
 - (6) Sunken anterior fontanel is usually seen with severe dehydration (more than 10%)
- c. Unusual head size or shape
- (1) Hydrocephalus—excessively large head at birth or head that grows abnormally rapid; usually associated with distended scalp veins, widely separated cranial sutures, large and tense anterior fontanel, and “sunset eyes”
 - (2) Microcephaly—head circumference > 2 standard deviations below the mean for age, sex and gestation; reflects an abnormally small brain; common causes are intrauterine infections (e.g., herpes, rubella, syphilis); genetic defects; drug usage during pregnancy (especially alcohol)
 - (3) Macrocephaly—head circumference > 2 standard deviations above the mean for age, sex and gestation; common causes are hydrocephalus, masses, increased intracranial pressure; skeletal dysplasias (osteogenesis imperfecta)
 - (4) Head tilt—common causes include strabismus, CNS lesions, or short sternocleidomastoid muscle
 - (5) Caput succedaneum—diffuse edema of the soft tissue of the scalp which usually crosses suture lines; may be seen with bruising due to traumatic vaginal birth; seen at birth; no specific treatment necessary; usually resolves in 2 to 3 days
 - (6) Cephalohematoma—subperiosteal collection of blood which does not cross suture lines; often does not appear until several hours after birth and may increase over 24 hours; no specific treatment indicated; resolves over a few weeks to months; observe for hyperbilirubinemia
- (7) Premature or irregular closure of suture lines can cause unusual head shape (craniosynostosis)
 - (8) Bossing (bulging) of frontal area is associated with rickets and prematurity
- d. Head Control
- (1) By 4 months of age, head should be held erect and in midline
 - (2) By 6 months of age, there should be no head lag when infant is pulled from supine to sitting position; if present, may indicate neuromuscular disorder; may be the first sign of cerebral palsy
- e. Neck
- (1) Pain and resistance to flexion may indicate meningeal irritation
 - (2) Torticollis (restriction of motion)—can result from birth trauma (e.g., injury to the sternocleidomastoid muscle with bleeding into the muscle), muscle spasm, viral infection, or drug ingestion
 - (3) Webbed neck—common in Turner’s syndrome, a chromosomal abnormality occurring 99% of the time in females which results in webbed neck, widespread nipples, abnormal ears, micrognathia, and lymphedema of hands and feet
 - (4) Unusual position of trachea could indicate serious lung problem
 - (5) Mass in the neck
 - (a) Thyroglossal duct cyst—usually seen near midline of neck; cyst moves up and down with protrusion of tongue; may become infected and present as an abscess; surgical excision recommended
 - (b) Brachial cleft cyst—can appear as swelling anterior to sternocleidomastoid (SCM) muscle or as opening along anterior border of SCM; may drain and become infected
 - (c) Hematoma (of sternocleidomastoid muscle)—more common in breech deliveries
 - (d) Enlarged thyroid—due to hyperthyroidism or hypothyroidism; visible thyroid gland is almost always enlarged
 - (e) Enlarged lymph node—most frequent cause of lateral neck mass

- Face
 1. History indicating possible abnormalities
 - a. Difficult delivery; use of forceps, vacuum
 - b. Asymmetry of face when crying or speaking
 - c. Facial features which are unusual or do not match family characteristics
 - d. Drug or alcohol use during pregnancy
 2. Selected physical examination findings
 - a. Asymmetry of nasolabial folds or drooping mouth indicates facial nerve impairment or Bell's palsy
 - b. Child who demonstrates open mouth breathing and facial contortions may have allergic rhinitis
 - c. Dysmorphic facial features are hallmark of numerous syndromes (e.g., fetal alcohol syndrome) and diagnosis should be pursued

- Eyes
 1. History indicating possible abnormalities
 - a. Premature infant who required resuscitation, needed ventilator or oxygen support, had retinopathy of prematurity
 - b. Infant who does not track faces or objects; absent blink in response to bright lights or sudden movements
 - c. Children less than 6 years of age who:
 - (1) Rub eyes excessively, squint, have photophobia
 - (2) Have difficulty reaching for or picking up small objects
 - (3) Engage in head tilting
 - (4) Hold objects close to face
 - d. School-age children (same as young children) who:
 - (1) Sit close to blackboard or TV in order to see
 - (2) Are making poor progress in school not explained by intellectual deficit or learning disability
 - e. Any age child who:
 - (1) Demonstrates white area in pupil visible in photographs (retinoblastoma)
 - (2) Complains of headaches not present upon awakening, but progress during the day (accommodative errors)
 - (3) Has problems with excessive tearing—allergies (accommodative errors)
 - (4) Has an eye which turns in or out (strabismus)
 2. Selected physical examination findings
 - a. Position and placement—inner canthal distance averages 2.5 cm; epicanthal folds present in Asian children; palpebral fissures lie horizontally
 - (1) Hypertelorism (wide set eyes)—present in Down syndrome
 - (2) Epicanthal folds can be frequently seen in Down syndrome, renal agenesis, or glycogen storage disease
 - (3) Ptosis could be normal or may indicate paralysis of oculomotor nerve
 - (4) Exophthalmos (protruding eyeballs)—can be seen with hyperthyroidism
 - b. Eyelids normally same color as surrounding skin
 - (1) “Stork bite” mark—telangiectatic nevi disappear by 12 months
 - (2) Blocked tear duct (dacryostenosis)—may lead to infection of lacrimal sac evidenced by swelling, redness, and purulent discharge (dacryocystitis)
 - (3) Periorbital edema—soft swelling that may be associated with renal or cardiac problems or sinusitis; acute onset of unilateral eyelid edema with erythema, induration, and tenderness indicates periorbital cellulitis
 - c. “Allergic shiners”—bluish discoloration and soft edema below eyes usually indicates allergies
 - d. Sclera and conjunctiva
 - (1) Sclera is shiny, clear, and white
 - (2) Bulbar conjunctiva (covers sclera) is moist and transparent and palpebral conjunctiva (lines the eyelids) is pink and moist
 - (3) Spots of brown melanin may be seen in dark skinned races
 - (4) Yellow sclera indicates jaundice
 - (5) Redness may indicate bacterial or viral infection, allergy or irritation, e.g., chemicals
 - (6) Excessive pallor of the palpebral conjunctiva indicates anemia
 - (7) Cobblestone appearance of palpebral conjunctiva (lining the eyelids) can indicate severe allergy or contact lens irritation
 - e. Pupils and iris
 - (1) Unequal pupils (anisocoria)—usually congenital and normal, but can indicate increased intracranial pressure from head trauma or other intracranial disease processes, e.g., meningitis
 - (2) Dilated, fixed pupils—usually indicate severe brain damage
 - (3) Dilated pupils—may result from use of anticholinergic drugs (e.g., atropine) and substance abuse (e.g., amphetamines)

- (4) Abnormally small pupils—may result from brain damage, use of morphine, or substance abuse (e.g., cocaine)
 - f. By 3 to 4 months, infants should have binocular vision (ability to fixate on one visual field with both eyes simultaneously)
 - (1) Assessment techniques to elicit phoria (movement of eye when covered) or tropia (obvious turning in or out of eye without coverage) (e.g., strabismus)
 - (a) Cover-uncover test (when eye is covered, it may deviate in (esophoria) or out (exophoria) and return to midline when uncovered)
 - (b) Corneal light reflex (Hirschberg's test)—with light held 12 to 14 inches from eyes, reflection of light should be the same on both corneas; if unequal, it is suggestive of phoria or tropia
 - (2) Intermittent alternating convergent strabismus—normal from 0 to 6 months of age
3. Ophthalmoscopic examination—red reflex should be elicited before discharge from the nursery and at all subsequent routine health supervision visits
- a. The result of the red reflex examination is to be rated as normal when the reflection of the two eyes viewed, both individually and simultaneously, are equivalent in color, intensity, and clarity and there are no opacities or white spots (leukokoria) within the area of either or both red reflexes.
 - b. A child with an abnormal red reflex should be immediately referred to an ophthalmologist for a more complete examination.
 - c. All infants and children with a positive family history of retinoblastoma; congenital, infantile, or juvenile cataracts; glaucoma; or retinal abnormalities should be referred to an ophthalmologist for a more complete eye examination, regardless of the status of the red reflex.
- Ears
 - 1. History indicating possible abnormalities
 - a. Prenatal exposure to maternal infection, irradiation, or drug abuse
 - b. Birth weight less than 1500 g
 - c. Anoxia in neonatal period
 - d. Ototoxic antibiotic usage (e.g., gentamycin)
 - e. Cleft palate
 - f. Infections
 - g. Meningitis
 - h. Encephalitis
 - i. Recurrent or chronic otitis media
 - 2. Behaviors suggestive of hearing loss
 - a. No reaction to loud or strange noises
 - b. No babbling in infant after 6 months
 - c. No communicative speech; reliance on gestures after 15 months of age
 - d. Language delays
 - 3. Selected physical examination findings
 - a. Position and placement—low or obliquely set ears may indicate genitourinary or chromosomal abnormality or a multisystem syndrome
 - b. Pain
 - (1) Pain produced by manipulation of auricle or pressure on tragus may indicate otitis externa
 - (2) Pain and tenderness over mastoid process may indicate mastoiditis
 - c. Examination of tympanic membrane (TM)
 - (1) For best visualization of TM—pull auricle down and back in children under 3 years of age; pull auricle up and back for children over 3 years of age
 - (2) Crying produces erythema of TM bilaterally; landmarks are still visible with succinct light reflexes and +4 TM mobility
 - (3) Pneumatic otoscopy is critical for assessment of fluid in middle ear
 - (a) Decreased TM mobility—indicates fluid in middle ear
 - (b) In child with pressure equalization (PE) tubes, decreased mobility of TM indicates obstruction or dysfunction of tubes
- Nose and Sinuses
 - 1. History indicating possible abnormalities
 - a. Inability to move air through both nares
 - b. Discharge
 - c. Nasal flaring or narrowing on inspection
 - d. Hypernasal voice—snoring, hypertrophied adenoids
 - 2. Selected physical examination findings
 - a. Flattened nasal bridge (in other than Asian or African-American children) may indicate congenital anomalies
 - b. Boggy nasal mucous membranes (bluish, pale, edematous) with serous drainage indicates allergic rhinitis
 - c. Persistent copious or purulent discharge is indicative of sinusitis

- d. Unilateral purulent discharge suggests foreign body
- Throat/Mouth
 1. History indicating possible abnormalities
 - a. Lack of, or excessive fluoride supplementation or fluoridated water
 - b. Infant or toddler who goes to sleep with bottle of milk or juice
 - c. Thumb sucking or pacifier use beyond 2 years of age
 - d. Unusual sequence of tooth eruption
 2. Selected physical examination findings
 - a. Lips
 - (1) Cherry red color indicates acidosis
 - (2) Drooping of one side of lips indicates facial nerve impairment
 - (3) Fissures at corners of mouth may indicate riboflavin or niacin deficiency
 - b. Teeth
 - (1) Mottling may indicate excessive fluoride intake
 - (2) Green or black staining can result from oral iron intake
 - c. Palate—decay of maxillary incisors may result from baby bottle caries syndrome
 - (1) Palpation of palate is important in newborns to detect submucosal cleft
 - (2) Uvula rises and remains in midline when saying “ah”; deviation or absence of movement indicates involvement of glossopharyngeal or vagus nerves
 - (3) Bifid uvula is suggestive of a submucosal cleft palate
 - d. Tonsils
 - (1) During childhood, tonsillar hypertrophy is a normal immunological response; largest in size between 8 and 9 years of age and decreases in size after puberty
 - (2) Asymmetrically enlarged tonsil without infection may suggest tonsillar lymphoma
 - e. Voice
 - (1) Nasal quality indicates enlarged adenoids
 - (2) Hoarse cry may indicate croup, cretinism, or tetany
 - (3) Chronic hoarseness may indicate vocal cord polyps
 - (4) Shrill, high-pitched cry may indicate increased intracranial pressure
 - f. Temporomandibular joint (TMJ)
 - (1) Findings indicative of TMJ dysfunction—pain upon palpation of TMJ, decrease in mandibular movement, TMJ sounds (popping and clicking), malocclusion, and abnormal morphology of mandible (micrognathia)
- (2) Inability to open jaw (trismus) associated with fever and sore throat is suggestive of peritonsillar abscess
- Heart
 1. History indicating possible abnormalities
 - a. Infant
 - (1) Increased respirations, especially during sleep
 - (2) Prolonged feeding time, tires during feedings
 - (3) Cyanosis of mucous membranes of mouth
 - (4) Eyelid edema
 - b. Child
 - (1) Increased respirations, especially during sleep
 - (2) Squatting or sleeping in knee–chest position
 - (3) Eyelid edema
 - (4) Cyanosis of mucous membranes of mouth
 - (5) Exercise intolerance
 2. Selected physical examination findings
 - a. Heart sounds and area of clearest auscultation
 - (1) S_1 (closure of mitral and tricuspid valves)—heard best at apex
 - (2) S_2 (closure of aortic and pulmonic valves)—heard best at aortic and pulmonic areas
 - (3) Physiological splitting of S_2 during inspiration is normal; if fixed (heard upon inspiration and expiration), may indicate atrial septal defect or pulmonic stenosis
 - (4) S_3 —heard best at apex (sounds like Kentucky); due to blood rushing through mitral valve and hitting an empty ventricle; normal in almost all children; if loud in character, may indicate high diastolic pressure in involved ventricle as found in acute ventricular failure
 - (5) S_4 —heard best at apex (sounds like Tennessee); almost never normal; indicates high pressure in either ventricle as found in pulmonic and aortic stenosis and systemic hypertension
 - b. Normal variations in heart rhythm—in sinus arrhythmia, heart rate increases with inspiration and decreases with expiration; disappears with exercise or holding breath

- c. Innocent (functional) murmurs—present in approximately 50% of children
- (1) Characteristics
 - (a) Usually systolic in timing
 - (b) Usually soft; never more than Grade III
 - (c) Rarely transmitted
 - (d) Low pitched, vibratory, musical, or twangy
 - (e) Short duration
 - (f) Usually loudest at left lower sternal border or at the second or third intercostal space
 - (g) Varies in loudness and presence from time to time
 - (h) Heard loudest in the recumbent position, during expiration and after exercise
 - (i) Diminishes with change in positioning from recumbent to sitting
 - (j) No cyanosis
 - (k) Normal pulses, respiratory rate, and BP
 - (l) Normal growth and development
 - (m) Absence of a thrill (vibratory sensation felt over murmur with palm of hand)
 - (2) Types of innocent heart murmurs in children
 - (a) Pulmonary ejection murmur (heard at the pulmonic area)—early to mid systole; distinct gap between first heart sound and murmur and end of murmur and second heart sound
 - (b) Vibratory or Still's murmur—musical or vibratory murmur; heard best at the lower left sternal border
 - (c) Venous hum—heard best above or below clavicles, second or third interspaces; more coarse quality; very dependent upon position; disappears when child lies down or turns neck, which decreases blood velocity through internal jugular veins
 - (3) Conditions that increase intensity of innocent heart murmurs—exercise, fever, and anemia due to increased cardiac output
 - (4) Innocent murmurs in the newborn
 - (a) Transition from fetal to adult circulation may take up to 48 hours
 - (b) Usually Grade I or II
 - (c) Systolic
 - (d) Not associated with other signs and symptoms
 - (5) Point of Maximal Impulse (PMI)
 - (a) Children less than 8 years of age—4th intercostal space, midclavicular line
 - (b) Children more than 8 years of age—5th intercostal space, slightly right of midclavicular line
 - (c) Displacement of PMI with cardiac enlargement
 - (d) Increased pulsation of PMI indicates conditions, which increase cardiac output, e.g., anemia, anxiety, fever, fluid overload
 - (6) Peripheral pulses—normally palpable, equal in intensity and rhythm; weak or absent femoral pulses may indicate coarctation of the aorta
- Lungs
 1. History indicating possible abnormalities
 - a. Family history of tuberculosis, cystic fibrosis, allergy, asthma, atopic dermatitis
 - b. Infants and young children
 - (1) Premature infant with any respiratory complications
 - (2) Sudden onset of coughing or difficulty breathing
 - (3) Difficulty feeding
 - (4) Apnea episodes
 - c. Older children and adolescents
 - (1) Smoking
 - (2) Cocaine use
 - (3) Recurrent or chronic cough
 - (4) Exercise intolerance
 2. Selected physical examination findings
 - a. Normal breath sounds—breath sounds are best heard by having child breathe through mouth
 - (1) Vesicular—low pitch, soft intensity; inspiration is more than expiration with a ratio of 5:2; heard over peripheral lung fields
 - (2) Bronchovesicular—medium pitch, moderate intensity; inspiration equals expiration with a ratio of 1:1; heard over main bronchus
 - (3) Bronchial/tracheal—high pitch, loud intensity; inspiration less than expiration with a ratio of 1:2
 - b. Abnormal breath sounds
 - (1) Rhonchi—coarse sounds heard on expiration that are indicative of secretions in the large airways; usually present in bronchitis; clear with coughing; associated with bronchial

- fremitus (coarse vibrations felt with hand on chest as air passes through exudate in bronchi)
 - (2) Transmitted rhonchi—coarse sounds that result from the transmission of sound from congested nasal passages to the chest; can be avoided by having child breathe through mouth
 - (3) Wheezing—high pitched musical or whistling sounds produced as air passes through narrowed airways; heard in bronchiolitis, asthma; cystic fibrosis; foreign body aspiration (unilateral wheezing)
 - (4) Crackles—fine crackling sounds heard upon inspiration indicative of air passing through moisture in alveoli; usually suggests pneumonia or congestive heart failure
 - (5) Pleural friction rub—creaking or grating sound caused by inflamed parietal and visceral pleural linings rubbing together; usually inspiratory and expiratory; subsides when child holds breath
- c. Chest movement
 - (1) Children under 7 years of age are diaphragmatic (abdominal) breathers
 - (2) Girls over 7 years of age become thoracic breathers; boys continue as abdominal breathers
 - (3) Chest structural abnormalities may compromise lung function
 - (a) Pectus carinatum—protuberant sternum
 - (b) Pectus excavatum—depressed sternum
- Breasts (see Chapter 14, Endocrine Disorders)
 - 1. History indicating possible abnormalities
 - a. Prepubertal breast enlargement in girls
 - b. Gynecomastia in boys at any age
 - c. Breast mass
 - d. Galactorrhea not associated with childbearing
 - 2. Selected physical examination findings
 - a. Neonate may have gynecomastia and milky discharge which disappears within 2 weeks (or at least 3 months)
 - b. Gynecomastia can be normal variant in males due to temporary estrogen/testosterone imbalance (usually begins at Tanner Stage II to III and can last for 1 to 2 years); most commonly felt as small, tender, oval subareolar mass measuring up to 2 to 3 cm in diameter
 - c. Gynecomastia also may be indicative of
 - (1) Obesity or increased muscle (pseudogynecomastia)
 - (2) Testicular tumor (testes must be palpated in any male with gynecomastia)
 - (3) Medication usage—estrogen, steroids, tricyclic antidepressants (e.g., imipramine), resperidol, mellaril, amphetamines, digoxin, cimetidine
 - (4) Klinefelter's syndrome (47XXY)—associated with small penis and testes, scoliosis, aspermia, decreased testosterone levels, and height greater than 6 feet
 - d. Asymmetric breast development is normal in the adolescent female
 - e. Galactorrhea may be indicative of:
 - (1) Pregnancy
 - (2) Recent abortion
 - (3) Pituitary tumor—associated with increased prolactin level, increased headaches, amenorrhea, peripheral vision loss
 - (4) Drug use—marijuana; opiates (codeine, heroin, morphine); amphetamines; hormones (oral contraceptives); digoxin; valium; cimetidine; phenothiazines (thorazine, mellaril); haloperidol; tricyclic antidepressants; resperidol
 - (5) Hypothyroidism
 - f. Breast masses in adolescents
 - (1) Benign breast masses (obtain ultrasound versus mammogram due to dense breast tissue in adolescents)
 - (a) Fibroadenoma—most common breast mass in adolescents; increased incidence in African-Americans
 - (i) Characteristics—single, unilateral mass; round or discoid in shape; firm and smooth in consistency; no retraction; mobile; nontender
 - (ii) No variation with menstrual cycle
 - (b) Fibrocystic breasts—usually result of hormonal imbalance
 - (i) Characteristics—breast pain with or without lumps; symptoms worsen a few days before menses and resolve with completion of menses
 - (ii) Mobile cysts or areas are more dense and fibrous; usually resolve in 1 to 3 months

- (2) Neoplastic breast masses (very rare)—firm, nonmobile, painless, overlying skin changes, nipple discharge

- Abdomen

1. History indicating possible abnormalities
 - a. Birth weight under 1500 g places infant at high risk for necrotizing enterocolitis
 - b. Failure to pass first meconium stool within 24 hours
 - c. Jaundice
 - d. Failure to grow or unexplained weight loss
 - e. Projectile vomiting or blood in emesis
 - f. Chronic diarrhea or constipation
 - g. Enlargement of the abdomen with or without pain
 - h. Abdominal or pelvic pain
2. Selected physical examination findings—flexion of knees and hips facilitates examination
 - a. Prominent abdomen (potbelly)—normal in early childhood in sitting and supine positions due to poorly developed musculature; children up to 13 years of age may have prominent abdomen in standing position
 - b. Liver edge—may be palpable 1 to 2 cm below right costal margin, especially with deep inspiration
 - c. Spleen tip—may be palpable 1 to 2 cm below left costal margin, especially with deep inspiration
 - d. Diastasis recti—separation of rectus abdominis muscles several cm wide from xiphoid bone to umbilicus; may extend to symphysis pubis; normal as long as not associated with hernia
 - e. Bowel sounds
 - (1) Active bowel sounds—heard every 5 to 15 seconds; range of 4 to 12 sounds per minute
 - (2) Hypoactive bowel sounds—heard at more than 15 second intervals or less than 4 sounds per minute
 - (3) Hyperactive bowel sounds—heard less than every 5 seconds or more than 12 sounds per minute
 - f. Palpation of fecal mass in LLQ common with constipation

- Reproductive System

1. History indicating possible abnormalities
 - a. Discharge or bleeding from vagina or penis
 - b. History or suspicion of sexual abuse
 - c. Sexual intercourse without use of contraceptives

- d. Scrotal swelling with crying or bowel movement
- e. “Empty” scrotum versus retractable testes
- f. Unusual voiding pattern

2. Selected physical examination findings (refer to Human Growth and Development Infancy through Adolescence (Chapter 2) for Tanner stages and sequence of pubertal development)

- a. Signs and symptoms of possible sexual abuse
 - (1) Evidence of general physical abuse or neglect
 - (2) Evidence of trauma or scarring in genital, anal, or perianal areas
 - (3) Changes in skin color or pigmentation in genital or anal area
 - (4) Any sexually transmitted disease
 - (5) Anorectal itching, bleeding, pain, or poor sphincter tone
 - (6) Rashes, sores, or discharge in genital area
 - (7) Dysuria, frequency of urination, enuresis
 - (8) Behavioral or emotional changes
 - (9) Deterioration in school performance
 - (10) Inappropriate sexual behavior for developmental level
- b. Labial adhesions
 - (1) Rule out ambiguous genitalia
 - (2) Adhesions of labia minora most common in young infants
- c. Male genitalia
 - (1) Phimosis—foreskin that cannot be easily retracted over the glans penis before 5 years of age
 - (2) Undescended testes (cryptorchidism)—if testicle can be “milked” down into scrotum, consider it descended; refer undescended testicle after 1 year of age
 - (3) Hypospadias or epispadias should be referred
 - (4) Large scrotum—hydrocele, spermatocele, varicocele
 - (a) Testicular tumor (common in adolescents and young men)
 - (b) Indirect inguinal hernia

- Musculoskeletal System (refer to Chapter 10, Musculoskeletal Disorders: Infancy through Adolescence)

1. History indicating possible abnormalities
 - a. Birth history—large for gestational age, abnormal presentation, anoxia in perinatal period
 - b. Delay in motor developmental milestones

- c. Unusual style of movement—dragging of legs while crawling
 - d. Hand dominance in an infant—may indicate weakness of other hand
 - e. Leg pain/limp
 - 2. Selected physical examination findings
 - a. Barlow and Ortolani tests to detect hip dislocation or subluxation in infants—more common in females and breech deliveries
 - (1) Barlow’s test—hip is flexed and thigh is adducted which causes displacement of femoral head from acetabulum with a palpable “clunk”
 - (2) Ortolani’s test—hip is flexed at 90 degree angle and abducted with examiner’s finger over the greater trochanter which reduces dislocated hip; a “clunk” is heard and felt
 - b. Allis sign to detect hip dislocation or shortened femur—unequal leg length is abnormal
 - c. Trendelenburg test to detect hip dislocation—child stands and raises one leg off the ground; lowering of iliac crest on side opposite weight-bearing leg indicates defect in weight-bearing hip
 - d. Gower’s sign shows generalized muscle weakness; often indicative of muscular dystrophy—use of hands on legs to push self to standing position is abnormal
 - e. Scoliosis (lateral curvature of the spine)
 - (1) Functional—child can voluntarily straighten spine; disappears when child is recumbent
 - (2) Structural—persistent curvature, unequal height of shoulders and iliac crests when standing erect, presence of rib hump when leaning forward
 - f. Developmental differences
 - (1) Longitudinal arch of foot can be obscured by fat pad until 3 years of age; child appears “flat footed”
 - (2) Bowlegs (genu varum) and wide-based gait—common in toddlers
 - (3) Knock knees (genu valgum)—common in preschool children
 - g. “Turned-in” foot may have different causes
 - (1) Femoral anteversion—most common between 3 and 8 years; 99% resolve by 8 years of age
 - (2) Tibial torsion—most common between 1 and 3 years of age; growth alone will correct 99% of cases
 - (3) Metatarsus adductus
 - h. Hip should always be examined in child complaining of knee pain
- Neurologic System
 - 1. History indicating possible abnormalities
 - a. Delay or regression in developmental milestones, unusual behavior for age, learning or school difficulties
 - b. Headaches/seizures
 - c. Clumsiness or progressive weakness, irritability or lethargy
 - 2. Selected physical examination findings
 - a. Infant reflexes (automatisms)—see **Table 3-1**
 - b. Absence of infantile automatisms or persistence beyond expected time of disappearance may indicate severe CNS dysfunction
 - c. Spasticity—may be early sign of cerebral palsy
 - d. Use of Denver II or other general developmental screening tool yields much data regarding age appropriate skills
 - e. Babinski sign—normal up to 2 years of age
 - f. Age of disappearance of individual “soft signs” is typically 8 years
 - Dermatologic System (refer to Chapter 7, Dermatologic Conditions: Infancy through Adolescence)
 - 1. History indicating possible abnormalities
 - a. Family history of atopic dermatitis, allergic skin disorders, familial hair loss, or unusual pigmentation patterns

■ **Table 3-1** Infant Reflexes

Reflex	Appearance	Disappearance
Palmar grasp	birth	2–3 months
Plantar grasp	birth	10–12 months
Moro	birth	4–6 months
Stepping	birth	3–4 months
Tonic neck	Birth to 6 weeks	4–6 months
Rooting	birth	3–4 months, except during sleep up to 12 months

- b. Chronic or repeated acute episodes of skin lesions
- c. Frequent scratching or rubbing of body area
- 2. Selected physical examination findings
 - a. Birthmarks (nevi)
 - (1) Salmon patch (stork bite)—common on eyelids, nasolabial region, or nape of neck; disappears by 12 months
 - (2) Nevus flammeus (port-wine stain)—enlarges as child grows
 - (3) Strawberry nevus (raised hemangioma)
 - (a) Begins as circumscribed grayish white area; later becomes red and raised; not always present at birth
 - (b) Majority resolve spontaneously by 10 years of age
 - (4) Mongolian spot (hyperpigmented nevi)
 - (a) Usually in sacral or gluteal areas
 - (b) Generally seen in newborns of African-American, Asian, or Latin descent
 - (5) Café-au-lait spots (light brown, flat patches)—6 or more may be associated with neurofibromatosis
 - b. Common color changes in newborns
 - (1) Acrocyanosis—cyanosis of hands and feet due to vascular instability
 - (2) Cutis marmorata—transient mottling when infant is exposed to decreased temperature
 - (3) Erythema toxicum—transient, benign, pink papular rash with vesicles on thorax, back, buttocks, and abdomen
 - (4) Harlequin color change—as infant lies on side, dependent portion of body becomes pink and upper portion is pale
 - (5) Jaundice (refer to Chapter 13, Hematological/Oncological/Immunologic Disorders: Infancy through Adolescence)—skin blanches yellow with pressure instead of white as with carotinemia
 - (a) Jaundice appears first on head and then progresses down body
 - (b) Approximate level of hyperbilirubinemia by cephalocaudal distribution—nose (3 mg/dL); face (5 mg/dL); chest (7 mg/dL); abdomen (10 mg/dL); legs (12 mg/dL); palms (20 mg/dL)
 - (c) Physiological jaundice—appears 2 to 3 days after birth
 - (d) Appearance of jaundice in first 24 hours indicates hemolytic disease or infection
 - (e) Breastfeeding infants who develop jaundice in the second to third week may have breastmilk jaundice
 - (f) First appearance of jaundice at 2 to 3 weeks of age—must suspect biliary atresia (indirect bilirubin is elevated and AST and ALT also may be elevated)
 - (g) Factors contributing to development of hyperbilirubinemia—metabolic acidosis, lowered albumin levels, free fatty acids, drugs (maternal or fetal), other condition (e.g., fetal distress, hypoxia, hypothermia, hypoglycemia, infant bruising, maternal or fetal infection)
 - c. Degree of dehydration can be estimated by length of time skin retains “tenting” after it is pinched
 - (1) < 2 seconds = < 5% loss of body weight
 - (2) 2 to 3 seconds = 5% to 8% loss of body weight
 - (3) 3 to 4 seconds = 9% to 10% loss of body weight
 - (4) > 4 seconds = > 10% loss of body weight
- Lymph Nodes
 - 1. History indicating possible abnormalities
 - a. Recurrent infections—tonsillitis, adenoiditis, bacterial infections, oral candidiasis, chronic diarrhea
 - b. Poor growth, failure to thrive
 - c. Maternal HIV infection
 - d. IV drug use
 - e. Multiple and indiscriminate sexual contacts
 - 2. Selected physical examination findings
 - a. Normal size is up to 1 cm in inguinal area; 2 cm in cervical area; in other areas, up to 3 cm is normal
 - b. Nodes enlarged due to infection are firm or fluctuant, warm, tender, mobile, and may be accompanied by redness of overlying skin
 - c. “Shotty” nodes (e.g., under 0.5 cm in diameter, firm, mobile and nontender) can be present at any time in childhood and usually indicate past infection
 - d. Suspect malignancy or tuberculosis if supraclavicular nodes are palpated

■ SELECTED LABORATORY TESTS AND VALUES

- General Considerations
 1. Cost, pain, and invasiveness versus need for data to make accurate diagnosis
 2. Anesthetic cream used topically can ease venipuncture, especially in highly anxious children
 3. Laboratory values should be referenced against specified norms of laboratory conducting the testing since normal values may vary from laboratory to laboratory
- Hematology—CBC with differential
 1. Normal range of values for complete blood count (CBC) with differential (see **Table 3-2**)
 2. Common causes of variation in CBC with differential
 - a. Hemoglobin variations
 - (1) Increased—may indicate polycythemia (an overproduction of RBCs as a result of hypoxia); dehydration, or intravascular hemolysis
 - (2) Decreased—may indicate anemia, hemodilution, sickle cell anemia, thalassemia, hemorrhage, or hyperthyroidism
 - b. Hematocrit variations
 - (1) Increased—may indicate polycythemia, dehydration, or erythrocytosis
 - (2) Decreased—may indicate anemia, hemorrhage, hyperthyroidism, leukemia, or cirrhosis
 - c. Red blood cell variations
 - (1) Increased—may indicate dehydration, hemorrhage, severe diarrhea, acute poisoning
 - (2) Decreased—may indicate blood loss, low iron intake, lead poisoning, leukemia, rheumatic fever, systemic lupus erythematosus, or subacute bacterial endocarditis
 - d. White blood cell variations
 - (1) Increased—may signal bacterial infection (e.g., tonsillitis, sepsis, meningitis, appendicitis) or indicate acute hemorrhage, serum sickness, steroid use, hemolysis, or leukemia
 - (2) Decreased—indicates bone marrow depression which may result from viral infection; rickettsial infection; hypersplenism; leukemia; certain drugs (e.g., antiseizure)

■ **Table 3-2** Normal Range of Values CBC

	Newborn	1 month	1 year	2–6 years	6–12 years	12–18 years
Hgb (g/dL)	14–24	11–17	11–15	11–15	11.5–15	12–16 (Males) 11.5–16 (Females)
Hct (%)	42–60	33–55	33–41	34–42	34–43	37–50 (Males) 36–44 (Females)
RBC (mill/mm ³)	4.1–7.5	4.2–5.2	4.1–5.1	3.9–5.3	4.0–5.2	4.5–5.3 (Males) 4.1–5.1 (Females)
WBC × 1000/mm ³	9–30	5–19.5	6–17	5–15.5	4.5–13.5	4.5–13.5
Neutrophils (Segs %)	33–60	32–60	31–60	33–60	33–60	33–60
Bands (%) (immature neutrophils)	0–5	0–5	0–5	0–5	0–5	0–5
Lymphocytes (%)	25–33	31–57	31–61	31–59	31–57	20–45
Monocytes (%)	3–7	3–7	3–7	3–7	3–7	3–10
Eosinophils (%)	1–4	1–4	1–4	1–4	1–4	1–7
Basophils (%)	0–0.75	0–0.75	0–0.75	0–0.75	0–0.75	0–0.75
Platelets (10 ³ /mm ³)	84–478	150–400	150–400	150–400	150–400	150–400
Reticulocyte count (%)	3–7	0.2–2	0.2–0.28	0.5–4	0.5–4	0.5–1.5
MCV (Mean Cell Volume ⁺ ; μm ³)	95–120	70–95	70–95	70–95	70–95	78–100
MCHC (Mean Cell Hemoglobin Concentration*)	31–37	31–37	31–37	31–37	31–37	31–37

+ Mean cell volume—average size of red cells 31–37

* Mean cell hemoglobin concentration—ratio of hemoglobin weight to hematocrit; helps to distinguish between normal colored cells (normochromic cells) and paler (hypochromic) cells

- medications, antibiotics, antihistamines, diuretics, analgesics, tricyclic antidepressants)
- e. Neutrophils variations
 - (1) Increased—may indicate bacterial infection, ischemic necrosis from burn injuries, metabolic disorders (e.g., diabetic ketoacidosis), stress response, emotional distress, inflammatory diseases (e.g., rheumatic arthritis), or hemolysis
 - (2) Decreased (neutropenia)—viral infections (e.g., hepatitis, mononucleosis), chemotherapy or radiation, immune deficiencies, malignancies
 - f. Band cell or stab (immature neutrophil) variations—increased (known as a shift to the left); usually indicates severe bacterial infection (e.g., sepsis, pneumonia)
 - g. Lymphocytes variations
 - (1) Increased—may indicate a viral infection, lymphocytic leukemia, ulcerative colitis, immune diseases
 - (2) Decreased—indicates severe debilitating illnesses (e.g., congestive heart or renal failure, advanced TB), immunosuppressive therapy, Hodgkin's disease, after burns or trauma, Cushing syndrome, corticosteroid usage, HIV infection
 - (3) Atypical lymphocytes—common in premature and healthy newborn infants; may indicate viral infection (e.g., mononucleosis, hepatitis), lymphocytic leukemia, ulcerative colitis, immune diseases
 - h. Monocyte variations
 - (1) Increased—may indicate recovery from acute infection; SBE, leukemia, Hodgkin's disease, rickettsial infection, SLE, rheumatoid arthritis, hepatitis
 - (2) Decreased—may indicate rheumatoid arthritis, HIV infection, prednisone usage
 - i. Eosinophil variations
 - (1) Increased—may indicate allergic disorders (e.g., asthma, allergic rhinitis, atopic dermatitis) or parasite infection
 - (2) Decreased—stress responses due to trauma, shock, burns, and emotional stress; Cushing syndrome
 - j. Basophil variations
 - (1) Increased—may indicate certain leukemias, Hodgkin's disease, inflammatory conditions (e.g., ulcerative colitis), polycythemia, chronic hemolytic anemia; infections such as TB, varicella, influenza
 - (2) Decreased—may indicate hyperthyroidism, pregnancy, stress, prolonged use of steroids, allergic reaction
 - k. Platelet variations
 - (1) Increased—may indicate acute infection, malignancy, post-splenectomy, trauma, rheumatoid arthritis, Kawasaki disease
 - (2) Decreased (thrombocytopenia)—may indicate leukemia, idiopathic thrombocytopenic purpura (ITP), autoimmune disorders, drugs (e.g., penicillin, ampicillin, cephalothin), hemolytic uremic syndrome, DIC, viral infection, HIV infection
 - l. Reticulocyte count variations
 - (1) Increased—may indicate hemorrhage/blood loss, increased destruction of RBCs, response to initiation of iron therapy
 - (2) Decreased—may indicate iron deficiency anemia, chronic infection, radiation, aplastic anemia
 - m. MCV variations
 - (1) Increased—may indicate macrocytic anemia due to folic acid or vitamin B₁₂ deficiency
 - (2) Decreased—may indicate microcytic anemias caused by iron deficiency and thalassemia, anemia of chronic disease, or lead poisoning
 - n. MCHC variations
 - (1) Increased—may indicate spherocytosis
 - (2) Decreased—suggests hypochromic anemia caused by iron deficiency, chronic blood loss, or thalassemia
- Chemistry
 1. Sodium chloride (mEq/L)
 - a. Newborn—134 to 144
 - b. Child—135 to 145
 - c. Increased—dehydration, vomiting or diarrhea, diabetes insipidus, Cushing syndrome
 - d. Decreased—vomiting, diarrhea, burns, diabetic ketoacidosis, Addison's disease, acute or chronic renal failure, syndrome of inappropriate antidiuretic hormone (SIADH)
 2. Potassium (mEq/L)
 - a. Newborn—3.7 to 5.9
 - b. Child—3.5 to 5.0
 - c. Increased—may indicate acidosis or renal failure

- d. Decreased—may indicate diarrhea, vomiting, dehydration, malabsorption, use of diuretics or anti-inflammatory drugs
 3. Blood urea nitrogen (BUN)
 - a. Normal range—5 to 20 mg/dL
 - b. Increased—may indicate a high protein diet, renal or urinary obstruction or disease, GI hemorrhage, malignancies, dehydration, shock
 - c. Decreased—hemodilution, pregnancy, nephrotic syndrome, liver failure
 4. Creatinine (more sensitive indicator of renal function than BUN)
 - a. Normal value—0.3 to 1 mg/dL
 - b. Increased—may indicate renal dysfunction, urinary tract obstruction, dehydration, muscle disease
 5. Bilirubin (mg/dL)
 - a. Birth—1.5
 - b. Three to four days postnatal
 - (1) Breastfed—7.3
 - (2) Bottlefed—5.7
 - c. Older infant and child
 - (1) Total—less than 1.5
 - (2) Direct (conjugated)—0.2 to 0.4 (higher levels require investigation for pathology)
 - (3) Indirect (unconjugated) bilirubin—0.4 to 0.8 (levels greater than 20 mg/dL may be neurotoxic to brain)
 6. Cholesterol (mg/100 mL)
 - a. Full-term newborn—45 to 167
 - b. Infant—70 to 190
 - c. Child and adolescent—less than 170
 7. Lead—normal value (less than 10 μ g/dL)
- Urine
 1. pH
 - a. Newborn—5.0 to 7.0
 - b. Older infant and child—4.8 to 7.8
 - c. Increased (alkaline)—may indicate urinary tract infection, salicylate intoxication
 - d. Decreased (acidic)—may indicate acidosis, renal failure, diarrhea, or dehydration
 2. Specific gravity
 - a. Newborn—1.001 to 1.020
 - b. Older infant and child—1.001 to 1.030
 - c. Increased—may indicate dehydration, nephrosis, glomerulonephritis
 - d. Decreased—may indicate diabetes insipidus, severe renal damage
 3. Glucose (should be negative)—presence of sugar may indicate diabetes mellitus or other metabolic disorders; liver disease or renal tubular disorders
 4. Protein (should be negative)—presence of protein may indicate renal disease (e.g., nephritis, nephrosis), exercise, SLE, orthostatic proteinuria, asymptomatic proteinuria
 5. Ketones (should be negative)—presence of ketones may indicate fever, dehydration, anorexia, diarrhea, fasting, prolonged vomiting, or anorexia
 6. Nitrites (should be negative)—presence of nitrites strongly suggests urinary tract infection
 7. WBC
 - a. Normal range—0 to 4 WBC/HPF
 - b. Increased—may indicate urinary tract infection, fever, pyelonephritis, TB, nephrosis
 8. RBC
 - a. Normal range 1 to 2 RBCs/HPF
 - b. Increased—may indicate urinary tract infection, pyelonephritis, SLE, renal stones, trauma, TB, hemophilia, polyarteritis nodosa, malignant hypertension
 9. Bacteria (should be negative)—100,000 colonies/mL or more of a single pathogen on urine culture by clean catch method confirms a urinary tract infection; repeat urine culture should be obtained for a result of 10,000 to 100,000 colonies/mL
 - Cerebrospinal fluid (CSF)
 1. Pressure—70 to 180 mm H₂O; higher indicates increased intracranial pressure which may be the result of a tumor, cerebral hemorrhage, meningitis, obstructed shunt
 2. Appearance—Clear
 - a. Bloody—may indicate traumatic tap, cerebral hemorrhage
 - b. Yellow—may indicate hyperbilirubinemia or metastatic melanoma
 - c. Cloudy—suggests increased WBC as found in bacterial meningitis
 3. Glucose—60 to 80 mg/dL
 - a. Increased—diabetes
 - b. Decreased—may indicate bacterial meningitis, TB meningitis, hypoglycemia, leukemia with metastasis
 4. Protein—15 to 45 mg/dL; increased in encephalitis, bacterial meningitis, TB meningitis, acoustic neuroma
 5. Cell count
 - a. Infant—0 to 20 WBC/mm³
 - b. Child and adolescent—0 to 10 WBC/mm³
 - c. Increased—bacterial meningitis, early viral meningitis, cerebral abscess

■ SELECTED SCREENING TESTS

- Blood pressure screening on all children beginning at age 3. If child has renal or cardiovascular abnormality, perform at all visits regardless of age.
- Growth Parameters
 1. Height, weight, and head circumference from birth to age 3 plotted on appropriate growth chart at each well child visit. If head circumference has been appropriate for the first 2 years, routine measurements need not continue being obtained after age 2. Body mass index beginning at age 24 months.
 - a. infants should be undressed completely and young children should be wearing underpants only
 - b. recumbent length is plotted on the chart from birth to 3 years
 - c. when the child is old enough to be measured upright, height should be plotted on the chart for ages 2 through 18.
 2. Height, weight, and body mass index from age 3 through adolescence plotted on appropriate growth chart at each well child visit. A BMI \geq 95th percentile identifies a child who is overweight and a BMI \leq 5th percentile identifies a child who is underweight.
<http://www.cdc.gov/growthcharts/>
- Newborn Screening
 1. Screening should be conducted according to state law
 2. All states require testing for:
 - a. Hypothyroidism
 - b. Phenylketonuria (PKU)
 - c. Galactosemia
 - d. Hemoglobinopathies (e.g., sickle cell disease)
 3. Other tests may include screening for:
 - a. Maple syrup urine disease
 - b. Homocystinuria
 - c. Biotinidase deficiency
 - d. Tyrosinemia
 - e. Congenital adrenal hyperplasia
 - f. Cystic fibrosis
 - g. Toxoplasmosis
 4. Initial specimens should be obtained at least 24 hours after birth, but not more than 7 days of age
 5. Most states recommend a second metabolic screening in the first week of life for those infants discharged prior to 24 hours
 6. Infants with a positive screen result should receive close follow up with additional confirmatory studies performed
- Vision Screening
 1. Identify risk factors
 - a. Prenatal infections
 - b. Congenital cyanotic heart disease
 - c. Structural malformation
 - d. Family history of eye or vision problems
 - e. Excessive oxygenation in neonatal period
 - f. Hearing problems
 - g. Parent concern about the child's visual functioning
 - h. Deterioration in school performance
 2. Conduct vision screen at ages 3, 4, 5, 6, 8, 10, 12, 15, and 18.
 - a. Young infant
 - (1) Assess pupillary response to light
 - (2) Illicit blink reflex
 - (3) Determine ability to fix on and follow an object
 - (4) Assess red reflex
 - b. Older infant and toddler
 - (1) Determine ability to fix on and follow an object
 - (2) Perform corneal light reflex test (Hirschberg)
 - (3) Perform cover/uncover test
 - (4) Assess red reflex
 - c. Preschool child (same as toddler, plus)
 - (1) Conduct visual acuity tests (Allen figures, HOTV, Sjogren hand, illiterate E)
 - (2) Use Ishihara for color perception
 - (3) Assess red reflex
 - d. School-age child and adolescent (pre-school child, plus)
 - (1) Far vision—Snellen chart
 - (2) Near vision—Rosenbaum or Jaeger card
 - (3) Assess red reflex
 - e. Refer children for further evaluation with:
 - (1) Abnormal or asymmetric red reflex
 - (2) Asymmetric corneal light reflex
 - (3) Abnormal cover/uncover test
 - (4) Structural abnormality
 - (5) Failure to follow object equally when covering each eye
 - (6) Visual acuity minimal acceptable is 20/40 for age 3 to 5 and 20/30 for age 6 and older
 - (7) Two line difference or more in scores between eyes
- Hearing Screening
 1. Identify infants and children at risk for hearing problems
 - a. Neonatal risk criteria
 - (1) Affected family member
 - (2) Bilirubin $>$ 20 mg/dL
 - (3) Congenital CMV, herpes, rubella

- (4) Defects in ENT structure
 - (5) Birth weight < 1500 g
 - (6) Bacterial meningitis
 - (7) Use of ototoxic medications for more than 5 days
 - (8) Mechanical ventilation for cardio-pulmonary disease for more than 48 hours
 - (9) Intracranial hemorrhage
 - b. Risk criteria for children less than 2 years of age (neonatal risk factors, plus)
 - (1) Parental concerns regarding hearing or language development
 - (2) Head trauma with temporal bone fracture
 - (3) Infections known to cause sensorineural hearing loss (e.g., measles, mumps)
 - (4) Recurrent otitis media or middle ear effusion
 2. Conduct hearing screen at the following intervals and when indicated
 - a. Newborn performed prior to discharge using auditory brainstem response (ABR) or evoked otoacoustic emissions (EOAE)
 - b. Ages 4, 5, 6, 8, and 10
 - (1) Examine ears using an otoscope with pneumatic otoscopy before conducting audiometry
 - (2) Conduct pure tone audiometry—test each ear at 500, 1000, 2000, and 4000 Hz (hand-held audiometers have not proven effective in screening children)
 - (3) Use tympanometry to further assess middle ear air pressure and tympanic membrane compliance if pneumatic otoscopy was abnormal
 3. Refer children to an audiologist for further evaluation if hearing threshold levels are greater than 20 dB at any of the above frequencies; if the reliability of a test with an individual child is uncertain, repeat screening before referral
- Tuberculosis screening
 1. Mantoux test should be used for testing; use 0.1 cc of purified protein derivative (PPD) which contains 5 tuberculin units; administer via intradermal injection on the volar aspect of the forearm to produce a 6 to 10 mm wheal; multiple-puncture tests do not have adequate specificity and sensitivity
 2. Test should be read 48 to 72 hours following injection by a measurement of the area of induration, not redness
 3. BCG vaccination is NOT a contraindication to TB skin testing
 - 4. Positive skin tests include children who have reaction of:
 - a. At least 15 mm *INDURATION* with no risk factors
 - b. At least 10 mm *INDURATION* who are less than 4 years of age or those with medical risk factors (e.g., born, or whose parents were born, in areas of prevalent TB [Asia, Middle East, Africa, or Latin America] or exposed to adults who are HIV positive, homeless, illicit drug users, nursing home residents, incarcerated or institutionalized persons, or migrant farm workers)
 - c. At least 5 mm *INDURATION* who are household contacts of active or previously active TB cases as well as children with immunosuppressive conditions, including HIV infection
 - 5. Current recommendations for TB testing are based on degree of risk rather than routine, universal screening
 - 6. Immediate TB testing should be conducted on children who are contacts of individuals with confirmed or suspected infectious TBI, have been in contact with persons who have been incarcerated in the last five years, are immigrating from or with a history of travel to endemic countries, and those with clinical or radiologic findings of TB.
 - 7. Annual TB testing should be conducted for children at high risk, beginning as early as 3 months of age
 - a. HIV positive or living with an HIV infected person
 - b. Institutionalized and/or incarcerated children and adolescents
 - 8. Testing every two to three years. Children exposed to the homeless, HIV-infected individuals, nursing home residents, institutionalized or incarcerated adolescents, illicit drug users, and migrant farm workers
 - 9. Periodic TB testing between 4 to 6 years and again between 11 and 16 years for children living in high prevalence areas or with uncertain history of risk factors
 - 10. See most recent AAP *Red Book* for specific updated guidelines
<http://aapredbook.aappublications.org/>
- Lead Screening
 1. The Centers for Disease Control (CDC) no longer recommend universal screening; targeted screening beginning at age one to two years of age should be based on surveillance of risk (refer to Chapter 13, Hematological/Oncological/Immunologic Disorders). All

children receiving Medicaid are required by federal regulation to be tested for blood lead levels at one and two years of age.

2. Risk factors include children who live in a house, or are cared for in a house, built before 1950 or built before 1978, that has recently or is undergoing renovation; have a sibling or playmate being followed for lead poisoning; live with a person whose job involves exposure to lead; live near an industrial site; use pottery that are suspect of having lead content; and/or have exposure to burning lead-painted wood.
 3. Most children with lead poisoning are between the ages of 6 months and 6 years.
 4. Venous blood samples of lead levels are more reliable than capillary samples.
 5. The CDC defines lead poisoning as a blood lead level of 10 µg/dL or higher and these children should be rescreened at recommended intervals.
 6. Children who have blood lead levels above 14 µg/dL should have cognitive development evaluated and attempts made to identify the environmental source. Iron deficiency should be treated if present.
 7. Chelation of lead is indicated for levels of 45 µg/dL or more and is urgently required for levels over 70 µg/dL.
- Cholesterol Screening
 1. Universal screening for all children not recommended.
 2. Children 2 years of age and older who have a parent with a total cholesterol level of 240 mg/dL or greater should receive a total cholesterol screen.
 3. Children 2 years of age and older with a family history of premature cardiovascular disease (e.g., a parent or grandparent with a myocardial infarction, sudden cardiac death, angina pectoris, coronary arteriography for diagnostic purposes, or cardiac bypass surgery at the age of 55 years or younger) should be screened with a serum lipid profile.
 4. Overweight children are in a special risk category and should be screened regardless of family history or other risk factors.
 5. There are differences in cholesterol concentrations related to gender, as it is higher in females than males after pubertal development. There may be variations by ethnicity, with African-American children having a higher HDL and lower triglyceride than non-Hispanic white and Hispanic children.
 6. Children receiving total cholesterol screening may eat a normal diet before the test.
 - Cholesterol Screening
 7. Children receiving a serum lipid profile should fast for 12 hours (except for water) before their blood is drawn.
 8. An acceptable total cholesterol level in children and adolescents is less than 170 mg/dL; an acceptable LDL level is less than 110 mg/dL.
 9. An elevated total cholesterol level in children and adolescents is greater than or equal to 200 mg/dL; a high LDL level is greater than or equal to 130 mg/dL.
 - Urine Screening
 1. Routine screening recommended only if indicated by history and/or physical examination findings.
 2. Positive results of bacteriuria from a bagged urine specimen on an infant should be confirmed by catheterization or suprapubic aspiration.
 3. “Clean catch” midstream urine specimens in children and adolescents are best for reliable results.
 - Anemia Screening

Hemoglobin/hematocrit obtained between 9 and 12 months of age and for at risk children ages one through five
 - Sexually Transmitted Infections/HIV
 1. All sexually active adolescents (especially females who are often asymptomatic) should be screened for gonorrhea, chlamydia, syphilis, and trichomoniasis.
 3. Female adolescents who are sexually active and all females age 18 years and older should have a Pap smear performed at least every 3 years and more frequently if indicated by risk factors.
 4. High-risk adolescents offered HIV testing (e.g., those with multiple sexual partners, who reside in areas with a high prevalence of STI/HIV infection, who have been sexually abused by or have had sexual contact with individuals with documented STI/HIV infection or parenteral drug use)

■ SPECIAL EXAMINATIONS

- The Newborn Examination
 1. Immediately after birth, obtain APGAR scores at 1 and 5 minutes; composite scores range from 0 to 10 based on 5 criteria

Criteria	0	1	2
Heart rate	Absent	Slow (< 100)	> 100
Respiratory rate	Absent	Slow, irregular	Good strong cry
Muscle tone	Limp	Some flexion of extremities	Active motion
Reflex irritability	No response	Grimace	Cough or sneeze
Color	Blue, pale	Body pink, extremities blue	Completely pink

2. Obtain length, weight, head circumference percentiles, and vital signs
3. Assess vision and hearing
4. Assess gestational age (Ballard/Dubowitz exam)
5. HEENT
 - a. Palpate anterior and posterior fontanel—note presence of molding, craniosynostosis, cephalohematomas, and asymmetry
 - b. Assess presence of red reflex
 - c. Note size, shape, and position of ears and characteristics of TM
 - d. Assess for patency of nares, intact palate, and any unusual findings in the mouth
6. Assess neck for webbing, palpate for masses
7. Auscultate lungs and assess thorax for shape, symmetry, and character of respirations
8. Perform cardiac evaluation, including auscultation for murmurs and assessment of brachial and femoral pulses
9. Conduct abdominal examination, including the number of blood vessels in cord (two umbilical arteries and one umbilical vein), cord appearance, and condition of stump
10. Assess genitourinary system, including prominence of labia, number of testicles and position, position of urethra
11. Conduct musculoskeletal examination, including hips, feet, range of motion, presence of crepitus, Ortolani and Barlow maneuvers
12. Perform neurological examination, including assessment of head lag and muscle tone; illicit the following reflexes:
 - a. Root/suck
 - b. Gag reflex
 - c. Moro
 - d. Plantar/palmar
 - e. Stepping
 - f. Tonic neck

13. Assess skin for cyanosis, jaundice, meconium staining, rashes, lesions, and birthmarks
 - a. Major risk factors for jaundice*
 - (1) Jaundice in first 24 hours
 - (2) Blood group (ABO) incompatibility
 - (3) Gestational age less than 37 weeks, LBW
 - (4) Previous sibling received phototherapy
 - (5) Cephalohematoma or bruising
 - (6) Exclusive breastfeeding not going well
 - (7) East Asian race
 - b. Minor risk factors for jaundice*
 - (1) Gestational age 37–38 weeks
 - (2) Previous sibling with jaundice
 - (3) Macrosomic IDM
 - (4) Maternal age > 25
 - (5) Male
 - (6) Delayed cord clamping
- *AAP Guidelines for high risk factors for jaundice

- The Sports Evaluation

1. Purpose
 - a. Identify risk factors associated with morbidity and mortality
 - b. Identify conditions that place the child at risk for injury
 - c. Identify conditions which could worsen with sports participation
 - d. Determine appropriate sports activities for child's abilities
2. Preparticipation History
 - a. Cardiovascular—murmur, chest pain, syncope, shortness of breath, family history of cardiac diseases
 - b. Hypertension—130/75 mm Hg in a child under age 10 or 140/85 mm Hg in a child 10 years of age or older
 - c. History of chronic diseases
 - d. Musculoskeletal limitations and prior injuries
 - e. Menstrual history for females
 - f. Nutritional factors—recent weight loss or gain, eating patterns
 - g. Medication history—including performance enhancing compounds, stimulants, and narcotics
3. The physical examination should be comprehensive with emphasis on:
 - a. Cardiac examination, including blood pressure
 - (1) Presence of a systolic murmur that increases on sitting or with valsalva maneuver requires an echocardiogram

- (2) An arrhythmia that does not subside with exercise requires referral to a cardiologist
 - b. Musculoskeletal examination—including range of motion of neck, shoulders, upper and lower extremities, back, and gait.
 - (1) Perform scoliosis screen
 - (2) Have child perform the “duck walk” (squat on heels, walk 4 steps, and stand up)
 - (3) Assess shoulder flexion and rotation
 - c. Genitalia exam
 - (1) Tanner stage (refer to Chapter 2, Human Growth and Development)
 - (2) Presence of hernias
 - (3) Presence of both testicles in males
 - d. Skin for contagious lesions
 - e. Visual problems
 - f. Neurological problems
4. Estimate the relative risk of an acute injury to the athlete by categorizing sports as contact, limited contact, or noncontact sport. Contact sport includes the subset of collision sport. Collision implies greater injury risk. Clinical judgment involving the risk of acquiring a disease as a result of participation. Variables to consider:
- a. the advice of knowledgeable experts
 - b. the current health status of the athlete
 - c. the sport in which the athlete participates
 - d. the position played
 - e. the level of competition
 - f. the maturity of the competitor
 - g. the relative size of the athlete
 - h. the availability of effective protective equipment
 - i. the availability of efficacy of treatment
 - j. whether rehabilitation has been completed
 - k. whether the sport can be modified to allow safer participation
 - l. the ability of the athlete’s parent(s) or guardian and coach to understand and to accept the risks involved in participation
5. Exclusion from sports participation include:
- a. Carditis
 - b. Fever
6. Exclusion (until cleared) requiring consultation with a specialist and/or evaluation include:
- a. Atlantoaxial instability (as found in Down syndrome or juvenile rheumatoid arthritis with cervical involvement)
 - b. Bleeding disorder
 - c. Blood pressure reading at least 5 mm Hg above the 99th percentile
 - d. Congenital heart disease

- e. Dysrhythmia
- f. Heart murmur
- g. Structural or acquired heart disease
- h. Vasculitis/vascular disease
- i. Cerebral palsy
- j. Diarrhea
- k. Eating disorder
- l. Eye disorder
- m. Conjunctivitis, infectious
- n. Gastrointestinal disorder
- o. Heat illness, history of
- p. Absence of a kidney
- q. Hepatosplenomegaly
- r. Malignant neoplasm
- s. Neurological disorders
- t. Myopathies
- u. Recurrent plexopathy and cervical cord neuropraxia with persistent defects
- v. Seizure disorder, poorly controlled
- w. Organ transplant recipient
- x. Pregnancy/postpartum
- y. Respiratory conditions including pulmonary compromise such as cystic fibrosis, not asthma
- z. Acute upper respiratory infections
 - aa. Rheumatologic diseases
 - bb. Sickle cell disease
 - cc. Skin infections

If the athlete participates in sports despite known medical risks and against medical advice, the parents or guardian should be asked to sign a written consent statement indicating they have been advised of the potential dangers of participation and that they understand these dangers. The athlete should also sign the consent statement. It is recommended that the adult write the statement in their own words and handwriting.

- **Breast and Pelvic Examinations**

- 1. Breast
 - a. Although breast disease is rare in adolescent females, assessment of the breasts should be performed routinely as part of the well child physical examination from the start of puberty (as soon as breast budding occurs).
 - b. In addition to detection of disease, the breast examination is an opportunity to teach adolescents about breast development and self-breast examination as well as provide reassurance about the “normalcy” of their breasts.
- 2. Pelvic
 - a. Recognize the pelvic examination produces much fear and anxiety for adolescents, especially those who are experiencing it for the first time, have had

- a prior difficult experience, or have been sexually abused
- b. Prepare the adolescent for the examination by showing her illustrations of the reproductive system and explaining the procedure
- c. Provide concrete objective information about sensations the adolescent will feel during course of the examination which will help her cope better with the experience
- d. Suggest ways to maintain control and decrease anxiety during examination (e.g., relaxation techniques)
- e. Use largest speculum that will fit comfortably within vagina (usually small plastic or Pederson speculum work best for this age group)
- f. Warm speculum before insertion
- g. Inform adolescent of what you are doing throughout the examination
- h. Encourage the adolescent to become involved with the examination if she desires (e.g., a mirror can be positioned to see the area being examined)
- i. Verbal or visual modes of distraction are helpful (e.g., interesting posters on the ceiling or wall)
- j. Use examination to teach adolescent about her body and to reassure her of “normalcy”
- k. Provide as much privacy as possible, e.g., while dressing
- l. Give positive feedback to the adolescent for her cooperation or assistance
- m. Recognize that cervical ectopy is a normal finding during the examination, especially in adolescents taking hormonal contraceptives

■ PROBLEM-ORIENTED HEALTH RECORD

- Organized system for recording health visits to allow for thorough, concise data; easy retrieval of data; enhanced communication between health professionals; documentation of problem assessment/management; and decreased risk of liability
- Components of the problem-oriented health record
 1. Database—medical history, physical examination, growth charts, developmental flow sheets, screening and laboratory tests, and problem-specific progress notes
 2. Problem list—conditions that require diagnostic work-up or ongoing management

3. Management plan—includes information related to the diagnosis, management, education, and follow-up for specific health problem
4. Progress notes—includes documentation of each patient visit and is usually recorded in SOAP format
 - a. S—subjective information provided by child and parent or caregiver
 - b. O—objective information which consists of observations, physical examination, and laboratory findings
 - c. A—assessment (diagnosis)
 - d. P—plan
 - (1) Medications
 - (2) Treatments
 - (3) Further laboratory studies
 - (4) Consultations or referrals
 - (5) Diet or activity modifications
 - (6) Teaching
 - (7) Follow-up schedule (visit or telephone contact)

■ CHILD HEALTH SUPERVISION SCHEDULE OF VISITS WITH KEY ISSUES (NUTRITION, DEVELOPMENT, SCREENING, IMMUNIZATIONS, ANTICIPATORY GUIDANCE, AND HEALTH EDUCATION)

- Prenatal Visit
 1. General Considerations
 - a. Major purposes—ensure the health of the fetus, child, family; establish relationship with family; answer questions; provide anticipatory guidance and plan of care
 - b. Timing—between 30 to 35 weeks gestation
 - c. Include both parents and/or grandparents if single parent
 2. Family history—parents and siblings’ ages; health of parents, siblings, and blood relatives, including chronic illnesses such as asthma, cystic fibrosis, and heart disease, as well as mental disorders
 3. Obstetrical history—current gestational age; beginning of prenatal care; ultrasound or amniocentesis results; name of obstetrician; medications, drugs, cigarettes, or alcohol usage during this pregnancy; prior pregnancies and outcomes
 4. Preparation for childbirth and infant
 - a. Readiness for parenthood
 - b. Planned or unplanned pregnancy
 - c. Prenatal classes, childbirth preparation
 - d. Sibling preparation, if applicable
 - e. Choice of infant feeding
 - f. Circumcision, if applicable

- g. Genetic testing, if applicable
- h. Arrangements for child care
- i. Special concerns of prospective parents
- j. Current life stressors
- 5. Social history
 - a. Family type—single parent, nuclear
 - b. Number and types of pets
 - c. Perceived social support
 - d. Healthcare practices/religion
 - e. Financial information, including insurance
 - f. Cultural issues
- 6. Plan of care and anticipatory guidance
 - a. Timing of health supervision visits
 - b. Immunizations
 - c. Organization of practice, e.g., team approach with nurse practitioners and physicians
 - d. How and where to access care when needed; available hours
 - e. Fees/medical coverage
 - f. Need for transportation
 - g. General infant care and supplies needed
 - h. Safety information, e.g., car seats, pets
 - i. Psychological adjustment of parents and siblings
- Prenatal Breastfeeding Guidance
 1. Review the benefits of breastfeeding as appropriate during visit
 - a. Species specific
 - b. Benefits child's health
 - c. Maximizes potential of infant
 - d. Assists with birth spacing
 - e. Conserves resources
 - f. Benefits mother's health
 - g. Environmentally friendly
 - h. Empowers women
 - i. Promotes bonding
 2. Promote attainment of Healthy People 2010 breastfeeding goals
 - a. 75% initiate in the early postpartum period
 - b. 40% exclusively until 3 months of age
 - c. 50% continue until infant is 6 months
 - d. 17% exclusively until infant is 6 months
 - e. 25% continue until infant is 12 months
 3. Identify and clarify contraindications to breastfeeding prior to birth
 - a. Mother has active TB
 - b. Maternal HIV
 - c. Debilitating maternal disease (cancer)
 - d. Drug abuse—cocaine, heroin
 - e. Mother has HTLV1 (human T-cell leukemia virus type 1)
 4. Clarify misconceptions related to what is NOT a contraindication to breastfeeding prior to birth
 - a. Hepatitis B or C infection
 - b. Exposure to low level environmental contaminants
 - c. Alcohol use (limit to occasional drink)
 - d. Tobacco use (encourage to stop smoking or avoid infant exposure)
 - e. Cytomegalovirus (CMV) infection unless infant preterm
 5. Identify physical conditions of the breast that may inhibit breastfeeding and recommend lactation consult referral prior to delivery
 - a. Hypoplastic/Tubular breasts
 - b. No increase in breast size during pregnancy a risk factor
 - c. Breast surgery (augmentation/reduction)
 - d. Previous treatment for breast cancer
 - e. Trauma or burns to the breast
 6. Identify risk factors for lactation problems related to history/social factors and recommend lactation consult referral prior to delivery
 - a. Early intention to breast and formula feed
 - b. History of previous breastfeeding problems
 - c. History of hormone related infertility, intended OCP use
 - d. Significant medical problems (hypothyroidism, diabetes, PCOS)
 - e. Maternal age (adolescent or AMA)
 - f. Psychosocial problems, especially depression
 - g. Anticipated multiple birth or possible preterm delivery
 - h. Insufficient prenatal education about breastfeeding
 - i. Maternal employment and lack of knowledge about continuation of breastfeeding while employed
 - j. Lack of family and societal (workplace) support
 7. Incorporate best practices for the PNP to promote breastfeeding
 - a. Encourage prenatal breastfeeding education
 - b. Recommend human milk for all infants unless contraindicated
 - c. Encourage mother to initiate breastfeeding within the first hour of life
 - d. Encourage exclusive rooming in of newborn with mother

- Hospital Newborn Visit Breastfeeding Guidance
 1. Identify and clarify contraindications to breastfeeding present after birth of infant
 - a. Infant has galactosemia/inborn errors of metabolism
 - b. Mother has active herpetic lesions on her breast(s)
 - c. Mother has varicella that has been determined to be infectious to the infant
 - d. Medications including radioactive isotopes, antimetabolites, cancer chemotherapy
 2. Clarify misconceptions related to what is NOT a contraindication to breastfeeding present after birth of infant
 - a. Maternal fever in absence of other contraindications
 - b. Hepatitis B or C infection
 - c. Alcohol use (limit to occasional drink)
 - d. Tobacco use (encourage to stop smoking or avoid infant exposure)
 - e. Cytomegalovirus (CMV) infection unless infant preterm
 3. Promote breastfeeding in the first 24 hours
 - a. Promote skin to skin contact
 - b. Promote rooming in, avoid interruptions in breastfeeding
 - c. Teach feeding cues or feeding readiness; increased activity, mouthing, rooting
 - d. Remember the 4 As
 - (1) Alignment—body and head of infant
 - Positions
 1. Cross-cradle
 2. Clutch or football
 3. Side-lying
 4. Cradle or Madonna
 - (2) Areolar grasp
 - (3) Areolar compression
 - (4) Audible swallowing
 - e. Use positive supportive tone and body language
 4. Minimum output AAP guidelines:
 - a. First 24 hour: one stool one void
 - b. Second 24: 2 stools and 2 voids
 - c. Third and fourth days: 3 stools and 5 voids
 - d. Meconium stool: day 1–2
 - e. Transitional stool: day 3–4
 - f. Breastfed stool: day 4–5 on
 5. Normal breastfeeding patterns:
 - a. 1st day: baby will feed 6–8 times in 24 hrs
 - b. 2nd and 3rd days: baby will cluster feed
 - c. 4th and 5th days: baby will feed 10–12 times a day
 6. Identify risk factors for lactation problems identified after birth and recommend lactation consult prior to discharge
 - a. Nipple trauma
 - b. Pain throughout feed
 - c. Infant weight loss; –8% birth weight (less than 2–3% per day)
 - d. Inadequate output
 - e. None or little swallowing after 24 hours of age
 - f. Abnormal infant oral anatomy
 - g. Mother of NICU infant/separation from infant
 - h. Late preterm (34–36 6/7), SGA, LGA
 - i. Jaundice infant
 - j. Multiple births
 - k. Birth interventions
 - l. Unrelieved fullness or engorgement
 7. Pumping guidelines requiring a hospital pump
 - a. Infants in NICU
 - b. Infants with weight loss \geq 8%
 - c. Infants close to 24 hours old and have not breastfed
 - d. Inverted, flat, or sore nipples
 - e. Using breast shield
 - f. History of breast surgery
 - g. Medical conditions, i.e., hemorrhage or shock
 8. Provide information to mother regarding milk volume at week two if exclusively providing pumped breastmilk
 - a. Ideal: 750 to 1000 ml/day
 - b. Adequate: 500 ml/day
 - c. Borderline: < 350 ml/day
 9. Breastmilk storage guidelines

a. Room Temperature	4–8 hrs
b. Refrigerator	3–5 days
c. Freezer	3–6 months
d. Deep Freeze	1 year
e. Thawed, refrigerated	24 hours
f. Store in opaque, clear or brown glass, or breastmilk storage bags to maintain maximum number of functional cells	
 10. Signs of good feeding after discharge
 - a. Feeling good, strong deep sucking, without sharp pain
 - b. Audible swallowing (at 18 hours of life)
 - c. Breast softer
 - d. Adequate output
 - e. Seeing milk in baby's mouth
 - f. Leaking from other breast
 11. Signs of poor feeding after discharge
 - a. Sleepy baby
 - b. Baby has difficulty latching on
 - c. Pain throughout feeding
 - d. Clicking or popping sounds in baby's mouth
 - e. Inadequate output

- f. Less than 8 feedings per day or more than 12 feedings per day
- 12. Incorporate best practices for the PNP to promote breastfeeding
 - a. Avoid supplementation unless medically indicated
 - b. Avoid pacifier use until breastfeeding well established
 - c. Provide timely hospital follow-up
- Newborn Visit
 - 1. Purposes—obtain complete history; physical examination; anticipatory guidance, and plan of care
 - 2. Initial history—from chart review and parent(s)
 - a. General health of mother
 - (1) Mother's age and gestation at first prenatal visit, regularity of visits
 - (2) History of maternal infection and prenatal complications
 - (3) Substance abuse, tobacco use
 - (4) History of chronic disease
 - (5) Medications during pregnancy
 - (6) Weight gain, nutritional status
 - (7) Number of weeks of gestation
 - (8) Number of living children
 - b. Labor and delivery
 - (1) Length of labor
 - (2) Medications used—anesthesia
 - (3) Type of facility—birthing center, hospital, other
 - (4) Type of delivery—spontaneous, C-section (explanation needed if yes)
 - (5) Blood type, including Rh factor
 - c. Infant's status at delivery
 - (1) Identification verified
 - (2) Term, preterm (number of weeks)
 - (3) Determination of gestational age (Ballard/Dubowitz exam)
 - (4) Apgar scores
 - (5) Complications
 - (6) Oxygen or any other treatment(s) required
 - (7) Blood type, Rh factor; other values
 - (8) Length, weight, head and chest circumference (including percentiles)
 - (9) Physical examination results—note any abnormal findings
 - (10) Correct date of birth
 - d. Nursery course—presence of jaundice; type of treatments; medications, immunizations; length of stay; circumcision/cord condition
 - e. Family history—maternal/paternal
 - (1) Review of systems (ROS)—includes physical and mental health problems
- (2) Home environment—members of household, pets, smokers, infant supplies, and sleeping arrangements
- f. Common parental concerns
 - (1) Initial weight loss, appearance of infant (cephalohematoma, molding), normalcy of infant
 - (2) Rashes, skin markings—telangiectasis, café-au-lait, hemangiomas
 - (3) Infant's habits—feeding, stooling, sleeping, development, normal crying
 - (4) Female infant—breast engorgement, vaginal discharge
- g. Objective data
 - (1) Verify identification of mother, infant
 - (2) Parent/infant interaction—eye contact; holding; response to crying, vocalization
 - (3) Behavioral—consolability, self-quieting
 - (4) Physical examination (refer to Newborn Examination under Special Examinations in this chapter)
- h. Plan of care
 - (1) Reinforce infant care, cord care, circumcision care, instructions on when to seek medical advice
 - (2) Emphasize individual variability of infant temperament, noting positive aspects and challenges
 - (3) Breastfeeding mothers
 - (a) Quiet alert state optimal time for nursing
 - (b) Initial feedings 3 to 5 minutes/each breast every 1 ½ to 2 hours; increase time as tolerated
 - (c) Supplemental formula unnecessary
 - (d) Colostrum present first 2 to 4 days
 - (e) Increase maternal fluid intake especially at 6 weeks and 3 months during infant growth spurts
 - (f) Adequate infant intake indicated by 6 or more voids/day
 - (g) Vitamins unnecessary unless at risk for nutritional deficiencies, e.g., inadequate sunlight as vitamin D source
 - (4) Bottle-feeding mothers
 - (a) Amount and frequency varies
 - (1) 0 to 1 month—2 to 4 oz every 3 to 4 hours
 - (2) 2 to 4 months—5 to 7 oz every 4 to 5 hours
 - (b) Inform them that iron-fortified formulas are best

- (c) Suggest partial hydrolysate formula if infant at risk for atopic disease, e.g., strong family history of atopic disease
 - (5) Crying—infant's first way of communicating needs
 - (a) Less than 2 hours/day during first month; peaks at 6 weeks
 - (b) Decreases as infant learns other ways to communicate
 - (6) Colic—excessive crying in otherwise healthy infants beginning at 2 to 3 weeks; may last 3 to 4 months
 - (a) Rule out physical problems
 - (b) Assess for over/under feeding
 - (c) Acknowledge stressful situation; encourage breaks from infant if possible; reassure that it usually subsides by 3 to 4 months
 - (d) Avoid sudden and overstimulation of infant
 - (e) If nursing, eliminate possible offending sources from maternal diet, e.g., coffee, spices, chocolate, milk, gas-forming foods
 - (f) Soothing techniques—rocking, walking, background “white noise,” car rides
 - (g) Antiflatulent if indicated by history and examination
 - (7) Sleep patterns
 - (a) Usually sleeps 8 hours by 3 months
 - (b) Early introduction of solids—doesn't cause infants to sleep through the night
 - (c) Position on side or supine as SIDS preventive measure unless medically contraindicated
 - (8) Bowel movements
 - (a) Usually after feeding (especially if breast fed); normal variation may be up to one/week
 - (b) Avoid laxatives; use stool softener if constipated
 - (9) Reminder to schedule office visit for 2 weeks-of-age
- Newborn Breastfeeding Guidance
 1. Promote exclusive breastfeeding for approximately the first 6 months
 2. Provide a formal evaluation of breastfeeding including observation, position, latch, milk transfer
 3. Identify maternal risk factors for breastfeeding problems
 - a. Inverted, short, or flat nipple
 - b. Sore nipples/nipple pain and causes
 - (1) Overly full breasts-engorgement
 - (2) Poor positioning and/or latch or improper pumping
 - (3) Not breaking suction correctly
 - (4) Anatomical oral variations of newborn or poor fit of mother to infant or inappropriate infant sucking
 - (5) Inappropriate cleaning of nipples (alcohol/drying agents)
 - (6) Infection due to bacterial or candidal infection
 - c. Engorgement
 - (1) Normal breast fullness typically peaks day 4–5, resolving without treatment
 - (2) Signs of engorgement is evidenced by continual breast fullness/edema, breast pain, and/or flattened nipples often causing difficulty latching
 - (3) Predisposing Factors for Engorgement
 - (a) Mother—delayed or missed breastfeeding, early supplements, flat or inverted nipples, breast surgery, abrupt weaning
 - (b) Infant—ineffective or poor latch, sleepy infant or poor feeder, prematurity/SGA, jaundice, physical abnormalities of mouth (tight frenulum/tongue tie Ankyloglossia)
 - (4) Overproduction or pathologic engorgement
 - (a) Continual breast fullness at 3 weeks postpartum and beyond, usually with forceful milk ejection
 - (b) Possible infant signs include:
 - (1) Chokes, coughs, sputters, arches with feeding
 - (2) Gassy, irritable, restless, frequent crying
 - (3) Green, thin, frothy stools
 - (4) Rapid weight gain (note that many symptoms similar to reflux in infant)
 - (c) Milk duct obstruction
 - (d) Mastitis—cause
 - (1) Infrequent or missed feedings
 - (2) Poor latch leading to insufficient milk removal
 - (3) Nipple damage
 - (4) Illness in mother or baby
 - (5) Oversupply of milk
 - (6) Rapid weaning
 - (7) Pressure on the breast—tight bra, car seatbelt
 - (8) Blocked milk duct or blocked nipple pore

- (9) Maternal stress and fatigue
 - (10) Maternal malnutrition or anemia
 - d. Insufficient milk supply—causes
 - (1) Most common cause is infrequent feedings or early introduction of formula (without pumping)
 - (2) Insufficient milk transfer as evidenced by poor output or inadequate weight gain in infant
 - (3) Conditions of mother—fatigue, stress, medications that inhibit milk production (estrogen-containing contraception, sudafed), psychological inhibition, pregnancy, or smoking
 - (4) Fussy infant for other reasons or growth spurt of infant
 - (5) At four weeks postpartum breasts naturally less full prior to feeding
 - e. Bleb, Vasospasm
 - f. Maternal medical problems—separation
- 4. Identify infant risk factors for breastfeeding problems
 - a. Jaundice (physiologic and pathologic) or other medical problems (hypoglycemia, respiratory distress, infection)
 - b. Ankyloglossia (Tongue-tied) or other oral abnormality
 - (1) Incidence in the population is predicted to be 4.8%
 - (2) Genetic prevalence
 - (3) More prevalent in males 2.6 to 1.0
 - c. Late preterm/low birth weight
 - d. Multiples
 - e. Neurologic problems/genetic syndromes
- First Newborn Office History Questions Related to Breastfeeding
 - 1. Number of feedings/wet diapers/stools in last 24 hours
 - 2. Does newborn wake for feedings?
 - 3. Does newborn easily latch and breastfeed eagerly?
 - 4. Is breastfeeding painful or uncomfortable?
 - 5. Is newborn receiving any supplements?
 - 6. Has mother previously breastfed and if yes, successful?
 - 7. Is mother taking any medications or dietary supplements?
 - 8. How is mother's nutrition/weight reduction/special diet?
 - 9. Family support at home
- Examining Newborn and Mother Related to Breastfeeding
 - 1. Calculate newborn's weight gain or loss since birth
- 2. Perform routine exam and oral motor assessment
- 3. Assess state of hydration
- 4. Observe for jaundice
- 5. Observe breastfeeding
- 6. Examine mother's breasts or refer for examination if necessary
- 7. Consider test weight to estimate milk volume consumed
- Anticipatory Guidance Related to Breastfeeding
 - 1. Encourage breastfeeding on demand
 - 2. Review normal breastfeeding patterns
 - 3. Discourage use of pacifiers and discuss potential risks
 - a. AAP recommends avoiding use until breastfeeding is established (about 3 weeks) as use is associated with:
 - (a) Decreased weight gain
 - (b) Nipple confusion
 - (c) Early weaning
 - 4. Avoid long nighttime intervals without feeding
 - 5. Review normal elimination patterns
 - 6. Reinforce the importance of care of the mother
 - 7. Avoid maternal use of alcohol, tobacco, and caffeine
 - 8. Discuss maternal use of medications
 - a. Drugs may transfer into human milk if they:
 - (1) Attain high concentrations in maternal plasma
 - (2) Are low in molecular weight (< 500)
 - (3) Are low in protein binding (drugs that circulate are either bound to albumin or freely soluble in plasma). Look for levels > 90%
 - (4) Are very lipid soluble
 - b. General guidelines for medications and breastfeeding
 - (1) Premature infants may not be able to tolerate medications like full-term infants
 - (2) During early lactation (day 1–3) and late stage lactation (the amount of milk produced, 30–100 ml a day) is so low that infant receives limited amounts in milk
 - (3) Avoid nursing at times of peak drug concentration
 - (4) Administer the drug before the infant's longest sleep period
 - c. Lactation Risk Categories
 - (1) L1 safest
 - (2) L2 safer
 - (3) L3 moderately safe
 - (4) L4 possibly hazardous
 - (5) L5 contraindicated

- Breastfeeding Interventions
 1. Attempt to determine and treat the cause of inadequate milk supply before supplementing
 2. Consider referral to lactation consultant if problems ongoing
 3. Identify an appropriate peer support group
- How Father of Baby or Support Can Help with Breastfeeding
 1. Assist with positioning
 2. Help with sleepy baby
 3. Assist mother into comfortable position
 4. Get the baby ready for feed by changing diaper
 5. Monitor mom's fatigue level
 6. Monitor visitors
- Closing the Visit Related to Breastfeeding
 1. Congratulate parents on decision to breastfeed their infant
 2. Review some of the benefits of breastfeeding
 3. Remind mother to take the time to establish regular food and fluid intake to meet her needs
 4. Arrange for appropriate follow-up until weight gain is adequate and breastfeeding is going well
- Interim Visits—2 Weeks to 1 Year (2 weeks; 2, 4, 6, 9, & 12 months)
 1. Subjective information
 - a. Feeding—type, amounts, time at breast, name of formula
 - b. Elimination—frequency, color, consistency of stools, number of saturated diapers per day
 - c. Sleep
 - d. Development, including behavior and temperament
 - e. Concerns of parent, caregiver
 - f. Interval history—health since last visit, emergency care, illnesses, medications used
 2. Objective data
 - a. Caregiver/infant interaction
 - b. Length, weight, head circumference; including percentiles
 - c. Vital signs
 - d. Vision and hearing screening (refer to screening guidelines this chapter)
 - e. Physical examination
 - f. Developmental screening—Denver II or equivalent and results; autism screening at 9 months
 - g. Laboratory
 - (1) Newborn screening (metabolic screen)
 - (2) Hematocrit at 12 months or sooner if indicated
- (3) Lead screening by 12 months as appropriate based on well child surveillance of risk (CDC)
- 3. Plan of care, including health promotion strategies and anticipatory guidance
 - a. Administer immunizations as indicated after obtaining consent and assessing response to previous immunizations—contact Centers for Disease Control and Prevention and the American Academy of Pediatrics for updated recommended childhood and adolescent schedules
 - (1) Inform parents of common side effects of immunizations; there is a Web site designed and maintained by the Centers for Disease Control (CDC) that has the most up-to-date information about immunization and possible side effects.
<http://www.cdc.gov/vaccines/recs/schedules/default.htm>
 - (2) The National Childhood Vaccine Injury Act of 1986 requires all health providers who administer vaccines to report occurrences of certain adverse events stipulated in the Act—the Vaccine Adverse Event Reporting System (VAERS) Web site has the most up-to-date listing of reportable events and the associated time intervals from vaccine administration. Additionally, providers may report adverse effects online using this Web site;
<http://vaers.hhs.gov/>
 - b. Inform parents of any medications prescribed, including name, rationale, dosage, frequency, course, and potential side effects
 - c. Implement health promotion strategies and anticipatory guidance appropriate for age and developmental level (ideally should be done before or at the very beginning of a developmental stage or problem); best to focus on one or two per visit with input from the parent or caregiver
- 4. Anticipatory guidance—nutrition and feeding
 - a. Nutritional requirements are 110 to 120 calories/kg/day
 - b. Consumption of more than 32 oz of formula per day usually indicative of need for solids once an infant is at least 4 months of age
 - c. Formula recommended up to 1 year of age; whole milk until two years of age

- d. Judicious use of juices, some fortified with vitamin C; best to place in cup versus bottle
 - e. Best to avoid giving infants a bottle in bed, especially containing formula or juice (leads to dental caries)
 - f. Introduction of solids
 - (1) Usually best to delay introduction of solids until 6 months of age; earlier introduction may lead to overfeeding and allergies
 - (2) Cereal usually first food added to diet followed by vegetables, fruits, and meats
 - (3) Add only 1 new food every 3 to 5 days
 - (4) Do not give common allergenic foods until one year of age, e.g., cow's milk, egg whites, wheat, citrus fruits, peanut butter
 - (5) Avoid the following until 3 years of age to prevent choking—nuts; potato chips; popcorn; raw celery and carrots; hotdogs; fish with bones; tough meat; small, hard candies
 - g. Introduction of cup—when infant loses interest in bottle (7 to 10 months), spout cup may be used initially
 - h. Weaning (breast or bottle)—gradually decrease number of feedings over several weeks; night feeding usually last to be eliminated
5. Anticipatory guidance—sleep
 - a. 2- to 4-month-old infant—8 to 12 hours at night; 2 to 3 naps
 - b. 6- to 12-month-old infant—11 to 12 hours at night; 2 to 3 naps
 - c. Side-lying or supine sleep position through 6 months
 - d. Strategies for parents
 - (1) Put infant in crib when drowsy rather than asleep
 - (2) Quiet, nonstimulating night feedings
 - (3) Night waking once night feedings stop—briefly stroke infant lightly; don't pick infant up
 - (4) Avoid pattern of placing infant in parent's bed
 6. Anticipatory guidance—infant stimulation
 - a. Provide variety of age-appropriate stimulation
 - b. Formal infant stimulation programs for infants at risk for developmental delays
 7. Anticipatory guidance—teeth
 - a. Eruption typically begins at approximately 6 months of age
 - b. Signs and symptoms—local sensitivity and inflammation, increased drooling, biting, irritability
 - c. No evidence that teething causes fever, diarrhea, or other systemic illnesses
 - d. Comfort measures—hard rubber teething toy, chilled teething ring, wet wash cloth, avoid liquid filled teething rings
8. Stranger/anxiety—emerging awareness and preference for mother/primary caregiver; early indicator of healthy attachment process emerging around 6 months
 9. Separation anxiety—emerging awareness that infant is an individual distinct from primary attachment figure/caregiver
 - a. Develops around 9 months; peaks at around 18 months
 - b. Suggestions for parents
 - (1) Recognize that bedtime, going to childcare, having a childcare provider at home are all separations
 - (2) Gradually introduce child to new situations and caretakers
 - (3) The child learns to accept separation through multiple, brief separations and reunions
 - (4) Games such as “peek-a-boo” and “hide-and-seek” may be helpful
 10. Safety—prevention of ingestion/aspiration of foreign objects
 - a. Keep pins, buttons, and other small objects off the floor and out of reach
 - b. Do not feed infants hard foods (e.g., nuts, hard candies)
 - c. Discard broken or cracked rattles with beads
 - d. Do not give infant balloons
 - e. Do not prop infant's bottle
 - f. Select pacifier with shield too large to enter infant's mouth
 - g. Learn emergency procedure for dealing effectively with choking
 11. Injury prevention
 - a. Keep electrical wires out of reach; outlets covered; cabinet safety locks; medicines, poisons out of reach
 - b. Install smoke detectors in household
 - c. Keep Poison Control telephone number available, other emergency phone numbers
 - d. Use car seat appropriate for infant's weight and age; rear facing until 12 months and 20 pounds
 - e. Avoid walkers and stairs; use gate to barricade doorways and unsafe areas, e.g., kitchen and bathroom

- f. Do not place necklaces or pacifier cords around infant's neck
 - g. Discourage infant jewelry
 - h. Position crib or playpen away from window blind or curtain cords
 - i. Use a crib with side rails spaced no more than $2\frac{3}{8}$ inches apart
 - j. Secure infant with belt on changing table
 - k. Lower crib mattress when infant begins to stand
12. Water safety—prevention of drowning and burns
 - a. Set hot water heater temperature at 120°F or less
 - b. Do not leave infant unattended in bathtub
 - c. Do not allow child to play in water unattended, e.g., toilet bowls, sinks, buckets of water
 - d. Use of safety devices around swimming pools, lakes, and in boats, e.g., life jackets
 - e. Lock fences around swimming pools
 - f. Continuous supervision around water
 - g. Do not leave other children in charge of infants around any body of water
 13. Sun safety and sunburn protection—use lotion with sun protective factor (SPF) of at least 15; avoid use around eyes; use caps with sun visors or bonnets to protect eyes; keep infants in shaded areas, even with sunscreen; sun avoidance for infants < 6 months
 14. Alternative child care arrangements—discuss childcare provider options; appropriate supervision and stimulation/activities (language, visual, and motor)
 15. Emphasize positive qualities of the infant and the parent-child relationship and mutually agree upon a plan with the parent to strengthen needed areas
 16. Initiate referrals to other nurse practitioners, physicians, clinics, and community resources as needed
 17. Schedule for next visit
- Infant Breastfeeding Guidance 2 Weeks to One Year
 1. Early Infancy
 - a. Discuss stool pattern/feeding pattern/night waking
 - b. Discuss mother's plans to return to work and breastmilk expression and storage
 - c. Provide 400 IU of oral vitamin D at 2 months of age
 - d. Discuss maternal use of medications
 - e. Assess milk supply
 - f. Encourage exclusive breastfeeding until 6 months of age
 2. Six Months
 - a. Discuss stool pattern/feeding pattern/night waking/distractibility/teething
 - b. Discuss mother's plans/adjustment to return to work and breastmilk expression and storage
 - c. Provide supplementary fluoride at 6 months of age
 - d. Discuss maternal use of medications
 - e. Assess milk supply
 - f. Introduction solids/high iron foods
 3. Nine Months
 - a. Discuss stool pattern/breastfeeding pattern/decreasing frequency/fluids in cup
 - b. Assess milk supply
 - c. Assess pressures to wean
 - d. Discuss maternal use of medications
 4. Twelve Months
 - a. Discuss stool pattern/breastfeeding pattern/decreasing frequency/fluids in cup
 - b. Discuss weaning techniques if desired, benefits of breastfeeding a toddler
 - Interim Visits—Toddler (1 to 3 years) (12, 15, 18, & 24 months)
 1. Subjective data—nutrition/appetite, elimination, sleep, development, parental concerns, health since last visit
 2. Objective Data
 - a. Assess parent-child interaction and parenting style
 - b. Height, weight, head circumference (up to 2 years), including percentiles; Body Mass Index starting at 24 months, including percentiles
 - c. Vital signs
 - d. Vision and hearing screening (refer to screening guidelines this chapter)
 - e. Physical examination
 - f. Developmental screening—Denver II or equivalent and results; autism screening at 18 and 30 months
 - g. Laboratory
 - (1) Hematocrit at 1 year of age
 - (2) Lead screening by 2 years of age as appropriate based on well child surveillance of risk (CDC)
 3. Management
 - a. Update immunizations—contact Centers for Disease Control and Prevention and the American Academy of Pediatrics for updated recommended childhood and adolescent schedules; inform parents of common side-effects of immunizations <http://www.cdc.gov/vaccines/recs/schedules/default.htm>

- b. Advise parents of any medications prescribed, including vitamins and fluoride, e.g., name, rationale, dosage, frequency, course, and potential side effects
- c. Nutrition appropriate for age
 - (1) Nutritional requirements are 102 cal/kg
 - (2) Provide foods from the five major food groups in realistic amounts; $\frac{1}{4}$ to $\frac{1}{3}$ of adult portion or one measuring tablespoon for each year of the child's age
 - (3) Physiological anorexia is common between 15 and 18 months of age
 - (4) Parental concerns regarding nutrition—decreased appetite, food jags, rituals, variable intake, definite likes and dislikes
 - (5) Potential nutritional problems—iron deficiency anemia (especially if toddler is drinking more than 32 oz of milk per day); low intake of calcium, vitamin A, zinc, ascorbic acid; obesity
 - (6) Provide regular meals and snacks
 - (7) Avoid food battles
- d. Toilet Training
 - (1) Most children are psychologically and physiologically ready between 18 to 30 months
 - (2) Majority of children achieve daytime bowel and bladder training simultaneously; average age is 28 months
 - (3) Nighttime control generally occurs about 1 year after daytime control is achieved
 - (4) Toilet training should not be started when family is unduly stressed (e.g., new baby, moving, holidays, divorce)
 - (5) Suggestions for parents—praise all efforts; expect accidents to happen; don't punish; if child is resistant, try again in a few weeks; follow usual pattern of elimination; limit time on potty to 5 to 10 minutes
- e. Sleep
 - (1) One to 3 year olds usually sleep 10 to 12 hours at night and take 1 to 2 naps
 - (2) Toddlers need rituals and consistency at bedtime as well as security objects, e.g., blanket, special toy
 - (3) Nightmares start at approximately 3 years; child generally awakens and remembers dream
 - (4) Night terrors generally occur between 2 to 4 years; child does not waken
 - (5) Tips for parents—quietly reassure child; let child fall asleep in own bed
- f. Negativism
 - (1) Normal behaviors in toddlers as they strive to develop sense of autonomy
 - (2) Expressed as “no,” temper tantrums, breath holding
 - (3) Tips for parents
 - (a) Give child as much control as possible (e.g., offer choice of two acceptable objects or actions); allow independence when possible
 - (b) Ignore temper tantrums, however expect a “response burst” initially when ignored
- g. Sibling rivalry
 - (1) Can occur throughout childhood; is often most troublesome just after birth of new baby if older sibling is under 2 years
 - (2) Tips for parents
 - (a) Involve children in preparation for new baby
 - (b) Praise “big-kid” behaviors; ignore regression
 - (c) Provide special time for each child every day
 - (d) Stay out of minor sibling conflicts, but discipline if aggression occurs
 - (e) Foster individual interests of each child and avoid comparing children
- h. Thumb Sucking—peaks between 18 to 21 months; ignore before age 4 unless child is not thriving
- i. Safety—apply recommendations given earlier that are appropriate for toddlers, plus:
 - (1) Emphasize street safety—begin teaching the basic rules of pedestrian and traffic safety
 - (2) Avoid strangers—teach child methods to avoid encounters with strangers which can be harmful
 - (3) Continued use of car seats according to weight—all toddlers should ride forward-facing and upright (studies show safest place for the child is middle of back seat)
 - (4) Keep medicine and poisons out of reach in locked cabinets—highest incidence of poisoning occurs in 2-year-olds
 - (5) Use of helmets with any type of bicycle use, either as a passenger or alone
 - (6) Enroll child in swimming lessons and always supervise when in or around water

- j. Dental care—use of toothbrush; first dental visit recommended at approximately 3 years of age
 - k. Parenting and Discipline
 - (1) Use limit setting and time-out for inappropriate behavior—1 minute of time-out for each year of age
 - (2) Praise good behavior
 - (3) Emphasize consistency, especially between parents
 - (4) Spend time with child; read to child on a daily basis
 - (5) Parents set example as role models
 - (6) Assign chores appropriate for age, e.g., put toys away
 - (7) Begin socialization with other children
 - l. Television
 - (1) Limit television viewing to appropriate children's programs
 - (2) Limit time spent watching TV
 - (3) Strong correlation between viewing aggression-type programs and child's level of aggressive play
 - (4) TV fosters negative outcomes—rapid paced, superficial problem-solving; obesity
 - (5) Watch TV with child and provide reality base; opportunity to discuss values and stereotypes
 - (6) Set good example through parental TV habits
 - m. Child care arrangements—discuss child care provider options; head start programs, nursery school, preschool programs
 - n. Emphasize positive qualities of the child and the parent-child relationship and mutually agree upon a plan with the parent to strengthen needed areas
 - o. Initiate referrals to other nurse practitioners, physicians, clinics, and community resources as needed
 - p. Schedule for next visit
- Interim Visits for Preschool Child (Ages 3, 4, & 5 years)
 1. Subjective information—nutrition/appetite; elimination; sleep; development, including behavior and temperament; caregiver concerns; interim health since last visit including stressful life changes, e.g., move, divorce
 2. Objective Data
 - a. Assess parent-child interaction and parenting style
 - b. Height, weight, body mass index including percentiles
 - c. Vital signs, including blood pressure
 - d. Vision and hearing screening (refer to screening guidelines this chapter)
 - e. Physical examination
 - f. Developmental screening—Denver II or equivalent and results
 - g. Laboratory
 - (1) Mantoux test for TB screening between 4 and 6 years of age if at risk
 - (2) Hematocrit if indicated
 - (3) Cholesterol screening if indicated
3. Management
 - a. Immunizations—contact Centers for Disease Control and Prevention and the American Academy of Pediatrics for updated recommended childhood and adolescent schedules; review status and update as indicated; inform parents of common side-effects of immunizations <http://www.cdc.gov/vaccines/recs/schedules/default.htm>
 - b. Advise parents of any medications prescribed, including vitamins and fluoride, e.g., name, rationale, dosage frequency, and potential side effects
 - c. Nutrition appropriate for age
 - (1) Nutritional requirements are 90 cal/kg
 - (2) Provide foods from the five major food groups in realistic amounts
 - (3) Provide nutritious snacks (e.g., fruits, vegetables) instead of high-fat foods (e.g., potato chips, cookies)
 - (4) Potential nutritional problems (same as in the toddler years)
 - d. Elimination—expect occasional nighttime accidents until 4 years of age; do not punish or embarrass child
 - e. Sleep
 - (1) The 3- to 5-year-old child usually sleeps 8 to 12 hours per night and gradually eliminates naps
 - (2) Prepare parents for an increase in nightmares
 - f. Development
 - (1) Discuss the change from the pleasing 3-year-old to the sometimes aggressive, frustrating behavior of the 4-year-old; provide reassurance that “calmness” usually begins at age 5 years
 - (2) Emphasize the “magical thinking” of the preschool child and how it places the child at risk for unintentional injury and an increase in fears

- (3) Discuss the child's rigid superego and how children this age feel guilty for negative events
 - (4) Inform parents that they can expect "tall tales," the use of "toilet talk," and the construction of imaginary playmates
 - (5) Discuss need for peer companionship and school readiness
 - g. Sex education—begins in infancy and toddlerhood when parents label the genitals and accept genital exploration and masturbation as normal activities
 - (1) Preschool children are curious about gender differences and "how babies are made"; their questions should be answered briefly and accurately
 - (2) Explore parents' feelings about masturbation and prepare for a possible increase in this behavior
 - (3) Talk with child about inappropriate touch; encourage parents to reinforce prevention
 - h. Dental care
 - (1) Brush teeth after each meal if possible and at bedtime
 - (2) Recommend first dental visit if not previously done
 - (3) If thumb sucking is persistent at age 4, have dental evaluation to rule out malocclusion; speech evaluation if tongue thrust is suspected
 - i. Safety—apply recommendations given earlier that are appropriate for preschool children, plus:
 - (1) Reinforce pedestrian safety—children ages 3 to 7 years are more frequently involved in pedestrian-related motor vehicle accidents
 - (2) Teach guidelines for bicycle safety; avoid busy streets
 - (3) Devise fire escape plan and teach the child; conduct routine fire drills
 - (4) Teach child to stop, drop, and roll if clothing catches on fire
 - (5) Teach safety—the danger of matches, open flames
 - (6) Always supervise child when swimming or near any water
 - (7) Frequently check playground equipment for stability, loose nuts and bolts, and suitable landing surface
 - (8) Keep child away from power equipment, including lawn mowers
 - (9) Never clean or handle a gun in the presence of the child
 - (10) Discard old refrigerators and other large appliances or remove all doors during storage to avoid entrapment during play or exploration
 - (11) Do not allow chewing gum or eating while running or jumping; parent should be aware of intervention for choking
 - j. Exercise—encourage frequent periods of outdoor activities and limit television viewing
 - k. Emphasize positive qualities of the child and the parent-child relationship and mutually agree upon a plan with the parent to strengthen needed areas
 - l. Initiate referrals to other nurse practitioners, physicians, clinics, and community resources as needed
 - m. Schedule for next visit
- Interim Visits: School-Age Children—annual visit (6 to 12 years)
 - 1. Subjective Data
 - a. Nutrition/appetite
 - b. Elimination—especially constipation and enuresis
 - c. Sleep
 - d. Development—include school, activities, exercise, friends, behavior, and family relationships
 - e. Allergies and reactions
 - f. Medications
 - g. Drug, alcohol, cigarette, and caffeine usage (for older school-age child)
 - h. Concerns of child and caregiver; current/recent stressors, e.g., move, divorce
 - i. Interim health since last visit (include review of systems); for older school-age children, obtain sexual/reproductive history as indicated
 - 2. Objective
 - a. Assess parent-child interaction and child behavior
 - b. Height, weight, body mass index, including percentiles
 - c. Vital signs, including blood pressure
 - d. Vision and hearing screening (refer to screening guidelines this chapter)
 - e. Physical examination, including assessment for scoliosis and Tanner staging
 - f. Laboratory
 - (1) Mantoux test for TB screening of high-risk child
 - (2) Hematocrit if indicated
 - (3) Cholesterol if indicated

3. Management

- a. Immunizations—review status and update as indicated—contact Centers for Disease Control and Prevention and the American Academy of Pediatrics for updated recommended childhood and adolescent schedules; inform parents of common side effects
<http://www.cdc.gov/vaccines/recs/schedules/default.htm>
- b. Advise parents of any medications prescribed, including vitamins and fluoride, e.g., on name, rationale, dosage, frequency, and potential side effects
- c. Nutrition
 - (1) Nutritional requirements are approximately 70 cal/kg
 - (2) Most behavioral problems with food resolved
 - (3) Potential nutritional problems—obesity, iron deficiency
 - (4) Encourage good nutritional practices (e.g., balanced meals, nutritious snacks, no meal skipping)
- d. Development characteristics and behaviors
 - (1) Ages 6 to 7—nervous mannerisms, restless activity, egocentric thinking, rigid superego
 - (2) Ages 8 to 10—takes on idols and heroes, friends serve as allies against adults, less rigid superego
 - (3) Ages 10 to 12—increased self-awareness and self-consciousness; body image concerns; mood swings, stormy behavior; need for independence
 - (4) Inform parents that use of inappropriate language may occur
 - (5) Prepare for pubertal changes and menstruation in girls
 - (6) Discuss with parents the importance of and strategies for bolstering the child's self-esteem
- e. Good health habits—assist child and parents to establish early patterns of behavior, e.g., regular exercise; sufficient sleep; regular dental care; avoidance of drugs, alcohol, tobacco
- f. Communication—encourage parents to assist child in developing good communication skills, problem-solving strategies, and stress management
- g. Drugs, smoking, and alcohol—encourage parent/child discussions
- h. Sex education—anatomy and physiology, sexual activity, values clarification,

decision making, contraception, prevention of STIs and HIV

- i. Parenting strategies
 - (1) Although the child is maturing, quality time, attention, and affection from parents is important (evidence exists supporting “connectedness” to parents and/or another adult decreases risk-taking behaviors)
 - (2) Encourage independence and decision-making
 - (3) Promote responsibility and accountability by assigning appropriate chores
 - (4) Maintain adequate supervision
 - (5) Discuss methods of discipline; use positive reinforcement and appropriate consequences for inappropriate behavior
 - (6) Establish fair rules
 - (7) Respect the child's need for privacy
 - (8) Set example by being a good role model
 - (9) Provide child with an allowance
 - (10) Encourage movies and TV programming and video games that are appropriate for child's developmental level
 - (11) Praise child for achievements
 - (12) Encourage consistent school attendance
 - (13) Refer parents to resources that can assist them in building assets and preventing psychosocial morbidities; see Melnyk et al. (2003) for the KySS (Keep your children/yourself Safe and Secure) Campaign strategic plan, a national effort by the National Association of Pediatric Nurse Practitioners (NAPNAP) to decrease emotional and behavioral problems in children and adolescents.
- j. Safety
 - (1) Bicycle, skateboarding, rollerblading; use of helmets and protective padding
 - (2) Seat belt use
 - (3) Prevention of sexual abuse, e.g., inappropriate touching; what to do if it occurs
 - (4) Water safety; swimming
 - (5) Fire prevention safety; home fire drill, proper use of appliances
 - (6) Sunburn prevention
 - (7) Prevention of violence in home; lock up guns, ammunition; teach child firearm safety (avoidance of firearms)
 - (8) Pedestrian safety

- k. Lying and cheating (common in school-age children)
 - (1) School-age children lie to avoid trouble or gain an advantage
 - (2) Appropriate response—confront child in a positive way, try to understand reason for lie, follow through with age-appropriate discipline when needed
 - (3) Adults should model honesty
 - l. Sports
 - (1) For 6- to 8-year-olds, sports participation should be noncompetitive and focused on learning rules, teamwork, and having fun
 - (2) Older school-aged children and teenagers should have a preparticipation physical examination
 - m. Emphasize positive qualities of the child and the parent-child relationship and mutually agree upon a plan with the parent to strengthen needed areas
 - n. Initiate referrals as needed
 - o. Schedule for next visit
- Interim Visits: Adolescent—annual visit (13 to 18 years)
 1. Subjective information—gather “sensitive” information when alone with the adolescent
 - a. Nutrition—especially appetite; meal-skipping
 - b. Elimination—especially use of aids such as laxatives, diuretics
 - c. Sleep practices
 - d. Development—special emphasis on assessment of mental and emotional health (including school performance and attendance; self-esteem, friends and relationships; family functioning and “connectedness”; hobbies; activities; work; stress and anger management; coping skills; risk-taking behaviors, such as driving while drinking and use of weapons; violent or aggressive behavior; ideations about hurting self or others)
 - e. Allergies
 - f. Medications
 - g. Drug, alcohol, cigarette, and caffeine consumption
 - h. Sexual activities, reproductive issues
 - i. Concerns/worries; current and recent stressors
 - j. Interim health since last visit—include review of systems, with special emphasis on gathering psychological and sexual/reproductive data
 - k. Specific questions when alone with parent
 - (1) Family communication patterns and relationship with the adolescent
 - (2) Parent’s description of the adolescent’s strengths and areas needing improvement; attitudes and behaviors
 - (3) Discipline practices and response
 - (4) Specific concerns and worries about the adolescent
 2. Objective data
 - a. Observation of parent-adolescent interaction (e.g., parental support of adolescent; does the parent allow the adolescent to answer questions)
 - b. Height and weight including percentiles
 - c. Vital signs, including blood pressure
 - d. Vision and hearing screening (refer to screening guidelines this chapter)
 - e. Physical examination—including scoliosis screen, Tanner stage, and breast examination; pelvic examination if adolescent female is sexually active, having irregular menses, or is more than 16 years of age
 - f. Laboratory
 - (1) Urinalysis if sexually active
 - (2) Hematocrit if indicated
 - (3) VDRL, GC, Chlamydia, and HIV if sexually active or history of sexual abuse
 - (4) Pap smear if pelvic examination performed
 - (5) Liver function tests (if history of drug usage); cholesterol, if indicated
 - (6) Mantoux test for TB screening if at risk
 3. Management
 - a. Update immunizations—contact Centers for Disease Control and Prevention and the American Academy of Pediatrics for updated recommended childhood and adolescent schedules; Hepatitis B vaccine series should be encouraged for all adolescents who may not have received the immunization in their earlier childhood years, especially sexually active adolescents or those who are using IV drugs; provide information on common side effects
<http://www.cdc.gov/vaccines/recs/schedules/default.htm>
 - b. Advise regarding any medications prescribed, e.g., include name, rationale, dosage, frequency, course, and potential side effects
 - c. Nutrition appropriate for age
 - (1) The adolescent diet should be similar to an active adult, with extra calories during rapid growth periods; prudent consumption of high fat foods, e.g., red meats, butter, and eggs

- (2) Nutritional issues—irregular meals, chaotic lifestyle; increase in meals eaten away from home; increase in snacks; skipping of meals; fad diets; vegetarianism
- (3) Potential nutritional problems—increased need for calcium, iron, zinc; eating disorders; obesity
- (4) Encourage well-balanced meals and nutritious snacks
- (5) Discuss adolescent's perception of his/her weight
- (6) Encourage healthy weight loss strategies if indicated, e.g., healthy food choices, regular exercise
- d. Safety—same as school age child, plus:
 - (1) Emphasize the possible consequences of drinking and use of drugs while driving
 - (2) Discourage being a passenger when the driver has been drinking or using drugs
 - (3) Discuss typical high-risk situations and how to avoid them; role play healthy behaviors to use in high-risk situations
 - (4) Encourage safe swimming and diving practices
 - (5) Discuss proper use of safe sports equipment and maintenance
 - (6) Instruct adolescent in proper training and warm-up exercises for sports and physical activities
 - (7) Educate regarding safe and proper use of firearms and other potentially dangerous objects such as firecrackers
 - (8) Discuss use and misuse of over-the-counter medications
- e. Developmental Issues
 - (1) Discuss dating and peer pressure
 - (2) Encourage open communication with parents, peers, and school personnel
 - (3) Teach stress reduction techniques and coping skills
 - (4) Educate regarding healthy outlets for anger
 - (5) Discuss plans for the future—further education, work, recreation, hobbies, marriage, parenthood
 - (6) Educate regarding acne—e.g., cause, myths, and proper skin care
- f. Discuss issues related to sexuality—decision making; mature relationships; assertiveness; safe sex/prevention of STIs, including HIV; pregnancy and contraception; implications of potential parenthood; “normalcy” of occasional masturbation
- g. Encourage good health habits—e.g., regular exercise, sufficient sleep, regular dental care
- h. Continue education regarding use of drugs, alcohol, cigarettes, and caffeine
- i. Inform parents of major developmental characteristics of adolescents
 - (1) Increased self-awareness, self-consciousness, and self-appraisal
 - (2) Body image concerns
 - (3) Mood swings and stormy behavior
 - (4) Need for independence
 - (5) Using peer values as criteria with which to judge own values, but still needing family to provide acceptance and feeling of self-worth
 - (6) Interest in opposite sex
- j. Parenting strategies with adolescents
 - (1) Fairness in rules and reasonable limit setting
 - (2) Allow decision making
 - (3) Respect adolescent's privacy
 - (4) Expect periods of estrangement (be available; adolescent needs supportive family)
 - (5) Praise achievements at home, school, extracurricular activities
 - (6) Bolster self-esteem
 - (7) Supervision as needed
 - (8) Encourage independence, new experiences, after-school activities, including part-time job
 - (9) Promote family communication
 - (10) Serve as role model—practice good health habits, e.g., parents should not smoke if they do not want child to imitate their behavior
 - (11) Recognize signs of probable substance abuse—drop in school performance, personality change, mood swings, sleepiness or fatigue, depression
- k. Emphasize positive qualities of the adolescent and the parent-adolescent relationship and mutually agree upon a plan to strengthen needed areas
- l. Make referrals as needed
- m. Refer parents to resources that can assist them in building their teen's assets as well as with their parenting skills
- n. Schedule for next visit

■ QUESTIONS

Select the best answer

1. Sixteen-year-old Sarah makes the following statements to you during a health visit. Which of the following pieces of information should not be kept confidential?
 - a. "I have been sexually active with three of my boyfriends."
 - b. "I sometimes smoke marijuana."
 - c. "I want to get pregnant."
 - d. "Sometimes I feel like ending my life."
2. In performing a physical examination on a nine-month-old infant, which of the following developmental fears would not be appropriate for you to consider?
 - a. Stranger anxiety
 - b. Pain
 - c. Separation from parents
 - d. Bodily harm
3. When performing a physical examination on a toddler, which of the following body parts would you examine last?
 - a. Heart and lungs
 - b. Abdomen and genitals
 - c. Ears and throat
 - d. Hips and extremities
4. Role play with equipment during the course of a physical examination would be most beneficial with which of the following age groups?
 - a. Toddlers
 - b. Preschoolers
 - c. Young school-age children
 - d. Older school-age children
5. Providing reassurance of "normalcy" during the course of an examination would be most important for:
 - a. Preschool children
 - b. Young school-age children
 - c. Older school-age children
 - d. Adolescents
6. Which of the following would not elevate the pulse of a child?
 - a. Fever
 - b. Anemia
 - c. Hypothyroidism
 - d. Exercise
7. The PNP recognizes which of the following signs as indicators that baby is not receiving sufficient breastmilk?
 - a. sleepiness, jaundice, decreased urine and stool
 - b. diarrhea, nausea, and vomiting
 - c. bulging fontanel and irritability
 - d. sleeplessness and excitability
8. Blood pressure should be measured at well child visits, beginning at age:
 - a. 2 years
 - b. 3 years
 - c. 4 years
 - d. 5 years
9. A wide pulse pressure that results from a high systolic blood pressure is usually not due to which of the following?
 - a. Fever
 - b. Exercise
 - c. Excitement
 - d. A patent ductus arteriosus
10. Head and chest circumferences should be equal at:
 - a. 6 months of age
 - b. 1 year of age
 - c. 2 years of age
 - d. 3 years of age
11. The anterior fontanel usually closes by:
 - a. 2 months of age
 - b. 6 months of age
 - c. 18 months of age
 - d. 24 months of age
12. Diffuse edema of the soft tissue of the scalp which usually crosses suture lines in the newborn is:
 - a. Bossing
 - b. Caput succedaneum
 - c. Cephalohematoma
 - d. Macrocephaly
13. An infant should no longer have a head lag when pulled from the supine to sitting position at what age?
 - a. 2 months
 - b. 4 months
 - c. 6 months
 - d. 9 months
14. "Boggy" nasal mucous membranes with serous drainage upon examination usually suggests:
 - a. Sinusitis
 - b. Polyps

- c. URI
- d. Allergic rhinitis
15. A white instead of red reflex upon eye examination of a 1-year-old child would suggest:
 - a. An accommodative error
 - b. Retinoblastoma
 - c. Papilledema
 - d. Retinal detachment
16. A cobblestone appearance of the palpebral conjunctiva usually indicates:
 - a. Bacterial infection
 - b. Chemical irritation
 - c. Viral infection
 - d. Severe allergy
17. An eye that deviates in when covered but returns to midline when uncovered is an:
 - a. Esophoria
 - b. Exophoria
 - c. Esotropia
 - d. Exotropia
18. Pain produced by manipulation of the auricle or pressure on the tragus suggests:
 - a. Acute otitis media
 - b. Otitis externa
 - c. Otitis media with effusion
 - d. Mastoiditis
19. A hypernasal voice and snoring in a child is suggestive of:
 - a. Polyps of the larynx
 - b. Nasopharyngeal tumor
 - c. Hypertrophied adenoids
 - d. Cleft palate
20. Physiological splitting of the second heart sound during inspiration in a child:
 - a. Is normal
 - b. Should be evaluated with an EKG
 - c. Suggests an ASD
 - d. Should be referred to a cardiologist
21. Which of the following is not characteristic of innocent heart murmurs in children?
 - a. Systolic in timing
 - b. Varies in loudness with positioning
 - c. Usually transmitted to the neck
 - d. Usually loudest at lower left sternal border or at second or third intercostal space
22. A Grade II musical or vibratory murmur heard best at the lower left sternal border that changes with positioning is suggestive of a:
 - a. Pulmonary ejection murmur
 - b. Ventricular septal defect
 - c. Venous hum
 - d. Vibratory or Still's murmur
23. Wheezing in a child may not be found in which of the following conditions?
 - a. Asthma
 - b. Bronchiolitis
 - c. Pleural friction rub
 - d. Cystic fibrosis
24. Gynecomastia in a male may not be a finding in which of the following?
 - a. Normal pubertal development
 - b. Steroid usage
 - c. Hyperthyroidism
 - d. Testicular tumor
25. Which of the following would usually not be considered a sign of a pituitary tumor in an adolescent female?
 - a. Dysfunctional uterine bleeding
 - b. Galactorrhea
 - c. Loss of peripheral vision
 - d. Increase in headaches
26. Which of the following is not a specific examination test for a dislocated hip?
 - a. Barlow's test
 - b. Ortolani's test
 - c. Trendelenburg test
 - d. Gower's test
27. In addition to the knee, which of the following should be examined in a child complaining of knee pain?
 - a. Foot
 - b. Ankle
 - c. Hip
 - d. Spine
28. Which of the following infant reflexes should not disappear by 6 months of age?
 - a. Moro
 - b. Rooting
 - c. Tonic neck
 - d. Plantar grasp
29. Spasticity in an infant may be an early sign of:
 - a. Neurofibromatosis
 - b. Hydrocephalus

- c. Cerebral palsy
 - d. Muscular dystrophy
30. A shift to the left is present when which of the following are elevated?
- a. Neutrophils
 - b. Bands or stabs
 - c. Lymphocytes
 - d. Eosinophils
31. Which of the following is usually elevated with viral infections?
- a. Neutrophils
 - b. Eosinophils
 - c. Lymphocytes
 - d. Basophils
32. Decreased platelets may not be found in which of the following?
- a. Leukemia
 - b. Anemia
 - c. ITP
 - d. Medication usage (e.g., ampicillin, cephalothin)
33. Which of the following does not suggest a urinary tract infection?
- a. Increased protein
 - b. Increased WBCs
 - c. Increased RBCs
 - d. Increased nitrites
34. A Mantoux test in a child with no risk factors is considered positive with a reaction of:
- a. At least 5 mm induration
 - b. At least 8 mm induration
 - c. At least 10 mm induration
 - d. At least 15 mm induration
35. The PNP teaches new parents that when breastfeeding is well established they can expect baby to have:
- a. as many as 4 wet diapers each day
 - b. a stool every 2–3 days
 - c. 1 wet diaper an hour
 - d. 5–6 wet diapers and 2–3 stools each day
36. Cholesterol screening should be done:
- a. Children 2 years of age and older who have a parent with a total cholesterol level of 240 mg/dL or greater
 - b. Once for all children at 6 years of age
 - c. Overweight children with a family history of premature cardiovascular disease
 - d. Once for all children at 12 years of age
37. For which of the following screening tests should children fast for 12 hours before the test is done?
- a. Total cholesterol
 - b. Serum chemistry profile
 - c. Serum lipid profile
 - d. Hematocrit
38. Which of the following is the most important history-taking question for a sports evaluation?
- a. Has the child ever had a head injury?
 - b. Has the child ever fainted or lost consciousness during exercise?
 - c. Does the child ever get short of breath with exercise?
 - d. Has the child ever had prior surgeries?
39. Which of the following conditions would exclude a child from participating in contact collision sports?
- a. Fever
 - b. Absence of a paired organ
 - c. Atlantoaxial instability
 - d. Prior head injury
40. Which of the following topics would not be appropriate to include when providing anticipatory guidance to the parent of a 4-month-old infant?
- a. Introduction of solid foods
 - b. Teething
 - c. Negativism
 - d. Introduction of a cup
41. Which is the correct order for introduction of solid foods to an infant?
- a. Fruits, cereal, vegetables, and meats
 - b. Cereal, meats, vegetables, and fruits
 - c. Fruits, cereal, meats, and vegetables
 - d. Cereal, vegetables, fruits, and meats
42. Which of the following topics is not appropriate to include when providing anticipatory guidance to the parent of an 18-month-old?
- a. Temper tantrums
 - b. Toilet training
 - c. Dental care
 - d. Stranger anxiety
43. Appropriate anticipatory guidance for the parents of an 8-year-old girl should not include:
- a. Preparation for an increase in nervous mannerisms and restless activity
 - b. Preparation for pubertal changes
 - c. Information that friends begin to serve as allies against adults

- d. Information that their daughter will take on idols and heroes
44. A new mom calls the PNP on post partum day 5. She reports her newborn wants to nurse for 30 minutes every 2 hours. Which of the following is your best response?
- "This is a very healthy breastfeeding pattern. Be sure to rest when you can. You are doing a great job."
 - "Your baby is too demanding. If you continue to feed that often you will spoil your child."
 - "You are not making enough milk and your baby will need to go to the ER to be evaluated."
 - "Your baby has an oral fixation and you should offer a pacifier to relieve stress."
45. A pelvic examination should not be performed on which of the following adolescents?
- A 14-year-old who is sexually active
 - A 15-year-old who has just started menarche
 - A 17-year-old who is having irregular menses
 - An 18-year-old healthy female

ANSWERS

- | | |
|-------|-------|
| 1. d | 24. c |
| 2. d | 25. a |
| 3. c | 26. d |
| 4. b | 27. c |
| 5. d | 28. d |
| 6. c | 29. c |
| 7. a | 30. b |
| 8. b | 31. c |
| 9. d | 32. b |
| 10. b | 33. a |
| 11. c | 34. d |
| 12. b | 35. d |
| 13. c | 36. a |
| 14. d | 37. c |
| 15. b | 38. b |
| 16. d | 39. a |
| 17. a | 40. c |
| 18. b | 41. d |
| 19. c | 42. d |
| 20. a | 43. a |
| 21. c | 44. a |
| 22. d | 45. b |
| 23. c | |

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Eye, Ear, Nose, Mouth, and Throat Disorders

Karen Buch

■ HEAD

■ NORMAL HEAD GROWTH

Birth head size : 32–38 cm (average 34 cm)	
0–3 months:	2 cm/month = 6 cm
4–6 months:	1 cm/month = 3 cm
6–12 months:	0.5 cm/month = 3 cm

GROWTH IN FIRST YEAR: 12 cm

Subsequent head growth—0.5 cm/year 2–7 years of age; 0.3 cm 8–12 years

Compare head circumference (HC) with chest circumference:

Newborn:	HC 2 cm > chest
Infancy:	HC equal or slightly larger;
> 2 years:	Chest > head

Microcephaly

- Definition: Head circumference (HC) 2 to 3 SD or more below the mean for age, sex, and gestation, or HC percentiles that drop with increasing age
- Etiology/Incidence (2.5% of total population)
 1. Primary: Caused by a variety of genetic/chromosomal disorders
 2. Secondary: Acquired from multiple causes including:
 - a. Intrauterine infections, e.g., TORCH—toxoplasmosis, other agents [HIV, listeria, syphilis, gonococcus, group β streptococcus, varicella-zoster, malaria], rubella,

cytomegalovirus inclusion disease, herpes simplex

- b. Fetal exposure to toxic substances including alcohol, medications (e.g., phenytoin), phenylketones (maternal PKU)
- c. Fetal exposure to radiation in first and/or second trimesters
- d. Placental insufficiency, prenatal hypoxia, trauma, maternal hypoglycemia, degenerative diseases (e.g., Tay Sachs)
- e. Extremely poor nutrition during the first six months of life
- f. Significant risk for isolated microcephaly—alcohol use, inadequate weight gain during pregnancy, inadequate prenatal care, African-American, and low educational level

- Signs and Symptoms
 1. Small head—in full term birth and up to six months of age, chest circumference surpasses head circumference (unless child is very obese)
 2. Delayed developmental milestones, neurological problems, mental retardation
 3. Family history of microcephaly
- Differential Diagnosis
 1. Craniosynostosis
 2. Endocrine disorders, e.g., hypopituitary function, hypothyroidism
 3. Severe malnutrition

- Physical Findings
 1. Early closure of fontanel; prominent cranial sutures
 2. Marked downward sloped forehead with narrowed temporal diameter and occipital flattening (familial microcephaly)
 3. Skull asymmetries, high arched palate, and dysplastic teeth (dysmorphic and chromosomal disorders)
- Diagnostic Tests/Findings
 1. Karyotyping for chromosomal disorders, e.g., fragile X syndrome
 2. Antibody titers for TORCH infections of the mother and child
 3. Test infant serum and urine for amino and organic acids
 4. CT or MRI to detect calcification, malformations, or atrophy
- Management/Treatment
 1. Complete history and physical findings crucial to finding treatable disorders, e.g., hypopituitarism, severe protein-calorie malnutrition
 2. Premature closure of sutures may be surgically corrected
 3. Most disorders are untreatable; accurate diagnosis is essential for appropriate genetic and family counseling
 4. Management is supportive; especially placement in a program that will maximize development since many are mentally retarded

Macrocephaly

- Definition: Head circumference greater than 2 to 3 SD from the mean for age, sex, and gestation, or one that increases too rapidly
- Etiology/Incidence
 1. Many causes including:
 - a. Hydrocephalus—excess cerebrospinal fluid
 - b. Intracranial lesions—neoplasms, subdural effusions
 - c. Enlargement of the skull—primary skeletal dysplasias
 - d. Increase in brain size (megalencephaly)—seen in neurofibromatosis
 2. Normal head growth with large heads: Familial macrocephaly (benign)
- Signs and Symptoms
 1. Excessive head growth
 2. Dependent on cause, e.g., hydrocephalus

- Differential Diagnosis
 1. Benign macrocephaly—familial or catch-up growth in thriving premature infant
 2. Pathological macrocephaly
- Physical Findings
 1. Progressive excessive head growth greater than 2 to 3 SD above the mean for age/sex as delineated on growth chart
 2. Transillumination of skull may reveal chronic subdural effusions, hydrocephaly, or large cystic defects
 3. Anterior fontanel may be enlarged/tense with wide suture lines
 4. Depending on the cause, may have signs of increased intracranial pressure
 5. May have extensive findings depending on underlying cause, such as developmental delays, skeletal dysplasia, ocular abnormalities, skin findings (café-au-lait spots)
- Diagnostic Tests/Findings: CT, MRI, or ultrasonography of anterior fontanel (if not closed) can:
 1. Define structural cause; determine if operable disorder
 2. If benign, provide more accurate diagnosis, prognosis, guide to management, and genetic counseling
 3. Provide basis of comparison if future study is necessary
 4. Skull x-rays can provide indirect information such as changes seen with increased intracranial pressure, primary skeletal dysplasias, or calcifications
- Management/Treatment
 1. CT, MRI, or ultrasound of head if any signs/symptoms of increased intracranial pressure
 2. Surgical correction; closure of sutures of some disorders such as shunt placement with hydrocephalus or tumor resection
 3. Genetic counseling

Hydrocephalus

- Definition: Increased production, impaired absorption, or obstruction in flow of cerebrospinal fluid (CSF), leading to excessive CSF in the cerebral ventricles and ventricular enlargement
- Etiology/Incidence
 1. Obstruction of CSF flow anywhere along its pathway; CSF is produced in the ventricles, exits into subarachnoid space (area of reabsorption)

2. Causes
 - a. Congenital, due to primary cerebral malformation
 - b. Acquired postnatally; secondary to tumor, hemorrhage, or CNS infection (e.g., meningitis)
 - c. Choroid plexus papilloma—overproduction of CSF; rare cause
 3. Types
 - a. Obstructive or noncommunicating hydrocephalus
 - (1) Major cause of hydrocephalus
 - (2) Obstruction of CSF within ventricular system before CSF reaches subarachnoid space
 - (3) Sites include aqueduct of Sylvius and obstruction of fourth ventricle
 - (4) Congenital defects—most common cause; can be isolated (aqueductal stenosis) or part of a syndrome (Arnold-Chiari and Dandy-Walker)
 - (5) Acquired conditions—common complication of tumors
 - b. Nonobstructive or communicating hydrocephalus
 - (1) Impairment of CSF absorption within subarachnoid space
 - (2) Usually caused by scarring due to subarachnoid hemorrhage (result of intraventricular hemorrhage in the premature infant) or meningitis
- Signs and Symptoms: Presentation is variable depending on age and etiology
 1. Birth to 12 months—apparent large head, sluggish feeding, vomiting, piercing cry, and irritability
 2. 12 months through adolescence—signs of increased intracranial pressure (ICP), headache following sleep, lethargy or irritability, confusion, personality changes, possible decline in academic performance, signs and symptoms related to specific focal lesion or tumor, possible sixth nerve palsies, chronic papilledema
 - Differential Diagnosis
 1. Macrocephaly
 2. Megalencephaly
 3. Benign large head
 4. Macrocrania
 5. Meningitis
 6. Sepsis
 7. Tumor
 - Physical Findings
 1. Infancy—bulging anterior fontanel, scalp vein distention, bossing, “setting sun sign,” separated skull sutures, slow PERRL, hypertonia, hyperreflexia, spasticity
 2. Childhood—strabismus, extrapyramidal tract signs (ataxia), papilledema, optic atrophy, growth failure from endocrine dysfunction
 - Diagnostic Tests/Findings
 1. Cranial radiograph—separated sutures
 2. CT scan/ultrasound—impaired CSF circulation causing ventricular enlargement
 3. Ventriculography—obstruction detection
 - Management/Treatment
 1. Most cases require extracranial shunts, particularly ventriculoperitoneal types
 2. Treatment of underlying cause, e.g., mass or lesion, inflammation, infection, vasogenic edema
 3. Anticipatory guidance for families and caregivers throughout child’s life
 - a. Referral to support groups
 - b. Teaching signs and symptoms of ICP
 - c. Daily head circumference measurements
 - d. Management of psychomotor challenges
- ## Caput Succedaneum
- Definition: Diffuse swelling of soft tissue of the scalp, which usually crosses suture lines with possible bruising
 - Etiology/Incidence: Result of compression or trauma to scalp during descent of the baby through birth canal, causing edema of scalp
 - Signs and Symptoms
 1. Non-pitting swelling of the scalp, typically present at birth, tending to overlie the occipital bones and some of the parietal bones.
 2. Some cases with bruising; often with molding
 - Differential Diagnosis
 1. Cephalohematoma
 2. Subgaleal hematoma
 - Diagnostic Tests/Findings: None
 - Management/Treatment
 1. Usually unnecessary with spontaneous resolution within a few days
 2. If extensive ecchymosis, may require phototherapy for hyperbilirubinemia

Cephalohematoma

- Definition: Subperiosteal collection of blood bound by suture lines, usually those surrounding the parietal bones
- Etiology/Incidence
 1. Approximately 0.4% to 2.5% of all deliveries
 2. Most commonly due to a low subperiosteal bleed, secondary to trauma from a difficult delivery (e.g., large infant, prolonged labor, forcep use, abnormal presentation)
 3. Uncommonly may be due to a coagulopathy or intracranial hemorrhage
 4. 25% overlies a linear skull fracture—not depressed; rarely of any clinical significance
- Signs and Symptoms
 1. Not evident at birth but typically presents hours to days after delivery
 2. Limited to periosteum at suture margins; usually unilateral, but occasionally bilateral
 3. May prolong neonatal jaundice from the resorption of a large hematoma
- Differential Diagnosis
 1. Caput succedaneum
 2. Cranial meningocele
- Physical Findings: Nonecchymotic swelling of parietal area that does not cross suture lines; soft, fluctuant
- Diagnostic Tests: None
- Management/Treatment
 1. Usually unnecessary with slow resolution over a few weeks/months
 2. Calcification of hematoma possible; may be felt as bony prominence
 3. Observance for hyperbilirubinemia

Craniosynostosis

- Definition: Premature closure of one or more of the cranial sutures resulting in a skull deformity; named according to the suture that is closed
- Etiology
 1. Primary—due to abnormalities of skull development; cause unknown in majority of cases but in approximately 10% to 20% of cases due to a genetic syndrome or familial pattern
 2. Secondary—due to failure of brain growth (microcephaly); uncommon cause

- Signs/Symptoms
 1. Abnormal head shape
 2. Head size often small but depends on which suture is closed
 3. Typically no symptoms unless part of a genetic syndrome or multiple suture closures; rarely may develop signs/symptoms of increased intracranial pressure
 4. FH of abnormal head shape and/or genetic syndrome
- Differential Diagnosis
 1. Molding in newborn
 2. Pseudo-synostosis (positioning)
- Physical Exam
 1. Abnormal head shape
 2. Palpable bony ridge along affected suture line
 3. Anterior fontanel may be large or small depending on suture closing
- Diagnostic Tests/Findings
 1. Plain skull x-rays can identify
 2. Chromosomal analysis if indicated
- Management
 1. Surgery to open fused suture
 2. Performed primarily for cosmetic reasons, unless neurologic complications

■ EYE

Conjunctivitis of the Newborn (Ophthalmia Neonatorum)

- Definition: Infection and/or inflammation of conjunctiva in first month of life
- Etiology/Incidence
 1. Chemical—irritation from use of silver nitrate or other ophthalmic preparations (occurs in 10% of newborns); erythromycin ophthalmic ointment commonly used in place of silver nitrate with less irritation
 2. Gonococcal—perinatal transmission of *Neisseria gonorrhoeae*; prophylactic treatment with erythromycin ophthalmic ointment 0.5%, topical silver nitrate 1%, tetracycline 1%, or povidone-iodine 2.5%
 3. Chlamydia (Inclusion)—perinatal transmission of *Chlamydia trachomatis* during vaginal birth; most common cause of neonatal conjunctivitis in the U.S. with an incidence of 8.2/1000 births; silver nitrate is not effective against chlamydia

4. Herpes simplex virus (HSV)—uncommon cause but may cause loss of vision and needs immediate attention; up to 70% of infants born to asymptomatic mothers
 5. Other pathogens after first week of life include *Haemophilus influenzae*, *Staphylococcus aureus*, enterococci Group B streptococcus
- Signs and Symptoms
 1. Chemical—mild injection of conjunctiva, presents several hours following ophthalmic drop/ointment instillation lasting no longer than 3 to 4 days
 2. Gonococcal—acute purulent discharge 2 to 4 days after birth
 3. Chlamydia—usually presents with mild mucopurulent discharge (may be profuse) a few days to several weeks after birth (typically 5th to 14th day)
 4. Herpes—symptoms variable; eye redness, watery discharge usually begins within 2 weeks. May have systemic symptoms.
 5. Other bacteria—purulent discharge normally seen on 2nd to 5th day
 - Differential Diagnosis
 1. Dacryostenosis
 2. Foreign body
 3. Corneal abrasion
 - Physical Findings
 1. Chemical—conjunctival hyperemia, minimal lid edema, scanty discharge
 2. Gonococcal—chemosis, significant lid edema, purulent discharge
 3. Chlamydia—minimal lid edema, conjunctival hyperemia, chemosis, possible concomitant pneumonia (afebrile, repetitive staccato cough, tachypnea, and rales)
 4. Herpes—usually unilateral, may have small eyelid vesicles, mild conjunctivitis; possible neurological or systemic symptoms (lethargy, poor feeding)
 5. Other bacteria—mucopurulent discharge, mild symptoms
 - Diagnostic Tests/Findings
 1. Gram and Giemsa stain—gonococci revealed as intracellular gram negative diplococci; chlamydia is revealed as basophilic intracytoplasmic inclusions in the conjunctival epithelial cells
 2. Cultures—positive for specific organism
 3. Direct fluorescent antibody staining or enzyme immunoassay—detection of HSV anti-
- gens; fluorescein staining of eye may detect ocular dendritic ulcers
4. Immunofluorescent antibody staining of conjunctival scrapings for chlamydia—highly sensitive and specific
- Management/Treatment
 1. Chemical—no treatment necessary; resolves spontaneously without sequelae; decreased incidence with use of erythromycin ophthalmic ointment in place of silver nitrate
 2. Gonococcal
 - a. Ocular emergency (can cause blindness); hospitalization is necessary;
 - b. Eye should be irrigated initially with normal saline every 10 to 30 minutes, gradually decreasing to every 2-hour intervals until purulent discharge is cleared
 - c. Ceftriaxone 25 to 50 mg/kg/day, IV or IM not to exceed 125 mg given as a single dose; cefotaxime 100 mg/kg/day, IV or IM given as a single dose is an alternative; topical treatment alone is inadequate and unnecessary when systemic therapy is given
 3. Chlamydia conjunctivitis and pneumonia—oral erythromycin 50 mg/kg/day in four divided doses for 10 to 14 days; oral sulfonamides may be used after immediate neonatal period for infants intolerant to erythromycin; topical treatment ineffective and unnecessary; efficacy of erythromycin therapy is 80%, second course sometimes required
 4. Herpes simplex virus
 - a. Hospitalization is required
 - b. Acyclovir is first line (parenteral)
 - c. Topical ophthalmic preparations adjunct therapy; 0.1% iododeoxyuridine, 1–2% trifluridine, or 3% vidarabine
 5. Other bacteria—topical antibiotics such as erythromycin, polymixin-bacitracin, and tobramycin are adequate; warm compresses to remove exudate
 6. Stress thorough hand washing, no sharing of washcloths or towels
 7. Mothers and sexual partner(s) should also be treated appropriately

Conjunctivitis of Childhood

- Definition: Inflammation and/or infection of palpebral (lining of eyelids) and bulbar (layer of tissue over the sclera) conjunctiva in children one month or older

- Etiology/Incidence
 1. Most frequent ocular disorder in children
 2. Very contagious, especially in daycare and school settings
 3. Bacterial conjunctivitis—most common pathogens include *Staphylococcus aureus*, *Haemophilus influenzae*, and *Streptococcus pneumoniae*; Conjunctivitis-otitis syndrome occurs in about 25% of young children (< 3 years) and most often associated with non-typeable *H. influenzae*; typically in ipsilateral eye/ear
 4. Viral conjunctivitis primarily due to adenovirus 3, 4, and 7; uncommonly caused by herpes simplex and varicella zoster; adenoviral conjunctivitis can occur in isolation or as part of a viral illness (e.g., pharyngo-conjunctival fever)
 5. Allergic and vernal (chronic allergic) conjunctivitis—often with unidentified cause; commonly associated with seasonal allergens (pollens, grasses)
- Signs and Symptoms
 1. Pruritus, foreign body sensation, tearing
 2. Mild eye discomfort (burning, pain); severe pain suggests another diagnosis
 3. Possible sensitivity to light (mild)
 4. Discharge
 - a. Viral, allergic—watery or thick/stringy mucoid
 - b. Bacterial—purulent, glued eyes (lids stuck together after sleeping)
- Differential Diagnosis
 1. Nasolacrimal duct obstruction
 2. Blepharitis in older children
 3. Keratitis—can be seen with herpes simplex conjunctivitis or severe adenovirus (e.g., epidemic keratoconjunctivitis)
 4. Systemic infection presenting with conjunctivitis, e.g., rubella, rubeola, Kawasaki
 5. Periorbital cellulitis
- Physical Findings
 1. Discharge, rhinitis
 2. Eyelid edema
 3. Moderate to severe erythema of conjunctiva; chemosis (especially allergic)
 4. Preauricular adenopathy, commonly seen with viral pathogens
 5. Malaise, pharyngitis, and fever may be present
 6. Cobblestone-like papillary hypertrophy along inner aspect of upper lid (vernal conjunctivitis)
 7. Follicular changes in the palpebral fornix, especially with viral infections
- Diagnostic Tests/Findings
 1. Cultures and sensitivities usually unnecessary unless *Pseudomonas*, *Neisseria*, or other virulent organism suspected
 2. Rule out corneal involvement with fluorescein stain
- Management/Treatment
 1. Bacterial—most respond readily to topical ophthalmic antibiotics, e.g., tobramycin, erythromycin, sulfacetamide, polymyxin B sulfate-trimethoprim, or fluoroquinolone drops or ointment; if concomitant otitis media, also treat with oral antimicrobials
 2. Viral—artificial tears for lubrication, topical steroids (only by ophthalmologist) if keratitis present
 3. Herpes—initial infection self limited, virus remains latent; can recur in children with recurrent keratitis; ophthalmologist referral necessary; any child with a history of a herpes ocular infection who presents with red eye, lid vesicles, or dendritic findings on corneal staining with fluorescein should be referred immediately; treatment includes topical and systemic antivirals
 4. Allergic—avoidance of known allergens, cold compresses, systemic or topical antihistamines, ophthalmic nonsteroidal anti-inflammatories (e.g., Ketorolac/Acular) or topical mast cell stabilizers (e.g., cromolyn sodium); refer to ophthalmologist for chronic conjunctivitis
 5. Good hand washing and hygiene; stress control of cross contamination; avoid shared linen; cleanse eyelashes with warm sterile water, wiping from inner canthus outward
 6. Ophthalmologist referral needed:
 - a. If conjunctivitis is unresponsive to treatment within 2 to 3 days
 - b. If associated with loss of vision, pain, photophobia
 - c. If severe conjunctivitis or with corneal/orbital cellulitis involvement

Dacryostenosis

- Definition: Unilateral or bilateral obstruction of the nasolacrimal duct, usually at nasal punctal opening (“blocked tear ducts”)
- Etiology/Incidence
 1. Due to failure of duct canalization during gestation
 2. May also occur secondary to trauma or infection
 3. 90% spontaneous resolution by 12 months

4. Most common lacrimal disorder in infants; occurs in approximately 30% of newborns (Zitelli, 2007)
- Signs and Symptoms
 1. Onset typically 1 to 2 weeks of age (range newborn to one month)
 2. Continuous or intermittent tearing (wet look in the eye) with crusting of lashes
 3. Conjunctiva clear (no redness)
 - Differential Diagnosis
 1. Ophthalmia neonatorum, conjunctivitis
 2. Dacryocystitis—inflammation/infection of obstructed nasolacrimal duct
 3. Glaucoma
 4. Intraocular inflammation
 5. External irritation
 - Physical Findings
 1. Tearing
 2. Expression of thin mucopurulent exudate from lacrimal sac
 3. Signs of dacryocystitis—fever, erythema, and edema of lid and/or over lacrimal sac or duct
 - Management/Treatment
 1. Gently massage lacrimal sac and nasolacrimal duct by frequently stroking skin from brow area along lateral aspect of nose (no clear supporting data for effectiveness but often recommended)
 2. For purulent discharge, intermittent use of erythromycin ointment (when drainage becomes clear again, stop ointment)
 3. Referral to ophthalmologist by six months of age or earlier for evaluation and treatment if no improvement with antibiotics and massage
 4. Persistent obstruction and recurrent purulent drainage beyond six to twelve months; may require surgical probing of duct and/or silicone tube intubation
 5. Severe dacryocystitis requires referral to ophthalmologist and systemic antibiotics

Chalazion

- Definition: Chronic granulomatous inflammation of the meibomian glands, occurring on the conjunctival aspect (inner lining) on the mid-portion of the eyelid; results in a well-defined, nontender cyst
- Etiology/Incidence: Unknown cause but may be related to chronic inflammation, especially related to hordeolum

- Signs and Symptoms
 1. Progressive swelling of upper or lower eyelid over weeks
 2. Possible slight discomfort
 3. Possible minimal redness
- Differential Diagnosis
 1. Blepharitis
 2. Hordeolum
 3. Sebaceous cell carcinoma (rare)
- Physical Findings
 1. Firm, nontender, nonmovable nodule; often on palpebral surface of lid
 2. Large chalazions may cause chronic pressure on cornea leading to astigmatism
- Diagnostic Tests/Findings: None indicated
- Management/Treatment
 1. Small chalazions may resolve without treatment
 2. Warm compresses 2 to 3 times a day for 20 minutes for 2 to 3 days
 3. Treat large, recurrent, or infected lesions with local antibacterial drops or ointments (sulfacetamide sodium 10%), qid for one week; recurrent chalazions may benefit from systemic antibiotics
 4. If unresponsive to treatment, refer to ophthalmologist for surgical excision, curettage, or corticosteroid injections

Blepharitis (Granulation of Eyelids)

- Definition
 1. Common acute or chronic bilateral inflammation of the eyelid margins
 2. Types
 - a. Seborrheic
 - b. Staphylococcal—bacterial infection of eyelash follicles
 - c. Mixed—combination of both types
- Etiology/Incidence
 1. Seborrheic blepharitis may be associated with seborrheic dermatitis, psoriasis, eczema, or allergies; chemicals, smoke, air pollution, and cosmetics can aggravate condition
 2. Staphylococcus aureus is primary causative pathogen
- Signs and Symptoms
 1. Irritation, burning sensation to eyes
 2. Sensation of foreign body
 3. Erythema of eyelid margins
 4. Itching of eyelid margin
 5. Loss of eye lashes (especially with staph)

- Differential Diagnosis
 1. Chalazion, conjunctivitis, superficial keratitis
 2. Hordeolum
 3. Seborrhea, pediculosis of eyelashes
- Physical Findings
 1. Scaliness to eyelid margin (and scales can be seen on eyelashes)
 - a. Seborrheic—greasy scales
 - b. Staphylococcal—dry scales
 2. Redness to lid margin—more marked with staph, less red in seborrheic type
 3. Loss of eyelash—seen with staphylococcal type
 4. Ulcers—tiny ulcerated areas on lid margin with staph type; may lead to eyelid margin distortion and possible ectropion
 5. May be associated with conjunctivitis and superficial keratitis
- Diagnostic Tests/Findings: None indicated
- Management/Treatment: Similar for both forms
 1. Warm moist compresses to lid margins several times/day
 2. Daily mechanical scrubbing and cleansing of lid margins with cotton tipped applicator or soft cloth, dipped in dilute baby shampoo
 3. Application of topical antibiotic ointment massaged into lid margins (sulfacetamide sodium 10% or polymyxin B-bacitracin)
 4. When seborrheic dermatitis of the scalp is present, frequent shampooing with selenium sulfide recommended
 5. Continue treatment for several weeks if necessary; recurrences are common

Hordeolum (Stye)

- Definition: An acute localized inflammation of one or more sebaceous glands (meibomian or zeisian) of eyelids
- Etiology/Incidence: Most common infectious pathogen is *Staphylococcus aureus*; highest incidence in children/adolescents
- Signs and Symptoms
 1. Sudden onset of tenderness, redness, and swelling of affected lid; typically lid margin
 2. Foreign body sensation, occasional
- Differential Diagnosis
 1. Chalazion
 2. Blepharitis
 3. Inclusion cyst

- Physical Findings
 1. Painful erythematous swelling on either skin (external stye) or conjunctival (internal stye) surface
 2. May suppurate and drain spontaneously
- Diagnostic Tests/Findings: Culture not necessary
- Management/Treatment
 1. Warm moist compresses for 15 minutes three to four times a day
 2. Sulfacetamide sodium 10% or polymyxin B-bacitracin drops or ointment
 3. Cleanse eyelids with baby shampoo once a day
 4. Refer for incision and drainage if unresponsive to treatment
 5. Dispose of old eye makeup; discourage use of eye makeup until hordeolum is resolved; stress good hand and eye hygiene

Orbital/Periorbital Cellulitis

- Definition
 1. Orbital—inflammation of the orbital contents posterior to orbital septum
 2. Periorbital (preseptal)—inflammation/infection of the skin and subcutaneous tissue surrounding eye
- Etiology/Incidence
 1. Orbital cellulitis is more common in children than adults and the primary etiology is extension of a bacterial sinusitis (into the orbital tissues)
 2. Periorbital cellulitis is more common than orbital cellulitis; often associated with skin disorders or infections of the eyelid such as insect bites, impetigo, styes
 3. Most common organisms are *Staphylococcus aureus*, *Streptococcus pneumoniae*, *Haemophilus influenzae*, Group A strep, and anaerobic organisms
- Signs and Symptoms
 1. Orbital cellulitis
 - a. Insidious onset of unilateral, eyelid swelling
 - b. Orbital pain, headache
 - c. Decreased vision
 - d. Fever
 - e. Potentially devastating consequences
 2. Periorbital cellulitis
 - a. Acute onset of unilateral eyelid swelling
 - b. Warmth, swelling, and tenderness of overlying skin
 - c. Eye itself and vision are usually normal

- d. Systemic symptoms are often absent but occasionally with fever
- 3. Rhinorrhea, fever (102° to 104°F)
- Differential Diagnosis
 1. Differentiate between orbital (within true orbit), periorbital, or preseptal (surrounding orbital septum) cellulitis
 2. Edema secondary to trauma
 3. Allergic periorbital edema
 4. Insect bite
- Physical Findings
 1. Orbital cellulitis
 - a. Lid edema—not extending onto the eyebrow (as in periorbital cellulitis)
 - b. Chemosis
 - c. Proptosis
 - d. Decreased ocular mobility
 - e. Decreased visual acuity
 - f. Ophthalmoplegia (paralysis of eye muscles) and proptosis (protrusion of eyeball)—classic findings and distinguish orbital from periorbital cellulitis
 2. Periorbital cellulitis
 - a. Unilateral eyelid edema
 - b. Erythema
 - c. Tenderness of overlying skin
 - d. Visual acuity usually normal
- Diagnostic Tests/Findings
 1. Visual acuity exam—decreased vision with orbital cellulitis only
 2. CT scan of sinuses—determine sinus involvement
 3. Orbital sonography—determine orbit involvement
 4. CBC with differential—leukocytosis
 5. Blood and/or eye cultures—R/O concurrent sepsis and identify pathogen
 6. Lumbar punctures with infants
- Management/Treatment
 1. Prompt assessment, treatment, and referral
 2. Systemic antibiotic therapy, primarily IV
 3. Moderate to severe cases or children under 1 year old may need hospitalization
 4. Complications may include loss of vision, subperiosteal or orbital abscess, meningitis, epidural and subdural abscess, thrombosis in the retina or sinus
 5. *Streptococcus pneumoniae* and *Haemophilus influenzae* vaccines have decreased annual case rate of periorbital and orbital cellulitis (Prentiss, 2008)

Cataracts

- Definition: Partial or complete opacity of the lens
- Etiology/Incidence: May be congenital or acquired, unilateral or bilateral; can result in amblyopia, partial or complete blindness; some are not clinically significant
 1. Congenital—may result from congenital infections (e.g., rubella, toxoplasmosis, cytomegalovirus), genetic anomalies, prematurity and/or drug exposure, or metabolic abnormalities such as hypocalcemia; may be present at birth or develop in first few months of life
 2. Acquired cataracts due to:
 - a. Trauma to the eye; cataracts may be a manifestation of child abuse
 - b. Systemic disease—diabetes mellitus, Trisomy 21, hypoparathyroidism, galactosemia, atopic dermatitis, hypocalcemia, Marfan's syndrome, neurofibromatosis
 - c. Various toxins, drugs, radiation; long-term systemic corticosteroid or ocular steroid drops; require routine ophthalmology exams
 - d. Complications from other ocular abnormalities, e.g., glaucoma, uveitis, strabismus, pendibular nystagmus with severe amblyopia
 - e. Approximately 30% are hereditary, 30% syndrome/disease related and the remaining are the other etiologies noted or undetermined
- Signs and Symptoms: Severity of visual acuity deficits depends on location and degree of opacity; cataracts alone may produce no symptoms except possible decreased visual acuity; no associated pain but may experience light sensitivity (Schwartz, 2005); evaluate for underlying systemic disease or other eye disorders
- Differential Diagnosis
 1. Retinoblastoma
 2. Glaucoma
- Physical Findings
 1. Decreased visual acuity
 2. Strabismus—may be initial sign of cataract in child
 3. Absent red reflex (leukocoria), black dot(s) surrounded by red reflex, or white plaque-like opacities
 4. Signs of other systemic diseases and ocular abnormalities as noted in etiology

- Management/Treatment
 1. Prompt referral to ophthalmologist for diagnosis/treatment
 2. Surgical measures indicated if vision unable to develop due to extent of cataract; visual correction (e.g., eyeglasses/contact lenses)
 3. Determine etiology; any positive family history of congenital cataracts; treat any underlying disorder

Glaucoma

- Definition: Increased intraocular pressure from disruption of aqueous fluid circulation involving one or both eyes resulting in optic nerve damage with loss of visual acuity and eventual blindness if untreated
 1. Congenital—glaucoma that occurs in the first 3 years of life; 40% present at birth and 85% by one year; incidence 1 in 12,500 births
 2. Juvenile—glaucoma that begins between the ages of 3 and 30
- Etiology/Incidence
 1. Primary—the cause is an isolated anomaly of drainage apparatus; 50% of infantile glaucoma is primary; incidence 0.03%
 2. Secondary—other ocular or systemic abnormalities are associated such as trauma, intraocular hemorrhage, intraocular tumor, cataracts, corticosteroid use; syndromes such as Marfan's, neurofibromatosis, congenital rubella syndrome, Pierre Robins syndrome
- Signs and Symptoms
 1. Classic triad—photophobia, abnormal overflow of tears (epiphora), blepharospasm (eyelid spasm); triad occurs in about 30%
 2. Decreased vision (peripheral first) leading to tunnel vision
 3. Persistent, extreme pain (occasionally)
- Differential Diagnosis: Cataracts
- Physical Findings
 1. Corneal and ocular enlargement; corneal diameter > 12 mm if < one year of age needs immediate attention; eventually entire eye enlarges
 2. Other findings may include corneal haziness and edema; conjunctival injection
 3. Deep cupping of optic disc; enlargement of the optic cup and increase of cup; disc ratio 0.5
 4. Increase in intraocular pressure (IOP)—can be measured or suspected with firmness to palpation of the eye

- Diagnostic Tests/Findings: Glaucoma pressure test will show increased pressure
- Management/Treatment
 1. Immediate referral to ophthalmologist to confirm diagnosis and initiate therapy
 2. Surgery is often first line, followed by medical therapy; post-op steroids and cycloplegic drops are essential to prevent adhesions
 3. Same treatments as with adults: topical beta blockers, adrenergic agents, and carbonic anhydrase inhibitors; miotics not used
 4. Even with treatment, still has high risk for visual impairment; refer early for services for visually impaired

Strabismus

- Definition: Ocular misalignment; eyes may deviate outward (exotropia), inward (esotropia), downward (hypotropia), or upward (hypertropia); causes the eyes to not simultaneously view the same object (causes a diplopia); response to diplopia is visual axis suppression (to eliminate diplopia)
- Etiology/Incidence
 1. Affects approximately 4% of children < 6 years of age; 30%–50% develop amblyopia with some visual loss; needs early detection for normal visual outcome
 2. Constant strabismus is termed “tropia;” intermittent is termed “phoria;” esotropia is most common
 3. Normal variant in first four to six months of life
 4. Historical risk factors include prematurity, family history, cerebral palsy, most chromosomal and other major genetic anomalies, prenatal drug exposure, fetal alcohol syndrome, major head trauma, major congenital or acquired structural ocular defects
 5. Acquired strabismus occurring after 6 months of age usually from cataracts, retinoblastoma, anisometropia, or high refractive errors
 6. Paralytic strabismus—often due to a tumor
 7. Patients with congenital vision loss are more likely to develop exotropia and those with acquired vision loss are more likely to develop exotropia
 8. Pseudostrabismus—eyes appear to be crossed due to epicanthal folds on either side of bridge of nose; no ocular deviation
- Signs and Symptoms
 1. Varies with age
 2. Squinting

3. Decreased vision
 4. School problems
 5. Head tilting
 6. Face turning
 7. Over pointing
 8. Awkwardness
- Differential Diagnosis
 1. Cataracts
 2. Retinoblastoma
 3. Anisometropia
 4. High refractive errors
 5. Amblyopia
 6. Head trauma
 7. Other congenital eye muscle syndromes
 - Physical Findings
 1. Misalignment of eye(s)
 2. Intermittent, alternating, or continuous esotropia, exotropia, hypertropia, or hypotropia
 - Diagnostic Tests/Findings
 1. Abnormal Hirschberg (corneal light reflex unequal) and/or cover/uncover test (abnormal movement of eye)
 2. Vision screen—decreased visual acuity, may reveal refractive errors, amblyopia, or anisometropia
 - Management/Treatment
 1. Establish period of onset, progression frequency, and amplitude (constant versus intermittent), presence or absence of fixation preference, family history of strabismus or amblyopia, evidence of any associated neurologic or other disorders (Ticho, 2003)
 2. Referral to ophthalmologist
 - a. Any children over 6 months with fixed or continuous strabismus; constant exotropia under 3 months of age
 - b. Earlier if other developmental delays are present or deviations are continuous or fixed
 - c. Signs of differential diagnosis present
 - d. Suspicious history
 - e. Immediate if hypotropia or hypertropia is present at any age
 3. Acquired—appropriate treatment depending on pathology and/or refractive errors
 4. Treatment options to be determined by ophthalmology; patching of nonaffected eye (forces use of deviating eye); orthoptic exercises, corrective lenses, surgical alignment, and/or a combination

Nystagmus

- Definition: Involuntary, horizontal, vertical, rotary, or mixed rhythmic movement of eyes
- Etiology/Incidence
 1. Caused by abnormality in one of three basic mechanisms—fixation, conjugate gaze, or vestibular mechanisms
 2. May be familial
 3. Associated with albinism, refractive errors, congenital cataracts, central nervous system (CNS) abnormalities, and various diseases of inner ear and retina
 4. Classified according to direction of movement
- Signs and Symptoms
 1. Irregular eye movements
 2. Abnormal head movements; may be rhythmic
- Differential Diagnosis: Underlying cause
- Physical Findings
 1. Involuntary rhythmic eye movements
 2. Vision screen may be abnormal
- Diagnostic Tests/Findings: Abnormal visual acuity test
- Management/Treatment
 1. Refer to ophthalmologist and neurologist (as necessary)
 2. Treat underlying cause
 3. Monitor child closely

Retinoblastoma

- Definition: Congenital malignancy of retina
 1. Most common malignant intraocular tumor of childhood
 2. See discussion in Chapter 13, Hematologic/Oncologic/Immunologic Disorders

Refractive Errors

Hyperopia (Farsightedness)

- Definition: Alteration in refractive power when visual image is focused behind the retina; ability to see objects clearly at a distance, but not at close range
- Etiology/Incidence
 1. Axial length of eye too short and/or due to insufficient convexity of the refracting surfaces of the eye especially the cornea
 2. Familial pattern common

- Signs and Symptoms
 1. Often asymptomatic due to children's ability to easily accommodate for hyperopia
 2. Headache, eye strain, squinting, and eye rubbing during prolonged periods of close work in older children
 3. Strabismus often seen with severe hyperopia
- Differential Diagnosis
 1. Astigmatism
 2. Myopia
 3. Anisometropia
 4. Amblyopia
- Physical Findings: Abnormal vision screen
- Diagnostic Tests/Findings
 1. Vision screen shows hyperopia
 2. Passing vision screen is 20/40 (age 3 to 4 years), 20/30 (older children)
 3. A difference of two lines between the 2 eyes is significant
- Management/Treatment
 1. Refer to ophthalmologist or optometrist
 2. If strabismus present, see previous section for management
 3. Annual eye examinations

Myopia (Nearsightedness)

- Definition: Alteration in refractive power when visual image is focused in front of retina; ability to see objects clearly at close range, but not at a distance
- Etiology/Incidence
 1. Axial length of eye too long and/or increased curvature of the refracting surfaces of the eye, especially the cornea
 2. Familial pattern common; frequently associated with prematurity
 3. Myopia may be present at birth; usually appears around 8 to 10 years of age
- Signs and Symptoms
 1. Distant objects blurred
 2. Squinting forms physiologic pinhole to improve acuity
 3. Difficulty reading blackboard
- Differential Diagnosis
 1. Hyperopia
 2. Astigmatism
 3. Anisometropia
 4. Amblyopia

- Physical Findings: Abnormal vision screen
- Diagnostic Tests/Findings: Abnormal vision screen (see hyperopia)
- Management/Treatment
 1. Refer to ophthalmologist or optometrist
 2. Importance of annual eye examinations

Astigmatism

- Definition: Refractive error due to an irregular curvature of the cornea or changes in the lens causing light rays to bend in different directions
- Etiology/Incidence
 1. Familial developmental variations in the curvature of the cornea
 2. Depending on severity of curvature variation, retina cannot focus regardless of distance
 3. Increased risk secondary to injury, periorbital or eyelid hemangiomas, or ptosis (Behrman, 2007)
- Signs and Symptoms
 1. Commonly found in conjunction with other refractive errors
 2. Pain in and around eyes, headache, fatigue, reading problems, frowning
- Differential Diagnosis
 1. Myopia
 2. Hyperopia
 3. Amblyopia
 4. Anisometropia
- Physical Findings: Possible abnormal vision screen; asymmetrical Hirschberg
- Diagnostic Tests/Findings: Vision screen with possible abnormality
- Management/Treatment
 1. Refer to ophthalmologist or optometrist
 2. Corrective lenses or contacts may be prescribed; patching may be recommended

Blindness (Amaurosis)

- Definition
 1. Varies from inability to distinguish light from darkness to partial vision
 2. Best corrected visual acuity between 20/70 to 20/200
 3. Legal blindness is distant acuity 20/200 in better eye or visual field; includes an angle of no greater than 20 degrees
 4. Primary blindness—present at birth

- Etiology/Incidence
 1. $\frac{1}{3}$ of blindness in children from trauma
 2. Variety of pathological causes—cataracts, glaucoma, retinopathy of prematurity (ROP) (most common cause of severe visual impairment); retinoblastoma, trauma, detached retina, cranial nerve II problems, infection, hydrocephalus, genetic problems
- Signs and Symptoms: Varies with age and mode of onset, abilities of child, laterality, and severity of deficit
 1. Developmental delays, e.g., gross motor, walking often delayed to 18 to 24 months
 2. Social skills—increased passivity, increased anxiety around strangers
 3. Decreased social communication and school performance
 4. Increased self-stimulating behavior, e.g., hand flapping, rocking, rubbing eyes
 5. Language delay; cognitive delays, understanding of object permanence, cause/effect
 6. Delay in development of conversational skills
 7. Photophobia, chronic tearing, wandering eyes
 8. Lack of smiling response to visual stimuli
- Differential Diagnosis: If primary or from birth
 1. Developmental malformations
 2. Damage consequent to gestational/perinatal infection
 3. Anoxia, hypoxia, perinatal trauma
 4. Genetically determined diseases
- Physical Findings: In primary blindness
 1. Nystagmus (may be first clue)
 2. Enlarged or cloudy cornea; abnormal or absent red reflex
 3. Lack of pupillary reflex; optic disc pallor; pigmentary deposits
 4. Fixed or intermittent strabismus beyond 6 months of age
 5. History of retinopathy of prematurity (ROP)
 6. Possible neurological disorder
- Diagnostic Tests/Findings
 1. Ophthalmologic examination—abnormal as described above
 2. Developmental testing—delays as outlined
 3. CT or MRI—rule out physiological abnormalities
- Management/Treatment
 1. Obtain complete family genetic visual impairment history
 2. Address family issues, social and emotional needs of child

3. General medical and developmental history
4. Metabolic and genetic studies
5. Prompt referral to ophthalmologist, neurologist, and developmental specialist

Amblyopia (Lazy Eye)

- Definition
 1. Decreased visual acuity in one or both eyes that cannot be attributed to any structural abnormality and cannot be immediately corrected with any glasses
 2. Occurs in visually immature children, during the “sensitive” period for visual development; amblyopia is caused by a lack of a clear image onto the retina of the immature visual system, which results in suppression of vision in that eye; vision can likely be restored if identified and corrected early; important to identify anything that would cause obstruction of a clear image onto the retina (e.g., strabismus, tumor, and cataract)
- Etiology/Incidence
 1. Organic—trauma, organic lesion, cataract, diseases of the eye or visual pathways, ptosis
 2. Nonorganic—sensory stimulation deprivation or disuse; abnormal binocular interaction during infancy and early childhood (greatest risk between 2 to 3 years of age but can continue until 9 years of age); large difference in refractive errors between both eyes (anisometropia)
 3. Rarely bilateral; associated with strabismus
 4. Occurs in 2% to 2.5 % of general population (nearly 6 million Americans)
 5. Five types—deprivation (ptosis, opacities), strabismic, anisometric, occlusion (patching good eye too much), ametropic (both eyes large refractive errors, typically hyperopic and/or astigmatism)
- Signs and Symptoms: Wandering eye
- Differential Diagnosis
 1. Cataracts
 2. Blindness
 3. Ptosis
- Physical Examination
 1. Decreased visual acuity in one or both eyes
 2. Specific findings with underlying causes; abnormal red reflex with tumor, cataracts; presence of ptosis and strabismus are also causes

- Diagnostics Tests/Findings
 1. Abnormal vision and/or ophthalmic examination—abnormal red reflex, positive cover/uncover test, unequal corneal light reflex
 2. Abnormal ocular movements
- Management/Treatment
 1. Early detection, prompt intervention, referral to ophthalmologist
 2. Effective vision screening before 3 years of age
 3. Corrective lenses
 4. Therapy forcing stimulation of amblyopic eye; patching or use of atropine in good eye
 5. Close monitoring of therapy
 6. Reassurance and support throughout treatment

Eye Injuries

General information regarding corneal abrasion, foreign body, hyphema, ecchymosis, and chemical injuries

- Definition/Incidence/Etiology
 1. $\frac{1}{3}$ of blindness in children < ten years of age is due to trauma
 2. Sports and activities in which ocular trauma is common include BB guns (most common), archery, darts, motorcycling, bicycling, racquet sports, boxing, basketball, and baseball
- Physical Examination
 1. Steps in proper evaluation include:
 - a. Recognizing life threatening nonocular conditions, e.g., chemical injuries
 - b. Taking adequate history to assess potential risk of injury
 - c. Examining in detail (visual acuity, external ocular motility unless ruptured globe suspected), pupil, anterior segment, and fundus
 - d. Referral for signs and symptoms or history of severe ocular injury
 2. Use caution—severe intraocular injury may be concealed behind minimal external trauma
 3. Examine lids, lacrimal system, adnexa, sclera, and conjunctiva for lacerations, foreign body, or perforation
 4. Palpate orbital rim for crepitus and obtain a CT scan when orbital fracture suspected; can be associated with significant intracranial and ocular injuries
 5. A dislocated lens presents with a quivering iris
 6. Topical anesthetic recommended for examination only; slows healing of cornea

- Management/Treatment
 1. Ocular injuries requiring immediate referral include:
 - a. Chemical injuries
 - b. Globe lacerations, severe lacerations of lid
 - c. Hyphemas
 - d. Penetrating intraocular injury
 2. Avoid pressure to globe by placing a protective shield over injured eye
 3. Nonaccidental trauma or child abuse should be considered with presence of lid ecchymosis, conjunctival hemorrhages, hyphema, or retinal hemorrhages; ideally injuries should be photographed when possible
 4. Acetaminophen for pain, no acetylsalicylic acid (ASA)
 5. Injury prevention (e.g., protective eyewear) will reduce ocular trauma by 90%
 6. Date of last tetanus vaccine
 7. Rabies prophylaxis if trauma from animal bite
 8. Refer to ophthalmologist for further assessment

Corneal Abrasion

- Definition: Loss of epithelial lining from corneal surface of one or both eyes
- Etiology/Incidence: abrasion, trauma or foreign bodies in upper lid leading to corneal injury, e.g., paper, brushes, toys or fingernails, contact lenses, ultraviolet light exposure; most common ocular injury in children
- Signs and Symptoms
 1. Possible evidence of foreign body
 2. Severe pain and tearing
 3. Blepharospasm
 4. Injection of sclera
 5. Photophobia
- Physical Findings: Epithelial injury visible with use of fluorescein stain and cobalt blue light (e.g., Wood's lamp)
- Management/Treatment
 1. Topical anesthetic for evaluation only
 2. Remove foreign body via irrigation with normal saline
 3. Broad spectrum antibiotic ointment such as erythromycin and bacitracin; and a topical cycloplegic agent such as cyclopentolate to help alleviate ciliary spasm 2 to 3 times/day
 4. Non-steroidal anti-inflammatory (NSAID) medications can be used for pain control
 5. The trend is away from patching, but if applied make certain the eye is closed and patch is

applied properly; do NOT patch if possible infection or foreign body exists

6. Most abrasions heal within 24 to 48 hours; follow up in 24 hours and restrain to evaluate abrasion and refer to ophthalmologist if abrasion is not healed within 3 days

Foreign Body Eye Injury

- Definition: Foreign body in the eye
- Etiology/Incidence
 1. Surface—nonadherent/loosely adherent to cornea or conjunctival epithelium (most likely sources dirt, sand, grass); organic material has a higher rate of infection and metal can cause a reaction in the vitreous
 2. Penetrating—into but not through cornea or sclera
 3. Perforating—through cornea or sclera and into globe
- Signs and Symptoms
 1. Foreign body sensation
 2. Pain
 3. Sensitivity to light
 4. Tearing
 5. Eye rubbing
- Differential Diagnosis
 1. Corneal abrasion
 2. Chemical injury
- Physical Findings
 1. Foreign body may be visualized
 2. Positive fluorescein examination if corneal abrasion present
 3. Epiphora
 4. Possible visual acuity abnormality
- Diagnostic Tests/Findings
 1. Visual acuity—to determine any deviation from normal
 2. Fluorescein test—to determine presence of corneal abrasion
 3. Possible radiographs to visualize foreign body
- Management/Treatment
 1. All intraocular foreign bodies require referral to ophthalmologist
 2. Topical ophthalmic anesthetic drops for examination unless perforating wound suspected
 3. If persistent corneal abrasion after 24 hours with treatment, penetrating or perforation wound, refer to ophthalmologist
 4. Thoroughly examine lashes, lids, cornea, and conjunctival surfaces

5. Remove foreign body via irrigation with normal saline or a moistened cotton-tipped applicator
6. After removal, examine for corneal abrasion, treat appropriately

Hyphema

- Definition: Accumulation of blood in anterior chamber
- Etiology/Incidence
 1. Due to blunt or perforating trauma (e.g., balls, fists, and sticks) leading to rupture of iris or ciliary body blood vessels; increased risk of rebleeding in first 5 days and glaucoma with traumatic hyphema
 2. May also occur with bleeding disorders, e.g., sickle cell disease, hemophilia
 3. Usually lasts 5 to 6 days
- Signs and Symptoms
 1. History of ocular injury
 2. Drowsiness (but also consider head injury)
 3. Pain
 4. Light sensitivity
 5. Blurring of vision
- Physical Findings
 1. Blood in anterior chamber can be seen on gross inspection with patient sitting up
 2. Visual acuity changes
- Management/Treatment
 1. Refer to ophthalmologist
 2. Reduce activity for several days, bed rest in supine position with head of bed elevated 30 to 40 degrees to promote reabsorption of blood; hospitalization often necessary; no reading or activities
 3. Eye patch with a metal shield for 5 days on injured eye (to protect from reinjury); patch must have holes or clear plastic so patients can assess their vision because worsening of vision first sign of rebleed
 4. Medications: topical cyclopegics to decrease pain, aminocaproic acid (ACA) (topical or oral) for bleeding control, no aspirin (Boar, 2008)
 5. Surgery may be necessary to remove blood
 6. Close follow-up for complications: rebleed is most common complication, usually 2 to 5 days after the injury (> 50% chance in patients with sickle cell trait or anemia) (Boar, 2008); glaucoma, cataracts, and sympathetic ophthalmia (inflammation that occurs in the uninjured eye weeks to years later) are other complications (Boar, 2008)

Ecchymoses (“Black Eye”)

- Definition: Bruising of periorbital region
- Etiology/Incidence: Blunt contusion injury; due to isolated periorbital injury or other orbital/ocular injury
- Signs and Symptoms
 1. Pain
 2. Visual impairment due to occlusion from edematous eyelid
- Differential Diagnosis: Other orbital/ocular injury (e.g., lens dislocation, globe rupture, retinal detachment)
- Physical Findings
 1. Edema
 2. Ecchymosis
- Diagnostic Tests/Findings
 1. Ophthalmic examination—determine other orbital/ocular injuries
 2. Visual acuity
- Management/Treatment
 1. Uncomplicated—cold compresses for 24 to 48 hours, then warm compresses until swelling resolves; elevate head; inform parents/patient that ecchymosis and edema may spread
 2. Refer to ophthalmologist—closed head injury, damage to skull, facial bone fracture

Chemical Injuries

- Definition: Burns of the eyelids, conjunctiva, and/or cornea
- Etiology/Incidence
 1. Steam, intense heat, and common household agents; deployment of air bags can release chemicals potentially causing alkaline chemical damage
 2. Severe alkali injuries characterized by corneal opacification
 3. Amount of damage directly related to duration of exposure
- Management/Treatment
 1. Acute ocular emergency
 2. Copious irrigation with normal saline for 20 to 30 minutes—patch and refer to ophthalmologist immediately to rule out corneal or other ocular trauma
 3. Severe chemical injury to eye(s) requires hospitalization
 4. Topical anesthetic may reduce pain from injury and irrigation

5. Pseudomonal contamination common with any burn, possibly leading to corneal ulceration; prevention—antibiotic preparation containing polymyxin B, gentamicin, tobramycin, or colistin should be instilled into the injured eye(s) 3 to 4 times a day

■ EAR**External Otitis Media (EOM)**

- Definition: Acute infection and/or inflammation of external auditory canal; “swimmer’s ear”
- Etiology/Incidence
 1. More common in summer months due to excessive wetness (swimming, bathing, or increased environmental humidity) which changes the acidic environment and promotes bacterial/fungal growth
 2. Common organisms are *Pseudomonas aeruginosa* (most common), *Staphylococcus aureus*, *Streptococcus pyogenes*, *Enterobacter aerogenes*, *Proteus mirabilis*, *Klebsiella pneumoniae*, *Staphylococcus epidermidis*, and fungi, e.g., candida, aspergillus, trichophyton
 3. Loss of protective cerumen or chronic irritation
 4. Trauma disrupting lining of auditory canal, e.g., foreign body, digital irritation, cotton-tipped swabs, hearing aids
 5. Excessive dryness (eczema, psoriasis); contact dermatitis, e.g., poison ivy, medications, hair spray, perfumes
- Signs and Symptoms
 1. Itching of ear canal (early symptom)
 2. Acute and possibly severe ear pain upon manipulation of pinna/tragus or performance of otoscopic examination
 3. Pressure/fullness in ear, possible hearing loss
- Differential Diagnosis
 1. Acute otitis media with perforation
 2. Dental infection
 3. Mastoiditis, furunculosis
 4. Foreign body
 5. Eczema
 6. Parotitis
- Physical Findings
 1. Tympanic membrane (TM) is normal; may be difficult to visualize due to swelling and exudate in the canal
 2. Edematous/erythematous external canal; with or without exudate

3. Fluffy white debris and occasionally with black spots on TM indicative of fungal infection
 4. Furuncle may be noted with localized infection
 5. Possible pre or postauricular lymphadenopathy
 6. Observe for signs of mastoiditis or cellulitis beyond external canal
- Diagnostic Tests/Findings: Culture of discharge unnecessary unless unresponsive to treatment
 - Management/Treatment
 1. Withdraw any foreign bodies or debris from external canal by gentle irrigation with warm water or normal saline; do not irrigate if perforated TM is a consideration
 2. Topical antibiotic otic drops are sufficient: those containing combinations of neomycin, polymyxin, or fluoroquinolone are effective (ofloxacin is safe with PE tubes or TM perforation) (Stone, 2007); the addition of hydrocortisone (e.g., Cipro HC) is helpful when canal is edematous
 3. If significant swelling, insert cotton wick saturated with antibiotic solution for first 24 to 48 hours
 4. Systemic analgesic often required for severe pain (e.g., NSAIDs, acetaminophen with codeine)
 5. Avoid swimming and getting ear wet during the acute phase
 6. Reexamine 1 to 2 weeks for evaluation of TM and removal of any debris
 7. Prevention—instillation of white vinegar and rubbing alcohol (50/50) in both ear canals after swimming; avoid water in canals, vigorous cleaning, scratching, or prolonged use of cerumenolytic agents
 8. Systemic antibiotics when OE accompanied by fever, lymphadenitis, facial cellulitis (uncommon)
 9. Incision and drainage (I + D) of furuncle if present
 10. Fungal infection treated with 5% boric acid in ethanol solution for 5 to 7 days
- Etiology/Incidence
 1. Fluid/pathogen accumulation in middle ear due to Eustachian tube (ET) dysfunction
 - a. ET allows for proper ventilation and drainage of middle ear space; without this, an effusion develops in the middle ear space with subsequent bacterial contamination
 - b. ET dysfunction can be due to age-related characteristics of the ET (e.g., floppy ET) or obstruction of the ET (e.g., inflammation from allergies, viral infections, or enlarged adenoids) and/or other mechanical factors
 - c. Most ET dysfunction corrects with age; temporary ventilation tubes (PE tubes) provide ventilation of the middle ear until child is older
 2. Common pathogenic agents—varies according to geographic location; has recently shifted with the use of Hib and pneumococcal conjugate vaccines
 - a. *Streptococcus pneumoniae* (approximately 40%)
 - b. *Haemophilus influenzae* (25% to 30%)
 - c. *Moraxella catarrhalis* (10% to 20%)
 - d. Less common pathogens—*Staphylococcus aureus*, group A beta hemolytic streptococcus, and *Pseudomonas aeruginosa* (more common in chronic otitis media)
 - e. Viruses—Influenza, RSV (most common), adenovirus, parainfluenza, and coronavirus
 - f. Prevalence of β -lactamase producing strains highest in past 15 years
 - g. Increase in drug resistant bacteria, especially in children younger than 24 months; those who recently were treated with β -lactamase antibiotics and children exposed to large numbers of children
 - h. No growth found in approximately 16% of AOM
 3. Predisposing factors
 - a. Physiological considerations—Eustachian tube of child under 6 years of age is short, wide, and horizontal, allowing access of pathogens from nasopharynx to reach middle ear
 - b. Possible hereditary factors
 - c. Common occurrence with/following upper respiratory infection
 - d. Bottle-feeding in supine position and/or no breastfeeding
 - e. Craniofacial abnormalities, e.g., cleft palate and Down syndrome
 - f. Allergic rhinitis

Acute Otitis Media (AOM)

- Definition: Inflammation of the middle ear with fluid in the middle ear space (suppurative otitis media); the 2004 American Academy of Pediatrics specify 3 criteria that must be present: (1) acute onset of signs/symptoms, (2) evidence of middle ear effusion, and (3) evidence of middle ear inflammation (redness of TM)

- g. Tobacco smoke exposure
- h. Children in group daycare centers at higher risk than those in home care
- 4. One of the most common pediatric diagnoses—highest prevalence between 6 to 36 months
- 5. Highest incidence in winter/spring; males, Caucasians, American Indians, Eskimos, and lower socioeconomic groups
- 6. Natural history of untreated otitis media—70% to 90% spontaneous resolution
- Signs and Symptoms
 1. Fever in 30% to 50% of cases
 2. Complaints of ear fullness, pain, or discomfort 50% of the time
 3. Poor appetite/feeding, irritable with sleep disturbances (especially in infants)
 4. Nausea/vomiting
 5. Rhinorrhea and/or nasal congestion
 6. Tugging on ears
 7. Purulent otorrhea with TM rupture
- Differential Diagnosis
 1. Crying child with normal erythematous tympanic membranes
 2. Serous otitis media
 3. Tympanosclerosis
 4. Cholesteatoma
 5. Otitis externa
 6. Rare complications—mastoiditis, sepsis in young infant, hearing loss, developmental/speech delay
- Physical Findings: Diagnosis is determined by changes in color, contour, and mobility of TM
 1. Color is erythematous or yellow and opaque with dull appearance
 2. Contour may be bulging; light reflexes and bony landmarks usually distorted
 3. Mobility decreased or absent via tympanometry or pneumatic otoscopy
 4. Conductive hearing loss (to varying degrees; may not be evident to parent)
- Diagnostic Tests/Findings: Use depends on age of child and stage of middle ear disease
 1. Pneumatic otoscopy—visualize degree of mobility impairment
 2. Tympanometry using electroacoustic impedance bridge to measure compliance of the TM—identifies middle ear effusion, perforation, patent ventilation tubes, or excessive hard packed cerumen
 3. Hearing test—to determine if any hearing loss
- 4. Language screen—assess for language delay
- 5. Consider allergy evaluation and possibly immunologic evaluation for children with recurrent OM and other supporting symptoms (e.g., allergy symptoms)
- Management/Treatment
 1. Judicious use of antimicrobials due to increased bacterial resistance; consider no antibiotic use during the first 24 to 48 hours; if no better, an antibiotic should be given
 2. Select appropriate antibiotic—see **Table 4-1**
 3. Maintain realistic expectations—90% to 95% symptom relief within 48 to 72 hours; if no better, change antibiotic to 2nd line therapy
 4. Length of treatment is controversial, typically 10 days, however in older children (> 2 years) and with milder cases, may consider shorter courses
 5. Use tympanocentesis sparingly, only used for retreatment failures with severe symptoms
 6. Monitor residual OME—may last for several months; persistent OME for 6 to 8 weeks, consider treatment with second line antibiotics (see Table 4-1)
 7. Pain and fever control
 - a. Analgesics—acetaminophen, ibuprofen
 - b. Local anesthetic otic drops (contraindicated in acute/chronic perforations and ventilation tubes)
 8. Decongestant use not recommended
 9. Recheck AOM after 10 to 21 days
 10. Prevention
 - a. Proper feeding techniques for infants
 - b. Encourage breast feeding—possible protective effect
 - c. Eliminate exposure to tobacco smoke
 - d. Antibiotic prophylaxis—see Table 4-1; only used for control of chronic AOM, e.g., 3 or more episodes in 6 months or 4 episodes in 12 months
 - e. Immunization with pneumococcal and influenza vaccines
 11. Referral to otolaryngologist
 - a. Persistent AOM resistant to treatment over 1 to 2 months
 - b. Frequent recurrent OM—3 in 6 months, 4 to 5 episodes in one year, 6 episodes by 6 years of age
 - c. Persistent/chronic OME > 3 months
 - d. Evidence of hearing deficit and/or language delay
 - e. Determine need for tympanostomy tube placement

■ **Table 4-1** Antibiotic Therapy for Management of Otitis Media

Drug Category
First line therapy -
Amoxicillin—80–90 mg/kg/day (maximum 4 g/day) for 10 days (shorter courses possibly with older children with milder cases)
If rash (not anaphylaxis), may use cefuroxime, cefpodoxime, and cefdinir
Second line therapy -
Augmentin—90 mg/kg/day of amoxicillin and 6.4 mg/kg of clavulanate
If no improvement in 48 to 72 hours or recurrence of AOM within one month
Macrolide antibiotics such as azithromycin and clarithromycin are not recommended second line (may be used for penicillin anaphylaxis)
Third line therapy -
Ceftriaxone—50 mg/kg 2 doses IM, 48 hours apart with an option for a third dose and a tympanocentesis to determine pathogen (Hay, 2009)
Prophylactic therapy
1. Amoxicillin—first line; drug most studies
2. Sulfisoxazole
3. Selected cases only because of bacterial resistance; low dose daily
Pain Management:
topical analgesics, oral acetaminophen/NSAIDs
Observation without use of antibiotics is an option in select children depending on age, severity, ability to follow-up

Otitis Media with Effusion (OME)

- Definition: Inflammation/fluid accumulation in middle ear (serous, not purulent fluid) with decreased TM mobility on pneumatic otoscopy but without signs and symptoms of ear infection; also referred to as serous, secretory, mucoid, and allergic otitis media or “glue ear”
- Etiology/Incidence
 1. Caused by Eustachian tube dysfunction (negative pressure in the middle ear produces an effusion in the middle ear); also occurs as a frequent sequelae of acute otitis media
 2. OME accounts for 25% to 35% of all cases of OM
 3. 30% to 40% have OME associated with allergic rhinitis
 4. Majority (50% to 80%) clear spontaneously within 2 to 3 months

5. Mild to moderate hearing loss with concomitant language delays may occur with OME contributing to subsequent behavioral difficulties including poor attention and school performance

- Signs and Symptoms
 1. Sometimes none or mild discomfort, crackling or full sensation in ear
 2. Behavioral changes, e.g., hearing loss, decreased attention span
- Differential Diagnosis: Same as AOM
- Physical Findings
 1. Color—yellow, dull, opaque, or translucent TM; possible presence of fluid level or air bubbles
 2. Contour—appears retracted due to negative pressure in middle ear
 3. Vascularity—none visible
 4. Mobility—decreased; tympanometry reveals high negative pressure or flat line
 5. Assess for complications more commonly seen with OME; cholesteatoma and persistent TM perforation (Hay, 2009)
- Diagnostic Tests/Findings: Same as AOM
- Management/Treatment
 1. Most cases resolve without antibiotics
 2. Limit use of antibiotic prophylaxis due to marginal benefit
 3. Limit passive smoking exposure, treat other infections, control allergies
 4. Audiogram after 3 months; referral to otolaryngologist
 5. Consider surgery (PE tubes, adenoidectomy if indicated) for chronic OME accompanied by pain, recurrent AOM, speech or hearing problems (Paradise, 2005)
 6. Decongestants and antihistamines not recommended except if allergy symptoms present
 7. Follow up every 3 to 4 weeks
 8. Prevention, education, and referral recommendations, same as AOM

Tympanostomy Tubes

- Definition
 1. Surgical incision of eardrum (myringotomy with placement of ventilation tube) to relieve pressure and drain pus/fluid from middle ear
 2. Two types
 - a. Short term—intended to remain in TM 8 to 15 months

- b. Long term—intended for more than 15 months
- Etiology/Incidence
 1. One million tubes inserted annually
 2. Indicated for children with recurrent otitis media, persistent OM with effusion, failed antibiotic prophylaxis of 4 months or longer, those allergic to penicillin or sulfonamides, those with associated hearing loss of 20 dB, or other complications
 3. With increasing number of resistant bacteria, possibility of more frequent use of tubes
- Management/Treatment
 1. Fitted earplugs with swimming
 2. If drainage occurs from tubes, treat with antibiotic otic suspension, e.g., fluoroquinolone otic drops (first line therapy), polymyxin B with neomycin and hydrocortisone

Chronic/Acute Perforations

- Definition: Spontaneous perforation of tympanic membrane during episode of AOM
- Etiology/Incidence
 1. Chronic—perforation lasts longer than one month
 2. Chronic suppurative OM—associated with discharge
 3. Chronic sites of perforation
 - a. Central—relatively safe from cholesteatoma formation
 - b. Peripheral—especially in pars flaccida, increased risk of cholesteatoma
- Signs and Symptoms: Painless ear discharge if infection present
- Differential Diagnosis
 1. Cholesteatoma
 2. Mastoiditis
 3. Labyrinthitis
 4. Lateral sinus thrombophlebitis
- Physical Findings
 1. Thickened, inflamed middle ear mucosa; with or without discharge
 2. May contain granulation tissue or polyps
 3. Conductive hearing loss dependent on size of perforation
 4. Site of perforation important to note
- Diagnostic Tests/Findings
 1. Culture discharge—*Pseudomonas aeruginosa* and *Staphylococcus aureus* most often seen
 2. CT scan of mastoid—rule out mastoiditis

- Management/Treatment
 1. Oral antibiotics (see Table 4-1) for 14 days plus antibiotic eardrops (3 to 4 drops four times a day for 7 days); if not responsive, suspect mastoiditis or cholesteatoma
 2. Patients with central nervous system sequelae, refer immediately
 3. Hospitalization may be necessary if complications or underlying disorder
 4. Follow up every week to 3 months; most heal within 2 weeks
 5. Refer to otolaryngologist
 - a. Unresolved perforation after 1-year duration
 - b. Surgical repair delayed until 9 to 12 years of age
- Prevention of Recurrent Infection
 1. Cotton plugs with petroleum jelly (on outer surface) when bathing and hair washing
 2. Discourage swimming (use fitted earplugs if unavoidable)
 3. Diving, jumping into water, and underwater swimming forbidden
 4. Exposure to air is helpful

Mastoiditis

- Definition: Infection of the mastoid bone, more specifically the periosteum of the mastoid; can lead to destruction of the mastoid air system and abscess formation; considered an extension of a middle ear infection; a suppurative complication of otitis media
- Etiology/Incidence
 1. Uncommon, due to successful antibiotics for AOM; incidence 0.2% to 0.4%
 2. Develops secondary to OM leading to periostitis and osteitis with abscess formation
 3. About 60% are < 2 years of age, between 6 to 24 months (uncommon younger) (Schwartz, 2005)
 4. Common pathogens
 - a. Most common—*Streptococcus pneumoniae*, *Streptococcus pyogenes*, *Staphylococcus aureus*
 - b. Less common—*Haemophilus influenzae*
 - c. Other agents—*Pseudomonas* (chronic mastoiditis), *Mycobacterium tuberculosis* (rare), *Moraxella catarrhalis*, enteropathic gram negative rods
- Signs and Symptoms
 1. Pain, tenderness behind ear
 2. Fever or irritability
 3. Otorrhea

- Differential Diagnosis
 1. Meningitis
 2. Extradural abscess
 3. Subdural empyema
 4. Focal otic encephalitis
 5. Otitis externa
 6. Parotitis
- Physical Findings
 1. Severe tenderness over mastoid bone
 2. Mastoid area often edematous with erythema
 3. Presence of AOM, but may be normal if on antibiotics
 4. Pinna displaced downward and outward (late finding)
 5. Narrowing of ear canal in posterior superior wall due to pressure from mastoid abscess
 6. Purulent drainage and debris may be present in ear canal
- Diagnostic Tests/Findings
 1. CBC—elevated WBC
 2. Blood culture—rule out sepsis
 3. PPD—rule out exposure to tuberculosis
 4. Radiography or CT scan of mastoid(s)—diffuse clouding of mastoid cells; later in disease, bony destruction and resorption of the mastoid air cells; CT also done if brain abscess is suspected
 5. Tympanocentesis—identify pathogen
 6. Lumbar puncture—rule out meningitis
- Management/Treatment
 1. Prompt referral to ENT and hospitalization
 2. Incision and drainage of abscess; systemic intravenous antibiotics
 3. Complications—meningitis, brain abscess, cavernous sinus thrombosis, acute suppurative labyrinthitis, facial palsy
 4. Oral systemic antibiotics for 4 to 6 weeks after discharge

Cholesteatoma

- Definition: Cyst-like growth within the middle ear with lining of stratified squamous epithelium filled with desquamated debris
- Etiology/Incidence
 1. Congenital or acquired
 2. Varied theories explaining formation, e.g., inflammatory process, perforation or failure of desquamated tissue to clear from middle ear
 3. Most common cause of acquired cholesteatoma is long-standing chronic otitis media (Behrman, 2007)

4. If surgery delayed, it can invade and destroy other structures of the temporal bone and possibly spread to intracranial cavity, with life threatening consequences
5. If untreated, may lead to facial nerve paralysis, intracranial infection

- Signs and Symptoms
 1. Dizziness
 2. Hearing loss
- Differential Diagnosis: Aural polyps
- Physical Findings
 1. Pearly white, opacity, on or behind tympanic membrane
 2. History of chronic OM with foul smelling purulent otorrhea
- Diagnostic Tests/Findings
 1. CT scan of the temporal bone—detects presence and extent of the disease
 2. Audiogram to rule out hearing deficit
- Management/Treatment
 1. Referral to ENT for surgical excision
 2. Complications
 - a. Irreversible structural damage
 - b. Permanent bone damage
 - c. Facial nerve palsy
 - d. Hearing loss
 - e. Intracranial infection

Hearing Loss

- Definition: A deficit in hearing process classified as conductive, sensorineural, or mixed; can range from mild to severe, may be congenital or acquired; quantified by measured hearing threshold
 1. Conductive loss—normal bone conduction and reduced air conduction due to obstruction of transmission of sound waves through external auditory canal and middle ear to the inner ear; usual range of 15 to 40 dB loss
 2. Sensorineural loss—cochlea hair cells and/or auditory nerve damage; may range from mild to profound
 3. Mixed—components of conductive and sensorineural hearing loss present
 4. Hearing loss criteria in children differs from adults since children are in the process of speech and language development (Behrman, 2007)
 - a. Mild—15 to 30 dB
 - b. Moderate—30 to 50 dB
 - c. Severe—50 to 70 dB
 - d. Profound—70 dB and above

- Etiology/Incidence
 1. Hearing loss can be classified in five ways
 - a. Age of onset
 - b. Type—conductive, sensorineural, mixed, or central
 - c. Degree—ranging from mild to profound
 - d. Configuration—decibel (dB) loss
 - e. Hearing status of parent(s)
 2. Congenital
 - a. Sensorineural—moderate to profound loss; 1–2/000 live births (Behrman, 2007)
 - (1) Genetic—autosomal dominant (80%), autosomal recessive (20%)
 - (2) In utero infections—TORCH, CMV, and rubella most common causes in newborn
 - (3) Erythroblastosis fetalis
 - (4) Anoxia
 - (5) Birth trauma
 - (6) Birth weight < 1500 g
 - (7) Exposure to ototoxic drugs
 - (8) Prolonged mechanical ventilation
 - b. Conductive—congenital atresia, deformities, or stenosis of ossicles
 3. Acquired
 - a. Sensorineural
 - (1) Meningitis
 - (2) Mumps, measles
 - (3) Labyrinthitis, ototoxic drug exposure
 - (4) Severe head trauma
 - (5) Noise induced hearing loss (NIHL) from loud music, firecrackers, firearms, toy cap pistols, squeaking toys, recreational vehicles, farm equipment, lawn mowers, inappropriate hearing aids
 - b. Conductive
 - (1) Otitis media with middle effusion—75% of children have one episode of AOM with conductive hearing loss
 - (2) Cerumen impaction; foreign bodies in external canal
 - (3) Perforated/damaged tympanic membrane; severe head trauma
 - (4) Cholesteatoma, otosclerosis
- Signs and Symptoms
 1. Infants
 - a. Failure to elicit startle or blink reflex to loud sound
 - b. Failure to be awakened by loud sounds
 - c. General indifference to sound
 - d. Lack of babbling by 7 months
 2. Children
 - a. Substitution of gestures for words, especially after 15 months
 - b. Failure to develop intelligible speech by 24 months
 - c. Asking to have statements repeated
 - d. Inappropriate response to questions
 - e. Markedly inattentive
- Differential Diagnosis
 1. Mental retardation
 2. Profound deprivation
 3. Communication disorder—articulation disorders, expressive language delay, global language disorder, autism
- Physical Findings
 1. Careful evaluation of TM, e.g., decreased mobility, bulging, opacity
 2. Congenital abnormalities—external canal abnormalities, craniofacial malformations, structural abnormalities of external ear
- Diagnostic Test/Findings
 1. Audiogram appropriate for age to rule out hearing loss
 2. CT scan—rule out physiologic abnormalities
 3. Weber and Rinne tests—abnormal response
- Management/Treatment
 1. History screening at two week old visit—family history of hearing loss, perinatal infections (TORCH), anatomic malformations involving head or neck, birth weight < 1500 g, hyperbilirubinemia, bacterial meningitis, asphyxia
 2. Detect hearing loss as young as possible, take parental suspicions seriously; many tests available for all age groups including newborn (otoacoustic emissions—OAE, auditory brainstem response—ABR)
 3. Referral for full audiological testing and language evaluation as soon as deficit is strongly suspected; since the institution of universal newborn hearing screening, early detection of hearing loss has resulted in higher scores for language at school age
 4. Psychosocial considerations—rehabilitation, hearing aids, educational programs
 5. Prevention
 - a. Early identification and intervention; periodic hearing and language screening (birth to 4 months—responds appropriately to loud noises; 4 to 24 months—responds to noise out of field of vision; older children—pure tone audiometry)
 - b. Appropriate management and treatment of auditory canal obstruction and middle ear disease

- c. Avoid repeated exposure to loud noises to prevent NIHL
- d. Control erythroblastosis with use of RhoGAM; hyperbilirubinemia with phototherapy and exchange transfusions
- e. Prevent mumps and measles with immunization
- f. Avoid ototoxic medications

■ NOSE

Allergic Rhinitis

- Definition: IgE mediated response to inhaled allergens or irritants producing nasal mucosa inflammation
- Etiology/Incidence
 1. Types—seasonal or perennial depending on exposure/sensitization to allergen
 - a. Seasonal—inhaled pollens, e.g., trees, grasses; more common after age 6
 - b. Perennial—house dust mites, mold spores, animal dander; may occur in children under age of 6, uncommonly seen under 24 months
 2. Most common pediatric allergic disease; commonly associated with conjunctivitis, sinusitis, OME, and/or atopic dermatitis
 3. Strong genetic predisposition
- Signs and Symptoms
 1. Chronic, intermittent, or daily nasal congestion, and clear rhinorrhea
 2. Episodes of sneezing with itching of eyes, ears, nose, palate, pharynx
 3. Open mouth facies, snoring with sleep
 4. Excessive tearing
 5. Purulent secretions indicate secondary infection (e.g., sinusitis, foreign body)
 6. Symptoms year-round with perennial rhinitis
 7. Sinus headache
- Differential Diagnosis
 1. Bacterial or viral upper respiratory infection, e.g., strep pharyngitis, influenza, OM, sinusitis
 2. Vasomotor rhinitis
 3. Congenital or anatomical abnormalities leading to obstruction, e.g., nasal polyps, foreign body
- Physical Findings
 1. Allergic “shiners” and “salute” with nasal crease
 2. Hypertrophied turbinates; halitosis
 3. Nasal mucosa pale, boggy, and edematous with watery or mucoid secretions
- Diagnostic Tests/Findings
 1. Nasal smear for eosinophils—10% considered confirmatory
 2. RAST and skin testing—elevations or reactions to specific allergens
 3. CBC differential—elevated eosinophils and total IgE
- Management/Treatment: Referral to allergist may be necessary
 1. Allergen avoidance—first line of therapy
 2. Environmental controls—removal of carpets, drapes, and stuffed animals; plastic covers for mattresses and pillows; decrease humidity with air conditioner; use of air purifiers; avoidance of tobacco smoke
 3. Drug therapy—antihistamine H₁ receptor antagonists
 - a. Fexofenadine hydrochloride—
 - (1) 6 mo < 2 yr: 15 mg po bid;
 - (2) 2–11 yr: 30 mg bid;
 - (3) > 12 years: 60 mg bid;
 - (4) nonsedating
 - b. Loratadine—
 - (1) 2–5 yr: 5 mg qd;
 - (2) > 6 years: 10 mg daily;
 - (3) nonsedating
 - c. Cetirizine—
 - (1) 6 mo < 2 yr: 2.5 mg qd, may increase to 2.5 mg bid;
 - (2) 2–5 yr: 2.5 mg, may increase to 5 mg/24 hr;
 - (3) > 6 yr: 5–10 mg qd
 - d. Montelukast:
 - (1) 6 mo–5 yr: 4 mg oral granules or chewable tab qhs;
 - (2) 6–14 yr: 5 mg chewable qhs;
 - (3) > 14 yr: 10 mg qhs
 - e. Diphenhydramine—(5 mg/kg/day) every 6 hours; sedating
 - f. Oral sympathomimetic (e.g., pseudoephedrine) for short-term relief by producing vasoconstriction of respiratory tract mucosa
 - g. Mast cell stabilizer nasal spray to prevent allergic response—cromolyn sodium (> 2 years) one spray intranasally 4 times/day (not used as frequently now)
 - h. Topical nasal corticosteroids to decrease immediate and late phase allergic reactions with reduction of influx of inflammatory cells; start about 1 to 2 weeks before allergy season
 - (1) Beclomethasone—(> 6 years) by nasal inhalation
 - (2) Flunisolide—(> 6 years) by nasal inhalation

4. Referrals if symptoms not responsive to avoidance and/or medication
 - a. Allergist for skin testing for possible long-term immunotherapy
 - b. Immunotherapy effective in relieving symptoms due to dust mites, animal dander, pollens, molds, insect stings, and drug sensitivities
 - c. HEENT specialist for consultation and possible surgical interventions, e.g., myringotomy tubes

Chronic Rhinitis

- Definition: Chronic nasal discharge with or without acute exacerbations
- Etiology/Incidence
 1. May reflect underlying disorder, e.g., nasal polyps, chronic sinusitis, chronically infected tonsils, cystic fibrosis, allergy, foreign body, deviated septum, congenital malformation, syphilis
 2. May result from prolonged topical nasal decongestant use
- Signs and Symptoms: Variable
 1. Foul smelling, nasal discharge
 2. Possible bloody discharge, e.g., with foreign body, syphilis
 3. Disturbances in taste and smell
 4. Fever with superimposed infection
- Differential Diagnosis: Allergic rhinitis
- Physical Findings
 1. Excoriation of anterior nares and upper lip
 2. Nasal discharge—usually clear
- Management/Treatment: Directed at underlying cause
 1. Antibiotic for bacterial infection
 2. Environmental controls to minimize exposure to allergens
 3. Special attention to nutritional status, rest, and prevention of exposure to new infections

Foreign Body in Nose

- Definition: Foreign body in either nostril
- Etiology/Incidence: Common items include food, crayons, small toys, erasers, paper wads, beads, beans and stones, alkaline button batteries (batteries may release small amounts of chemicals leading to chemical burns/perforations and need immediate removal)

- Signs and Symptoms
 1. Initial—sneezing, mild discomfort, rarely pain; can increase with time
 2. Infection usually follows—unilateral purulent, malodorous, or bloody discharge is always suggestive of a foreign body
- Differential Diagnosis
 1. Nasal polyps, nasal tumors
 2. Purulent rhinitis, sinusitis
- Physical Findings
 1. Unilateral foul smelling nasal discharge
 2. Visualization of foreign body
- Diagnostic Tests/Findings: None usually; head mirror or light will provide visualization of object
- Management/Treatment
 1. Remove promptly with forceps or nasal suction
 2. Generally infection resolves after removal
 3. Referral to otolaryngologist if unable to remove

Epistaxis

- Definition: Bleeding from the nose
- Etiology/Incidence
 1. Most cases are benign and frequent in childhood due to increased vascularity of nasal mucosa
 2. Trauma and inflammation of mucosal lining (sudden onset) most common cause—nose picking, foreign body insertion, direct blunt trauma, violent sneezing
 3. Nasal mucosal drying (intermittent bleeding) from poorly humidified air, e.g., heating systems
 4. Chronic infection/inflammation of nasal tissue—viral/bacterial or allergies
 5. Substance abuse, e.g., cocaine, cannabis
 6. Systemic diseases—consider with bleeding that is severe, prolonged, or recurrent
 - a. Hypertension—exacerbates problem
 - b. Clotting abnormalities, e.g., hemophilia, aplastic anemia, leukemia, idiopathic thrombocytopenia, platelet dysfunction
 - c. ASA and NSAID overuse, neoplasms (gradual onset), cancer treatments, hormonal influences, e.g., menses, birth control pills (BCP), pregnancy
- Signs and Symptoms
 1. Bleeding from nares—usually unilateral
 2. Tarry stools—occasionally with frequent bleeds

- Differential Diagnosis
 1. Foreign bodies
 2. Infection
 3. Substance abuse
 4. Allergies
 5. Chronic rhinitis
 6. Chronic nasal spray use
- Physical Findings
 1. Determine location of bleeding
 - a. Anterior bleed—most common site (90%) Kiesselbach's plexus
 - b. Posterior bleed—can see only if nose is normal, no inflammation, and with special instruments; may see posterior oropharynx blood flow
 - c. High nasal bleed—may represent nasoethmoid or orbit fracture
 - d. Recurrent—consider bleeding disorder or chronic irritation
 2. Assess nares for growths, septal hematoma (needs immediate attention)
 3. Assess for other signs of bleeding, excess bruising, petechiae/purpura
- Diagnostic Tests/Findings
 1. Stool for occult blood—determine if child is swallowing blood
 2. Roentgenograms to determine if nasal fracture, foreign body
 3. CBC with differential, platelets, PT, PTT; coagulation profile (if bleeding disorder is suspected)
- Management/Treatment
 1. Complete history including—recurrent or acute, unilateral or bilateral, duration, recent URI, allergic rhinitis, ASA or NSAID use, signs of underlying disease
 2. Monitor vital signs, especially blood pressure
 3. Apply pressure to anterior nasal septum with patient sitting in upright position with head tilted forward (most stop within 10 to 15 minutes); application of ice, increase humidity of home, avoid nose blowing
 4. Antibiotics (topical and oral) may be indicated if infection is suggested
 5. Topical nasal vasoconstrictor drops (phenylephrine $\frac{1}{4}\%$ to $\frac{1}{2}\%$) and packing may be needed
 6. Recurrent or severe cases—refer to otolaryngologist
- Etiology/Incidence
 1. Common pathogens (also commonly found in AOM)
 - a. Predominately in acute sinusitis—*Streptococcus pneumoniae*, *Haemophilus influenzae*, and *Moraxella catarrhalis*
 - b. Prevalent in chronic sinusitis are group A beta hemolytic streptococcus and *Staphylococcus aureus* and anaerobes
 - c. Viruses—less common
 2. 5% to 10% of URIs in children develop into sinusitis
 3. May be secondary to allergies, adenoidal hypertrophy, anatomical abnormalities, dental abscess, diving and swimming
 4. Patients with cystic fibrosis and immune deficiencies are at a higher risk for sinus infections
 5. Maxillary and ethmoid sinuses most frequently involved
 6. Complications are uncommon overall but are serious; orbital cellulitis is the most common, intracranial abscesses and osteomyelitis of the frontal bone (Potts puffy tumor) can also occur
 7. Adolescent males are at higher risk for intracranial abscesses
 8. Acute sinusitis—persistent symptoms > 10 days and < one month
 9. Chronic sinusitis—> 30 days
- Signs and Symptoms: Children have less specific complaints; adolescents usually present with classical symptoms
 1. Persistent URI (beyond 7 to 10 days) with purulent or watery drainage
 2. Cough, low-grade fever
 3. Facial pain, toothache, headache or tenderness over involved sinus
 4. Postnasal drip, bad breath, sore throat
 5. Increased pain with cough, bending over, or abrupt head movement
 6. Fatigue, malaise, anorexia
 7. Periorbital swelling in morning
- Differential Diagnosis
 1. Dental infections, cleft palate, foreign bodies, tumors and polyps, nasal trauma, malformations
 2. Allergic and purulent rhinitis, common cold
 3. Cystic fibrosis, immunodeficiency states, allergy, or asthma
- Physical Findings
 1. Clear or mucopurulent rhinorrhea and/or postnasal drainage
 2. Erythema of nasal mucosa and/or throat
 3. Pain on percussion with possible erythema/edema in area of affected sinus

Sinusitis

- Definition: Acute, subacute, or chronic inflammation of mucosal lining in one or more of paranasal sinuses

4. OME common finding—especially in younger child
 5. Assess for complications—eyelid swelling with orbital cellulitis, persistent, recurrent or worsening fever/headache, vomiting, neurological deficits (intracranial abscess)
- Diagnostic Tests/Findings
 1. Nasal scrapings—eosinophils present with allergy
 2. CT scan—primary diagnostic tool to diagnose sinus disease and complications; obtained if recurrent, uncertain of diagnosis, unresponsive to treatment, assess for complications
 3. Sinus x-rays—not routine
 - Management/Treatment
 1. Antibiotic therapy—same as used in AOM (see Table 4-1)
 - a. To penetrate sinuses, need 14 to 21 days of treatment, up to 6 weeks in chronic cases
 - b. Clindamycin and Trimethoprim/sulfamethoxazole can be used but have limitations; clindamycin is not effective against gram-negative organisms such as *H. flu* and trimethoprim/sulfamethoxazole; is not effective against group A streptococci
 2. Decongestants/antihistamines not proven effective except with concomitant allergic manifestations
 3. Nasal sprays—topical inhaled steroids (budesonide) and normal saline nasal spray; nasal decongestants can be used but not for > 3 days
 4. Comfort measures—analgesics, increased humidity, increase oral fluids
 5. Diving/swimming in moderation; consider elimination of activity with chronic cases
 6. Refer to otolaryngologist and/or allergist for chronic sinusitis and allergy control

Nasal Polyps

- Definition: Benign pedunculated tumors
- Etiology/Incidence
 1. Originate from edematous, chronically inflamed nasal mucosa
 2. Common in children with cystic fibrosis; 25% develop nasal polyps
- Signs and Symptoms
 1. Nasal obstruction
 2. Mouth breathing

- Differential Diagnosis
 1. Chronic sinusitis
 2. Cystic fibrosis
 3. Chronic allergic rhinitis
 4. Asthma
- Physical Findings
 1. Hyponasal obstruction
 2. Profuse mucoid/mucopurulent rhinorrhea
 3. Shiny grey, grape-like mass(es) between nasal turbinates and septum
- Diagnostic Tests/Findings: Sweat test to rule out cystic fibrosis on every child with polyps
- Management/Treatment
 1. Care in distinguishing swollen turbinates from polyps
 2. Antihistamines if allergy related
 3. Local/systemic decongestants/corticosteroids not helpful
 4. Refer for surgical removal in those with complete obstruction or uncontrolled deformity

■ MOUTH

Oral Candidiasis (Thrush)

- Definition: Common fungal infection of oral cavity
- Etiology/Incidence
 1. *Candida albicans* (monilial)
 2. More common in neonate and infant than in other age groups (transmission during vaginal delivery), affecting 2 to 5% of newborns
 3. Predisposing factors
 - a. Steroid therapy
 - b. Antibiotic therapy
 - c. Compromised immune system
- Signs and Symptoms
 1. White patches in mouth
 2. May be painful or produce no discomfort
- Physical Findings
 1. Characteristic white, curd-like adherent plaques that are not easily removed; found on the buccal mucosa, tongue, pharynx, and/or tonsils
 2. Bleeding occurs with attempts to remove plaques
- Diagnostic Tests/Findings: Systemic/immune status evaluation if persistent or resistant to treatment

- Management/Treatment
 1. Antifungal therapy—nystatin oral suspension applied to oral mucosa 4 times/day for 10 days; if not responding, consider oral fluconazole; gentian violet can be used but is less effective than the newer antifungals
 2. If breastfeeding, consider examining and treating mother for candidiasis of breast (cross-infection)
 3. Check diaper area for concurrent diaper rash
 4. Sterilize nipples, pacifiers if bottle-fed; apply nystatin to nipples if breast feeding; if recurrent, look for other causes, e.g., H.I.V.

Cleft Lip/Palate

- Definition
 1. Cleft lip—failure of embryonic structures surrounding oral cavity to join
 2. Cleft palate—failure of palatal shelves to fuse
 3. Occurs in various degrees
- Etiology/Incidence
 1. Genetic influence with cleft lip more than palate
 2. Both can occur sporadically
 3. More common to have combination of both than one without the other
 4. Cleft lip with or without palate—1:800 births
 5. Cleft palate alone—1:1750 births
 6. More common in males, Asians, and with maternal drug exposure
 7. Lower incidence in African-Americans
 8. Increase in middle ear/nasopharyngeal/sinus infections with associated hearing loss
 9. Recurrent otitis media is common
- Signs and Symptoms: Separation of lip and/or palate
- Differential Diagnosis: None
- Physical Findings
 1. Degree of cleft varies from small notch to complete separation
 2. Unilateral or bilateral
 3. Involves soft and/or hard palate
 4. Bifid uvula indicates submucosal cleft palate
- Diagnostic Test/Findings: Audiogram to rule out hearing deficit
- Management/Treatment
 1. Surgical repair; timing individualized—lip usually by 2 months, palate by 9 to 12 months
 2. Teach feeding techniques (breast or bottle) before and after repair

3. Referral for dental restoration if needed
4. Referrals to otolaryngologist, plastic surgeon, pediatric dentist, prosthodontist, orthodontist, speech therapist, psychiatrist, social worker, and genetic counselors
5. Support family adjustment and management

Dental Caries

- Definition: Demineralization of tooth surface secondary to production of organic acids by bacterial fermentation of dietary carbohydrates
- Etiology/Incidence
 1. Mutans streptococci are primary etiological organism responsible for human dental caries
 2. Rate of formation depends on frequency of acid environment in mouth, availability of fluoride for remineralization, oral hygiene
 3. Nursing bottle tooth decay (nursing caries)
 - a. Tooth decay resulting from repeated/prolonged contact with milk, formula, or juice
 - b. Children put to bed with bottle or sleep at the breast or use either the breast/bottle as pacifier are at increased risk
 4. Most pediatric dental caries occur on the occlusal surfaces of posterior and lingual aspect of the maxillary incisors, molars, and cuspids; nursing bottle caries affect the upper central and lateral incisors
 5. New teeth are at greater risk for caries than established teeth
 6. Fluoride is critical in the process of remineralization
 7. Incidence has decreased in past 30 years but remains high in low-income children
- Signs and Symptoms
 1. Sensitive or painful tooth
 2. Severe decay—pain, edema and infection
 3. Weight loss—if severe
 4. Feeding problems
- Physical Findings
 1. Initial decalcification of enamel appears as opaque white spots that turn light brown progressing to dark brown with destruction of tooth
 2. Left untreated, caries can progress to dental abscess (pain, facial swelling)
- Management/Treatment
 1. Prevention of dental caries/periodontal disease
 - a. Well balanced diet with appropriate feeding practices; low sugar and complex carbohydrate consumption

- b. Wean from bottle, pacifier, and breast at one year of age
 - c. Brush/clean/wipe teeth as soon as they appear
 - d. Daily brushing and flossing of teeth; for children under 8 years, parental involvement needed
 - e. Fluoride supplement, dental sealants to occlusal surfaces of the posterior teeth
 - f. Early dental visits—American Academy of Pediatric Dentists recommend starting dental visits at 12 to 18 months of age for initial discussion of oral hygiene, weaning, and fluoride supplementation; routine dental checkups starting at 3 years
2. Dental referral for identified caries

Aphthous Ulcers (Canker Sores)

- Definition: Shallow, painful mouth ulceration, prone to recurrence, two types: minor and major
- Etiology/Incidence
 1. Minor aphthous ulcers, more common, occur on the unattached gingiva; major ulcerations are larger and appear on attached gingiva or palate
 2. Appearance: well defined lesion, appears as an ulcer with yellow/white necrotic base with surrounding erythema
 3. Cause is unknown but multiple theories—trauma, stress, sun, food allergies, endocrine or hematologic disorders, infectious agents (viral), autoimmune basis
 4. Onset often in adolescence (20%) and recurrent
 5. “Minor”—1 to 5 lesions, 1 cm, lasting 7 to 14 days
 6. “Major”—10% of cases are “major,” defined as lesions that are greater than 1 cm, lasting > 6 weeks
- Signs and Symptoms
 1. Burning or tingling before appearance of lesion
 2. Pain at lesion site
 3. Afebrile
- Differential Diagnosis
 1. Herpetic lesions (simplex zoster)
 2. Herpangina (coxsackievirus A)
 3. Trauma
 4. Chemical burns
 5. Hand, foot, and mouth disease (coxsackievirus A5, A10, A16)

- Physical Findings
 1. Single or multiple, small, oval, indurated papules with erythematous halo; develops pale center that erodes into ulcers
 2. No systemic symptoms
- Diagnostic Test/Findings: None
- Management/Treatment
 1. Oral analgesics, e.g., 6 to 12 years use chlor-aseptic spray; greater than 12 years use viscous xylocaine solution, steroid in orabase (i.e., triamcinalone in orabase)
 2. Antibacterial (tetracycline) rinses may shorten disease course in children > 9 years of age
 3. Referral to pediatric dentist if condition lasts more than 14 days

Herpes Labialis (Cold Sore, Fever Blister) and Herpes Simplex Stomatitis

- Definition
 1. Ulceration and inflammation of oral mucosa from the herpes virus
 2. Acute primary herpetic gingivostomatitis (APHGS) occurs in previously unexposed children
- Etiology/Incidence
 1. Virus acquired from individual who has mouth sore or herpetic whitlow on a finger or toe; caused by herpes simplex virus 1 (HSV-1)
 2. Illness starts 5 to 10 days after exposure
 3. Spontaneous recovery in 7 to 10 days
 4. 50% develop subsequent cold sore episodes after primary acute episode; reactivated from the sensory neurons, by sunlight, stress, fevers.
- Signs and Symptoms
 1. May be asymptomatic, especially cold sores/fever blisters
 2. Initial symptoms include fever, chills, irritability, tender submandibular adenopathy, ulcerative exanthem of the gingiva and mucous membranes of the mouth
 3. Sore throat, burning in mouth and throat
 4. Anorexia from pain; could lead to dehydration
 5. Recurrent herpes simplex limited to a few lesions on the lips.
- Differential Diagnosis
 1. Aphthous ulcers
 2. Hand, foot, and mouth syndrome
 3. Herpangina

- Physical Findings
 1. Cold sores—grouped vesicles on erythematous base; commonly found on mucocutaneous border of lips
 2. Primary gingivostomatitis—vesicles on oral mucosa, gingiva, tongue, and lips; ulcer formation following vesicle stage which bleed easily; diffuse erythematous, edematous gingiva, especially the interdental papillae
 3. Cervical adenopathy with gingivostomatitis
 4. May spread to perioral skin
- Diagnostic Tests/Findings: None
- Management/Treatment
 1. “Over the counter” (OTC) cold sore treatment
 2. Pain management—acetaminophen or ibuprofen; topical relief with occlusive gels, e.g., infant oral anesthetic agents; 1:1 mixture of diphenhydramine combined with antacid preparations consisting of magnesium and aluminum hydroxide *or* antidiarrheal preparations to provide a protective coating for the oral mucosa (severe cases add 2% viscous lidocaine sparingly)
 3. Acyclovir may be considered in select patients for primary episodes but is not routinely used; must be started in first 72 hours (Bernius, 2006)
 4. Spontaneous recovery within 2 weeks, rarely complications; dehydration is a concern in primary herpes

Hand, Foot, and Mouth Disease

- Definition: Acute viral illness presenting with vesicular exanthem on tongue, gums, palate, oral mucosa; papulovesicular exanthem on hands, feet, legs, and occasionally the buttocks
- Etiology/Incidence
 1. Coxsackievirus A16 (most common), A5, and A10
 2. Enterovirus 71—frequently more severe illness (aseptic meningitis, encephalitis, and paralytic disease)
 3. Seasonal, predominant in summer and fall
 4. Incubation 4 to 6 days
 5. Spontaneous resolution in one week
- Signs and Symptoms
 1. Fever if present, typically low-grade
 2. Anorexia
 3. Dysphagia
- Differential Diagnosis
 1. Acute primary herpetic gingivostomatitis
 2. Aphthous stomatitis, herpangina
 3. Trauma, burns

- Physical Findings
 1. Small vesicles erode to ulcers on buccal mucosa, hard palate, tonsils, and tongue
 2. Vesicular lesions appear as blanching red lesions on arms, legs, palms, soles
 3. Lesions on buttocks are not usually vesicular
- Diagnostic Tests/Findings: Unnecessary
- Management/Treatment: See Herpangina

Herpangina

- Definition: Acute viral illness presenting with ulceration and inflammation of oral mucosa
- Etiology/Incidence
 1. Coxsackievirus, group A (most common)
 2. Coxsackie B viruses and echoviruses (less common)
 3. Seasonal in U.S.—predominant in summer months
 4. Resolves spontaneously in 3 to 5 days
- Signs and Symptoms
 1. Fever in moderate range
 2. Headache, myalgia, malaise
 3. Dysphagia, vomiting (25%), anorexia, significant oral discomfort, drooling
- Differential Diagnosis
 1. Acute primary herpetic gingivostomatitis
 2. Aphthous stomatitis
 3. Trauma or burns
 4. Hand, foot, and mouth disease
 5. Pharyngitis/tonsillitis
- Physical Findings: Small vesicles or punched-out ulcers, especially on soft palate and tonsillar pillars; anterior structures (i.e., gingiva, buccal mucosa, and hard palate) are typically not affected
- Diagnostic Tests/Findings: Unnecessary
- Management/Treatment
 1. Fever and/or pain control—acetaminophen, ibuprofen
 2. Topical relief with 1:1 mixture of diphenhydramine combined with antacid preparations consisting of magnesium and aluminum hydroxide *or* antidiarrheal preparations to provide a protective coating for the oral mucosa (severe cases add 2% viscous lidocaine; use sparingly)
 3. Encourage fluids to ensure adequate hydration

■ THROAT

Pharyngitis and Tonsillitis

- Definition: Acute inflammation and infection of the throat; when tonsils are main focus of inflammation, tonsillitis is more appropriate term to use
- Etiology/Incidence
 1. Causes vary by geographic location, season, age; most common in 5- to 15-year-olds
 2. Viruses
 - a. Approximately 80% to 90% of sore throats are viral; numerous viruses, most are not distinguishable from each other (Hay, 2009)
 - b. Virus is probable cause in conjunction with nasal congestion and rhinorrhea
 - c. Adenovirus is common; other viruses include influenza, RSV, parainfluenza and cytomegalovirus
 - d. Epstein-Barr virus—associated with infectious mononucleosis
 - e. Enteroviruses (e.g., coxsackievirus A and echovirus) seen in summer and fall
 3. Bacteria
 - a. Group A beta hemolytic streptococcus (GABHS) account for 15% to 30% of all pharyngitis cases; typically occurring in late winter and early spring (Choby, 2009)
 - b. *Neisseria gonorrhoeae*—in sexually active adolescents or sexually abused children
 - c. *Corynebacterium haemolyticum* and *Corynebacterium diphtheriae*—characteristic presence of grey pseudomembranous exudate on pharynx and tonsils which bleeds with attempts at removal; quite rare in U.S.; still seen in developing countries
 4. Noninfectious causes (no fever or illness symptoms)
 - a. Trauma from tobacco smoke, heat, alcohol
 - b. Allergic rhinitis or postnasal drainage
 5. Transmission—through exposure to infected respiratory secretions, shared silverware
 6. Complications—most are complications of GABHS
 - a. Peritonsillar or retropharyngeal abscess or cellulitis
 - b. Cervical adenitis, AOM, sinusitis, pneumonia
 - c. Acute rheumatic fever in untreated group A beta hemolytic streptococcal pharyngitis—prevented if treatment started within 9 days of initial complaints of sore throat
 - d. Glomerulonephritis—host/immune response to infection with GABHS; not all strains are nephritogenic; manifests in 1 to 3 weeks after pharyngeal or skin infection of GABHS; unrelated to treatment
- Signs and Symptoms: Common symptomatology with some variability by causative organism
 1. Sudden or gradual onset of symptoms
 2. Sore throat
 3. Fever, variable
 4. Headache, anorexia, occasional nausea, vomiting, abdominal pain, and malaise
 5. Viral pharyngitis—hoarseness, conjunctivitis, runny nose, cough, cold symptoms
 6. GABHS pharyngitis—usually seen 2 years of age and older, sudden onset of fever with complaints of headache, abdominal pain, and vomiting; scarletina rash, “strawberry tongue” may be present
- Differential Diagnosis
 1. Stomatitis
 2. Peritonsillar or retropharyngeal abscess, epiglottitis
 3. Allergic rhinitis, postnasal drainage
- Physical Findings
 1. Erythema of pharynx, of varying degrees; one presentation of GABHS is a beefy red appearance; petechial lesions on the soft palate can be seen with GABHS
 2. Enlarged tonsils with exudate can be seen with both viral and strep infections
 3. Erythema of nasal mucosa with coryza—more consistent with viral sore throats
 4. Cervical nodes usually enlarged with possible tenderness
- Diagnostic Tests/Findings
 1. Rapid strep test to determine presence of GABHS
 - a. Newer rapid strep tests 90% to 99% sensitive (older latex agglutination assays were 70% sensitive)
 - b. Treat if positive; throat culture to confirm negative test
 2. CBC—WBC may be elevated with bacterial infection and normal or decreased with viral infections, but not entirely reliable
 3. Consider other studies—dependent on history, age, and clinical presentation, e.g., mono spot, EBV, culture for gonorrhea, diphtheria culture
- Management/Treatment
 1. Viral pharyngitis/tonsillitis—symptomatic/supportive care
 - a. Saline gargles, throat lozenges
 - b. Analgesics for fever/pain (acetaminophen, ibuprofen)

- c. Encourage fluids for maintaining hydration
- 2. Bacterial pharyngitis/tonsillitis
 - a. GABHS—penicillin drug of choice, 125 to 250 mg every eight hours for 10 days (500 mg bid in adults and children weighing 60 lbs) or IM penicillin G benzathine (600,000 U for less than 60 lbs, 1.2 million U for greater than 60 lbs); amoxicillin often substituted for penicillin because of better taste
 - b. Erythromycin or first generation cephalosporin for those with penicillin allergy (cephalosporins can be substituted if nonanaphylactic reaction)
 - c. Second line therapy include macrolides, cephalosporins, or clindamycin
 - d. Considered noncontagious after 24 hours on antibiotic
 - e. Gonococcal pharyngitis—one IM injection of ceftriaxone
 - f. Diphtheria—hospitalization and treatment with erythromycin or penicillin G

Acute Nasopharyngitis (Common Cold)

- Definition: Acute viral infection of upper respiratory tract with potential involvement of nasal passages, sinuses, Eustachian tubes, middle ears, conjunctiva, and nasopharynx
- Etiology/Incidence
 - 1. Causative pathogens
 - a. Over 100 infectious pathogens—respiratory syncytial virus (RSV) most common
 - b. Other common pathogens include parainfluenza viruses, corona viruses, adenoviruses, enterovirus, influenza viruses, *Mycoplasma pneumoniae*
 - 2. Pathogen shed in large amounts through nasal secretions and easily spread through self-inoculation from fingers and hands to objects (clothing, environmental surfaces)
 - 3. Universal susceptibility; children average 5 to 8 infections/year with a peak incidence during first 2 years
 - 4. Increased susceptibility associated with active/passive smoke exposure
 - 5. More frequent in crowded situations
 - 6. Occurrence in cooler months in temperate climates—peaks in early fall, late January, and early April
- Signs and Symptoms: Generally lasts one week; dry cough with rhinorrhea, may persist up to 3 weeks
 - 1. Infants
 - a. Irritability, restlessness, fever (100° to 102°F)
 - b. Rhinorrhea
 - c. Occasional diarrhea
 - d. Changes in feeding and sleep patterns
 - 2. Older children
 - a. Afebrile or low-grade fever, stuffy nose, watery nasal discharge
 - b. Sore throat, sneezing, cough, chills
 - c. Occasional headache, malaise
- Differential Diagnosis
 - 1. Underlying secondary bacterial infection—sinusitis, OM, pharyngitis, lower respiratory tract disease
 - 2. Allergic rhinitis
 - 3. Foreign body
 - 4. Substance abuse in older children and adolescents, or overuse of medicated nasal spray
- Physical Findings
 - 1. Coryza
 - 2. Inflamed, moist nasal mucosa and oropharynx
 - 3. Chest clear
- Diagnostic Tests/Findings
 - 1. Viral cultures expensive, generally unnecessary
 - 2. If suspicious of differential diagnosis, consider additional tests such as throat culture, chest or sinus x-rays, allergy testing
- Management/Treatment: Symptomatic/supportive care
 - 1. Analgesics for sore throat, muscle aches and fever > 101°F
 - 2. Relief of nasal congestion
 - a. Saline nose drops with nasal bulb syringe
 - b. Cool mist humidification
 - c. Antihistamines and decongestants not routinely recommended
 - d. Antibiotics are not indicated in viral infections
 - 3. If symptoms persistent beyond 7 to 10 days, consider secondary infection
 - 4. Maintain hydration
 - 5. Prevention
 - a. Good hygiene and cleaning of clothes, toys, and play areas
 - b. Limited exposure to crowded situations

Retropharyngeal Abscess

- Definition: Infection of the retropharyngeal lymph nodes; inflammation of posterior aspect of pharynx with suppurative retropharyngeal lymph nodes
- Etiology
 1. Usually preceded by URIs, pharyngitis, sinusitis, and cervical lymphadenitis
 2. In older children—usually superinfection from penetrating injury to posterior wall of oropharynx
 3. Most common organisms—GABHS and staph aureus
 4. Relatively rare infection, most common in children under 6 years of age with a peak incidence age 3
- Signs and Symptoms
 1. Acute onset of high fever with persistent severe throat pain
 2. Drooling due to difficulty in swallowing
 3. Tachypnea, dyspnea, stridor
 4. Neck and head hyperextension
- Differential Diagnosis
 1. Epiglottitis
 2. Peritonsillar abscess
 3. Laryngotracheobronchitis (croup)
 4. Acute infectious mononucleosis
 5. Acute pharyngitis
 6. Bacterial tracheitis
 7. Meningitis
- Physical Findings
 1. Toxic appearing child; neck and head in hyperextension
 2. Noisy, gurgling respiration
 3. Drooling, meningismus
 4. Stridor, airway obstruction
 5. Possible neck swelling, torticollis
 6. Prominent swelling of posterior pharyngeal wall—confirms diagnosis
- Diagnostic Tests/Findings
 1. Lateral neck radiography—retropharyngeal space wider than C4 vertebral body or > 6 mm at C2
 2. CBC—elevated WBC common
 3. Computed tomography (CT) scan to visualize abscess
- Management/Treatment
 1. Immediate emergency referral to ENT
 2. Emergency hospitalization necessary
 - a. Admission to pediatric ICU for continuous monitoring for airway obstruction and possible respiratory arrest

- b. Surgical incision and drainage necessary
- c. Intravenous (IV) antibiotics—penicillin, clindamycin

Peritonsillar Abscess

- Definition: Infection of tonsils spreading to tonsillar fossa and surrounding tissues (peritonsillar cellulitis); if left untreated, tonsillar abscess forms
- Etiology/Incidence
 1. GABHS (most common)
 2. Staphylococcus aureus, anaerobic microorganisms
 3. Can occur at any age; more common in pre-adolescent or adolescent age groups
 4. Complication of untreated peritonsillar abscess—lateral pharyngeal abscess leading to possible airway obstruction; aggressive early treatment needed
- Signs and Symptoms
 1. Severe sore throat with high fever
 2. Toxic appearance, muffled voice, spasms of jaw muscles
 3. Difficulty swallowing and drooling in severe cases
 4. Bad breath
- Differential Diagnosis
 1. Retropharyngeal abscess
 2. Epiglottitis
- Physical Findings
 1. Unilateral enlargement of tonsil(s), bulging medially with anterior pillar prominence (most common)
 2. Soft palate and uvula edematous, erythematous, with uvula displaced towards unaffected side
 3. Extreme tonsillar tenderness on palpation
 4. Trismus can occur, making visualization of the pharynx difficult
- Diagnostic Tests
 1. CBC—increased WBC
 2. Rapid strep test to rule out GABHS
- Management/Treatment
 1. Immediate referral to ENT
 2. Surgical incision and drainage often necessary
 3. Hospitalization common for 24 hours (Hay, 2009); if not hospitalized, daily follow-up visits until stable
 4. Antibiotics (penicillin or clindamycin), IV initially, and discharged on oral antibiotics

Cervical Lymphadenitis

- Definition: Inflammation/infection affecting one or more cervical lymph nodes
- Etiology/Incidence
 1. Pathogens
 - a. Streptococcus pyogenes and Staphylococcus aureus account for approximately 80% of cases
 - b. Mycobacterium tuberculosis
 - c. Other organisms (e.g., viral, fungal, or parasitic)
 2. Secondary to local infections of the ear, nose, and throat (most common)
 3. Prevalent among preschool children
- Signs and Symptoms
 1. Complaints of swollen neck or face
 2. Fever commonly present
 3. Stridor, hoarseness, drooling if adenopathy impinges on airway
- Differential Diagnosis
 1. Bilateral cervical adenitis—mononucleosis, tularemia, diphtheria
 2. Subacute or chronic adenitis—cat scratch fever, nonspecific viral infections
 3. Atypical mycobacterium—tuberculosis
 4. Cervical node tumors, e.g., leukemia
 5. Mumps, cyst, hematoma
- Physical Findings
 1. Large unilateral cervical mass, greater than 2 to 6 cm
 2. Erythema may be present
 3. Tenderness on palpation
- Diagnostic Tests/Findings
 1. CBC—moderate to marked WBC increase
 2. PPD—rule out tuberculosis
 3. Mono spot—rule out mononucleosis
 4. Throat culture—rule out GABHS
 5. Serology tests if not resolving (e.g., EBV, toxoplasmosis, CMV, histoplasmosis)
 6. Aspiration of node if fluctuant—aerobic/anaerobic culture
- Management/Treatment
 1. With no evidence of sepsis, treat empirically with oral antibiotics—dicloxacillin, amoxicillin clavulanate, or cephalexin for a minimum of 10 days and no less than 5 days after resolution of symptoms
 2. Measure and follow size of node
 3. Analgesics for fever and pain; application of cold compresses

4. Reevaluation after 36 to 48 hours; if no improvement, possible hospitalization for IV antibiotics, especially with infants and young children
5. Referral to otolaryngologist if not improving
6. Surgical aspiration may be necessary
7. Persistent unexplained, symptomatic node, increasing in size despite treatment, refer for biopsy

Epiglottitis (Supraglottitis)

- Definition: Severe, rapidly developing inflammation and swelling of the supraglottic structures leading to life threatening upper airway obstruction
- Etiology/Incidence
 1. Can occur at any age, highest incidence 2 to 7 years of age
 2. Pathogens—group A beta-hemolytic streptococci, pneumococci, Haemophilus influenzae
 3. Decreasing incidence from Haemophilus influenzae with use of Hib vaccine by 99% in children under 5 years, but can still occur even with a complete set of vaccines (Rafei, 2006)
- Signs and Symptoms
 1. Acute, sudden onset of high fever, severe sore throat, muffled voice, drooling, poor color, labored breathing in a previously well child
 2. Choking sensation, refuses to speak
 3. Restless, irritable, anxious, apprehensive, frightened
 4. Hyperextension of neck, leaning forward and chin thrust out, prostration; “sniffing dog” or tripod position—provides best possible airway
- Differential Diagnosis
 1. Acute laryngeal edema, croup syndrome, foreign body aspiration
 2. Pertussis, diphtheria, Kawasaki syndrome
 3. Bacterial tracheitis, retropharyngeal abscess
- Physical Findings
 1. Rapidly progressive respiratory distress—suprasternal and subcostal retractions, soft inspiratory stridor, nasal flaring leading to possible respiratory arrest
 2. Toxic, distressed appearance
 3. Beefy erythematous epiglottis
 4. If epiglottitis suspected, do not attempt to visualize

- Diagnostic Tests/Findings: Often deferred to minimize distress
 1. CBC—WBC (greater than 18,800); laboratory examination is low priority in child with severe respiratory distress
 2. Cultures of blood, tracheal, or epiglottis secretions—identifies pathogen
 3. Radiograph—lateral neck shows a thickened/swollen epiglottis (“thumb sign”); may elect not to perform radiograph due to possibility of airway obstruction and respiratory arrest; airway can be safely visualized in surgery
- Management/Treatment
 1. Requires prompt recognition and treatment; represents a true medical emergency; death can occur within hours
 2. While waiting for emergency transport; provide oxygen, keep child calm, be prepared for emergency cardiopulmonary resuscitation
 3. Following diagnosis, airway must be established by nasotracheal or endotracheal intubation or elective tracheotomy immediately; usually extubated within 24 to 48 hours after reducing epiglottis and afebrile
 4. IV antibiotic therapy for 2 to 3 days
 - a. Third generation cephalosporins until initial pathogen identified; antibiotics most commonly used include ceftriaxone and ampicillin-sulbactam (Rafei, 2006)
 - b. In areas with penicillin and cephalosporin resistant pneumococci, vancomycin is drug of choice
 5. Oral antibiotic to follow IV to complete 10-day course
 6. Corticosteroid therapy to reduce swelling
 7. Prevention—Haemophilus influenzae type b vaccine at 2, 4, 6 or 12, and 15 months of age; no vaccines against other pathogens

■ QUESTIONS

Select the best answer

1. Which one of the following may cause microcephaly?
 - a. Hypocalcemia
 - b. Craniosynostosis
 - c. Skull fracture
 - d. Seizure disorder
2. What finding may accompany macrocephaly?
 - a. Pulsating anterior fontanel
 - b. Sunken fontanel
 - c. Premature closure of suture lines
 - d. Widened suture lines
3. Obtaining a CT of the head would be indicated in which of the conditions?
 - a. Macrocephaly
 - b. Cephalohematoma
 - c. Craniosynostosis
 - d. Caput succedaneum
4. Which one of the following conditions increases the risk of developing hydrocephalus?
 - a. Bilateral cephalohematomas
 - b. Craniosynostosis
 - c. Prematurity
 - d. Familial macrocephaly
5. A conjunctivitis appearing in a 2-day-old newborn is likely due to:
 - a. Chemical irritation from eye drops
 - b. Group B streptococcus
 - c. Chlamydia
 - d. Gonorrhea
6. Confirming the diagnosis of chlamydia conjunctivitis in a newborn would best be done by obtaining which one of the following?
 - a. Cervical swab of the mother
 - b. Urine PCR from the mother
 - c. Culture of the eye discharge
 - d. Culture of the conjunctival scrapings
7. Which one of the following eye findings would be considered an ophthalmic emergency?
 - a. Unilateral vesicular lesions on the upper eyelid in a 3-week-old
 - b. Presence of chemosis in a 5-year-old with bilateral upper eyelid edema
 - c. Cobblestone-like appearance along the inner aspect of the upper eyelid in a 15-year-old
 - d. Bilateral redness along eyelid margins with tiny ulcerated areas in a 16-year-old
8. The most appropriate management of a 5-year-old with a firm, nontender nodule in the mid-upper eyelid for 3 weeks would be:
 - a. Cool compresses
 - b. Topical ophthalmic ointment
 - c. Oral antibiotics
 - d. Oral steroids
9. Daily eyelid cleansing with diluted baby shampoo and a cotton-tipped applicator would be appropriate in the treatment of which one of the following conditions?
 - a. Dacryostenosis
 - b. Chalazion
 - c. Hordeolum
 - d. Blepharitis

10. A 3-year-old has an edematous, mildly erythematous right upper eyelid for one day with a fever of 103°F. An important eye assessment would be:
 - a. Ocular mobility
 - b. Conjunctival inflammation
 - c. Pupillary reaction
 - d. Optic disc papilledema
11. Concurrent otitis media and conjunctivitis is likely due to which organism?
 - a. *Streptococcus pneumoniae*
 - b. *Haemophilus influenzae*
 - c. *Moraxella catarrhalis*
 - d. *Staphylococcus aureus*
12. All but which one of the following is consistent with glaucoma?
 - a. Photophobia
 - b. Epiphora (increase tears)
 - c. Blepharospasm
 - d. Leukocoria (white red reflex)
13. All but which one of the following assessments is used to determine the presence of a strabismus?
 - a. Hirschberg test
 - b. Cover-uncover test
 - c. Extraocular movements
 - d. Pupillary response
14. A 3-month-old has a mild asymmetrical corneal light reflex on physical exam. What is the next appropriate step?
 - a. Observe and reevaluate at the next well check
 - b. Refer immediately to ophthalmology
 - c. Begin atropine drops or eye patching
 - d. Protect eyes from sunlight
15. Prematurity increases the risk of developing which one of the following?
 - a. Nystagmus
 - b. Astigmatism
 - c. Myopia
 - d. Glaucoma
16. Fluorescein staining of the eye is used to detect a:
 - a. Keratitis
 - b. Foreign body
 - c. Corneal abrasion
 - d. Hyphema
17. Trauma to the eye increases the risk of developing all but which one of the following?
 - a. Strabismus
 - b. Glaucoma
 - c. Cataracts
 - d. Hyphema
18. Corneal abrasions can be managed with topical application of which one of the following?
 - a. Anesthetic for pain control
 - b. Steroids to prevent adhesions
 - c. Antibiotics to prevent infection
 - d. Atropine to prevent ciliary spasm
19. The greatest risk in a patient with a hyphema is which one of the following?
 - a. Glaucoma
 - b. Infection
 - c. Rebleed
 - d. Cataracts
20. A 16-year-old was hit in the eye 1 day ago and now has ecchymoses on the upper and lower lids with 5 out of 10 eye pain. All but which of the following would be appropriate to obtain at this time?
 - a. Visual acuity
 - b. Intraocular pressure
 - c. CT scan
 - d. Fluorescein stain
21. A 10-year-old has marked ear pain, not wanting anyone to touch his ear. The canal is edematous and exudate is present. TM is normal. How should this be managed?
 - a. Topical fluoroquinolone
 - b. Oral steroids and topical neomycin
 - c. Oral amoxicillin and topical anesthetic
 - d. Oral amoxicillin and topical steroid
22. Patients with otitis externa should be instructed to do which one of the following?
 - a. Keep ear dry until symptoms improve
 - b. Limit swimming for remainder of summer
 - c. Wear ear plugs at all times with swimming
 - d. Use alcohol drops before swimming each day
23. All but which one of the following patients are at an increased risk of developing otitis media?
 - a. 2-year-old with cleft palate repair at 1 year of age
 - b. 15-month-old with Down syndrome
 - c. 9-month-old with lactose intolerance
 - d. 3-year-old with IgA immune deficiency

24. A 15-month-old failed treatment with amoxicillin for an otitis media. At his 2-week recheck, his TM remained red with distorted landmarks and he persisted with nasal congestion, poor nighttime sleeping, and with a 101°F fever for the past 2 days. The next best step would be to treat with:
- A 10-day course of Augmentin
 - A 3-week course of a cephalosporin
 - A higher dose Amoxicillin and topical antibiotics
 - Ceftriaxone and an antihistamine
25. A 2-year-old male with a history of chronic serous otitis media is noted to have a pearly white opacity in the upper outer quadrant of his TM. He currently has no symptoms and appears to hear “okay.” The most likely diagnosis and appropriate management would be:
- Tympanosclerosis; no treatment is necessary
 - Persistent perforation; prescribe topical antibiotic drops
 - Foreign body; perform an ear wash for removal
 - Cholesteatoma; refer to otolaryngology
26. A 7-year-old has experienced recurrent nosebleeds in the past 2 months. What finding on the physical exam would suggest an underlying medical cause for the epistaxis?
- Wheezing
 - Grade II murmur
 - Petechiae
 - Tonsil hypertrophy
27. An 8-year-old has chronic intermittent nasal congestion. All but which one of the following would support allergic rhinitis?
- Pale boggy turbinates
 - Darkened areas on lower eyelids
 - Increased basophils on CBC
 - Itchy, watery eyes
28. Acceptable management options for allergic rhinitis include all of the following except:
- Oral Cetirizine
 - Oral Montelukast
 - Nasal beclomethasone
 - Nasal neosynephrine
29. Which foreign body in the nose needs immediate removal?
- Bean
 - Bead
 - Stone
 - Battery
30. What complication of sinusitis are adolescent males more prone to?
- Intracranial abscess
 - Potts puffy tumor
 - Orbital cellulitis
 - Dental infection
31. Patients with sinusitis should be instructed not to participate in what activity?
- Swimming/diving
 - Boxing/wrestling
 - Weight lifting
 - Cross country running
32. All of the following may predispose a patient to thrush except:
- Age
 - Steroid therapy
 - Antibiotics
 - Poor oral hygiene
33. A 9-month-old is noted to have a bifid uvula. This would increase his risk of developing which disorder?
- Otitis media
 - Retropharyngeal abscess
 - Sinusitis
 - Dental malocclusion
34. A 10-year-old has a single painful ulcerated lesion on an erythematous base on the inner buccal mucosa. The most likely diagnosis and treatment would be:
- Herpes simplex stomatitis—oral acyclovir
 - Herpangina—viscous xylocaine
 - Aphthous ulcer—triamcinalone in Orabase
 - Hand, foot, mouth syndrome—antibiotic mouthwash
35. Initial exposure to the herpes virus may produce all of the following except:
- Fever and dehydration
 - Submandibular lymph nodes
 - Vesicular lesions on tonsils
 - Friable and edematous gingivitis
36. The organism that causes hand, foot, mouth syndrome is what virus?
- Cytomegalovirus
 - Parainfluenza
 - Varicella-zoster
 - Coxsackie

37. Which one of the following complications of strep pharyngitis cannot be prevented with antibiotics?
- Peritonsillar abscess
 - Cervical adenitis
 - Glomerulonephritis
 - Acute rheumatic fever
38. In addition to penicillin, all of the following antibiotics can be used to treat strep pharyngitis except:
- Clindamycin
 - Erythromycin
 - Bactrim
 - Ceftriaxone
39. Retropharyngeal abscess is typically seen in what age group and mainstay treatment includes:
- Neonates; hospitalization for IV antibiotics
 - 2 to 6 year old; ICU admission and IV antibiotics
 - 6 to 12 year old; outpatient oral antibiotics
 - Adolescent; ENT drainage of abscess
40. Findings consistent of peritonsillar abscess include all of the following except:
- Muffled voice
 - Unilateral enlargement of tonsil
 - Trismus
 - Exudate on tonsils
41. Appropriate lab tests to obtain in assessment of cervical adenitis include all of the following except:
- Throat culture
 - Mono test
 - PPD test
 - Blood culture
42. The incidence of epiglottitis has decreased because of which vaccine?
- Hib
 - Pevnar
 - Varicella
 - Meningococcal
43. Patients with epiglottitis prefer to sit in what position?
- Sitting up and leaning forward
 - Left lateral position
 - Supine with neck hyperextended
 - 45-degree upright, resting back
44. All but which one of the following conditions requires urgent inpatient admission?
- Cervical adenitis
 - Retropharyngeal abscess
 - Epiglottitis
 - Orbital cellulitis
45. Conductive hearing loss can be caused by:
- Brain tumor
 - Ototoxic drug exposure
 - Loud noises
 - Serous otitis

■ ANSWERS

- | | |
|--------------|--------------|
| 1. b | 24. a |
| 2. d | 25. d |
| 3. a | 26. c |
| 4. c | 27. c |
| 5. a | 28. d |
| 6. d | 29. d |
| 7. a | 30. a |
| 8. b | 31. a |
| 9. d | 32. d |
| 10. a | 33. a |
| 11. b | 34. c |
| 12. d | 35. c |
| 13. d | 36. d |
| 14. a | 37. c |
| 15. c | 38. c |
| 16. c | 39. b |
| 17. a | 40. d |
| 18. c | 41. d |
| 19. c | 42. d |
| 20. b | 43. a |
| 21. a | 44. a |
| 22. a | 45. d |
| 23. c | |

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Cardiovascular Disorders

Peggy Dorr

■ CONGENITAL HEART DISEASE/ DEFECTS

- Definition: Cardiovascular malformations that result from abnormal structural development of the heart and/or vessels; most heart defects occur within first 8 weeks of gestation
- Etiology/Incidence
 1. Etiology of most defects unknown
 2. Probably genetic predisposition interacting with environmental trigger
 3. Chromosomal abnormalities account for nearly 10% of cardiac malformations (Down syndrome, Turner's syndrome, DiGeorge syndrome, others)
 4. Environmental or adverse maternal conditions account for 2% to 4%
 - a. Maternal diabetes mellitus, phenylketonuria, systemic lupus erythematosus, rubella, or other viruses
 - b. Maternal ingestion of thalidomide, alcohol, lithium, anticonvulsant agents, other drugs
 5. Approximately 8 per 1000 live births; about 32,000 new cases of congenital heart disease per year in the U.S.
 6. Ventricular septal defect (VSD) is most common (25% to 30% of all lesions)
- Signs and Symptoms
 1. Cyanosis—usually apparent at oxygen saturation of 85% or less
 - a. Central (arterial desaturation)—generalized, mucous membranes
 - b. Peripheral
 2. Increased respiratory rate and/or effort at rest or with activity
 3. Poor feeding; fatigue during feeding
 4. Excessive sweating in infant, unrelated to the environment, especially while feeding
 5. Recurrent respiratory infections
 6. Decreased exercise tolerance
 7. Syncope (uncommon)
- Differential Diagnosis
 1. Pulmonary disease
 2. Arrhythmias
 3. Myocardial diseases
 4. Rheumatic fever
 5. Sepsis
 6. Hypoglycemia, anemia, polycythemia, especially in neonates
 7. Central nervous system disorders
- Physical Findings
 1. Cyanosis (central or peripheral); pallor
 2. Poor growth/weight gain
 3. Abnormal respiratory patterns
 - a. Tachypnea
 - b. Hyperpnea
 - c. Dyspnea
 4. Tachycardia
 5. Hepatic enlargement with some defects
 6. Precordial prominence or increased precordial activity
 7. Palpable thrill

8. Abnormal heart sounds
 - a. Increased intensity
 - b. Abnormal splitting
 - c. Murmurs (may be absent or soft in spite of serious heart defect)
 - d. Ejection clicks
 - e. Fourth heart sound (gallop)
 9. Abnormal peripheral pulses
 - a. Decreased
 - b. Bounding (seen w/defects that increase blood volume to left heart, i.e., PDA)
 - c. Unequal (decreased lower extremity pulses suggest coarctation of the aorta)
 10. Abnormal blood pressure
 - a. Hypotension
 - b. Upper extremity blood pressure (systolic) greater than 10 mm Hg higher than lower extremity blood pressure suggestive of coarctation of aorta
 11. Peripheral edema, uncommon in infants
 12. Clubbing of the fingers or toes (with long-standing arterial desaturation)
- Diagnostic Tests/Findings
 1. Chest radiograph to evaluate heart size and pulmonary vascular markings
 2. Electrocardiogram to evaluate rhythm, chamber enlargement, or hypertrophy
 3. Arterial blood gas and hemoglobin in infant with cyanosis—if decreased PO₂ in room air, repeat arterial blood gas after 10 to 15 minutes in 100% inspired oxygen (oxygen challenge) to help differentiate between cardiac and pulmonary cyanosis; if minimal increase in PO₂, cardiac etiology suggested
 4. Echocardiogram for diagnosis of specific congenital heart defect; evaluate function, estimate right heart and pulmonary pressures
 5. Imaging studies (CT/CTA/MRI—helpful to evaluate pulmonary veins, coronary arteries, aortic arch abnormalities)
 6. Cardiac catheterization—gold standard for diagnosis
 - Management/Treatment
 1. Prompt referral to pediatric cardiologist or institution with pediatric cardiology and/or cardiothoracic surgical services
 - a. Symptomatic infant or child
 - b. Infant with cyanosis
 2. Initiation of prostaglandin infusion for cyanosis in the newborn, until CHD ruled out
 3. Monitoring and counseling to promote optimal growth and development
 4. Primary care, immunizations
 5. Endocarditis prophylaxis with dental or surgical procedures

6. Psychosocial support of child and family
7. Surgical management/treatment of most significant congenital heart defects—see **Table 5-1**

■ CONGESTIVE HEART FAILURE (CHF)

- Definition: Clinical syndrome that reflects the inability of the heart to meet metabolic requirements of the body; failure may initially be left- or right-sided, but if left untreated, the entire heart will fail
- Etiology/Incidence
 1. Congenital heart defects with volume or pressure overload (most common cause in pediatric age group)—see **Table 5-2**
 - a. Hypoplastic left heart syndrome
 - b. Coarctation of the aorta (obstruction to flow)
 - c. Ventricular septal defect (alteration in volume)
 - d. Atrioventricular septal defect (AV Canal)
 - e. Tetralogy of Fallot
 - f. Patent ductus arteriosus
 2. Acquired heart disease (less common cause)
 - a. Myocarditis
 - b. Metabolic abnormalities
 - c. Cardiomyopathy
 - d. Rheumatic heart disease
 3. Other causes
 - a. Tachyarrhythmias, complete heart block in infancy
 - b. Severe anemia, hydrops fetalis
 - c. Acute hypertension
 - d. Infection/sepsis
 - e. Endocrinopathies, renal failure
 - f. Arteriovenous malformations
 4. Incidence unknown as congestive heart failure is secondary to other disease processes
- Signs and Symptoms
 1. Increased respiratory rate and/or effort at rest or with activity
 2. Poor feeding
 3. Excessive sweating, especially with feeding in infants
 4. Decreased exercise tolerance
 5. Fatigue, persistently tired
 6. Poor weight gain
 7. Orthopnea in older child, chronic cough
- Differential Diagnosis
 1. Pulmonary diseases
 2. Cardiac diseases (see etiology)

■ **Table 5-1** Congenital Heart Defects Requiring Surgical Intervention

Transposition of the Great Arteries (cyanotic)
Anatomy: The aorta arises from right ventricle; pulmonary artery from the left ventricle; have PDA at birth; usually create/enlarge atrial septal opening (balloon atrial septostomy [BAS]) to allow for unrestricted mixing until surgery
Surgical Management: Arterial switch (Jatene) operation in neonatal period (within first 14 days of life optimal) or intra-atrial baffle (Mustard or Senning procedure) if anatomy doesn't allow for switch procedure (rare)
Tetralogy of Fallot (cyanotic)
Anatomy: Combination of four defects: (1) pulmonary stenosis, (2) ventricular septal defect, (3) overriding aorta, and (4) right ventricular hypertrophy
Surgical Management:
<ul style="list-style-type: none"> • <i>Palliative:</i> Blalock-Taussig (BT) shunt (subclavian artery to pulmonary artery) or other aortopulmonary shunts in infancy if necessary; infrequently performed today, most children undergo the complete repair only • <i>Repair:</i> Patch closure of VSD and resection of infundibular pulmonary stenosis ± pulmonary valvulotomy if needed
Atrioventricular Canal (aka AV canal, AV septal defect, Endocardial cushion defect)
Anatomy: Failure of central portion of heart to form resulting in a primum (low) ASD, high VSD, and abnormal mitral and tricuspid valves due to failure of endocardial cushion to develop appropriately
Surgical Management:
<i>Repair:</i> Patch closure of septal defects, repair of mitral valve to ensure competence (limit leakage without causing more damage to valve, trying to prevent need for replacement)
Coarctation of the Aorta (acyanotic)
Anatomy: Congenital narrowing of the aorta, usually distal to the origin of the left subclavian artery, opposite the area of the ductus arteriosus
Surgical Management:
<i>Repair:</i> Performed via thoracotomy, several types of repair possible; typically "end-to-end anastomosis" where narrowed area is cut out and ends of aorta are sewn back together, or a "subclavian flap repair" where the subclavian artery is isolated and opened longitudinally and extended over narrowed portion of aorta
Total Anomalous Pulmonary Venous Connection
Anatomy: Pulmonary veins do not enter left atrium but are connected either directly or indirectly to right atrium; if veins return below the diaphragm, emergency surgery is usually required
Surgical Management:
<i>Repair:</i> Pulmonary venous confluence connected to left atrium in infancy
Truncus Arteriosus
Anatomy: Single arterial trunk that didn't divide completely in utero; single vessel provides blood flow to both pulmonary and systemic circulation, overrides ventricles and receives blood from them through a ventricular septal defect
Surgical Management:
<i>Repair:</i> Closure of ventricular septal defect, removing origin of the pulmonary arteries from trunk, and connecting pulmonary arteries to right ventricle with a conduit in infancy; the truncus portion becomes the aorta
Single Ventricle Physiology (secondary to Pulmonary Atresia with Intact Ventricular Septum, Tricuspid Atresia, Hypoplastic Left or Right Heart Syndrome, etc.)
Anatomy: Abnormal cardiac anatomy and development results in only one functioning ventricle; anatomical basis for this are numerous but the repair is essentially the same
Surgical Management:
<i>Repair:</i> Repair is broken out into three stages. The first stage is a shunt or conduit that provides consistent pulmonary blood flow and arch augmentation if necessary. This is the Norwood procedure and either a BT shunt or an RV-PA conduit is placed.
The second stage is a bidirectional Glenn which involves removing the SVC from the right atrium and attaching it to the branch pulmonary artery. The final stage is the Fontan, which involves directing IVC blood flow directly to the branch pulmonary artery. The goal of these surgeries is to allow venous return to flow to the lungs passively and use the single ventricle as the systemic ventricle.

■ **Table 5-2** Heart Defects Commonly Causing Congestive Heart Failure in Infancy

Defect	Etiology of Heart Failure
Hypoplastic left heart syndrome (any single ventricle physiology)	Cardiac output must be balanced between pulmonary and systemic circulation. Alterations in metabolic status (alkalosis, hyperoxygenation) will cause increased blood flow to lungs
Coarctation of the aorta	Severe coarctation will cause obstruction to left ventricular output
Ventricular septal defect	Abnormal opening between right and left ventricle that may result in congestive heart failure if left-to-right shunt is large, allowing excess blood flow to pulmonary system
Atrioventricular septal defect (AV canal, endocardial cushion defect)	Defect in lower part of atrial septum (ostium primum) and Ventricular septum allows excessive pulmonary blood flow, which may increase pulmonary pressures and resistance over time
Patent ductus arteriosus (PDA)	If large enough, the excess flow of blood through the PDA to the pulmonary system would alter the pulmonary pressures and resistance over time

- Physical Findings
 1. Tachycardia (common)
 2. Tachypnea (common)
 3. Hepatomegaly (right-sided failure)
 4. Puffy eyelids common in infants, peripheral edema less common in children
 5. Wheezing, pulmonary rales may be present (with left-sided failure)
 6. Pallor, mottling of extremities
 7. Weakly palpable peripheral pulses; cool extremities due to peripheral vasoconstriction
 8. Gallop rhythm with myocardial failure
 9. Cyanosis with alveolar edema
- Diagnostic Tests/Findings
 1. Chest radiograph—cardiomegaly almost always present; pulmonary vascular congestion dependent on etiology
 2. Echocardiogram to assess ventricular function, chamber enlargement, anatomy
 3. Electrocardiogram not diagnostic, may help define etiology
- Management/Treatment
 1. Referral to cardiologist to determine etiology if heart disease suspected
 2. Increase oxygen supply (supplemental oxygen, correct anemia)
 3. Drug therapy
 - a. Inotropic agents—usually digoxin
 - b. Diuretics
 - c. Afterload-reducing agents (ACE inhibitors such as captopril for systemic afterload reduction, sildenafil for pulmonary afterload reduction)
 4. Other measures
 - a. Semi-Fowler position (infant seat, pillows)
 - b. Prostaglandin E₁ if systemic perfusion dependent on patency of ductus arteriosus
 - c. Caloric supplementation of formula, breast milk fortifier (low sodium formulas not recommended)
 - d. Frequent rest periods
 5. Treatment of underlying condition if specific therapy available
 6. Possible referral for cardiac transplantation if refractory, end-stage heart failure

■ MURMURS

- Definition: A murmur is the result of turbulent blood flow through an abnormal or obstructed area; evaluated based on timing (systolic vs diastolic), intensity (I–VI scale), location, radiation, pitch, and quality; can be innocent or pathologic
 1. Innocent murmurs are murmurs not associated with any anatomic abnormality; result from turbulence of blood flow
 2. Innocent murmurs are also referred to as functional, normal, benign, or insignificant

3. Common innocent murmurs
 - a. Still's murmur—vibratory, groaning, or musical systolic murmur heard best between lower-left sternal border and apex; attributed to turbulence in left ventricular outflow tract or vibration of the tendonae in the ventricles
 - b. Pulmonary or aortic flow murmur—slightly harsh systolic ejection murmur heard best at second to third left intercostal space; attributed to turbulent flow in right or left ventricular outflow tract
 - c. Physiologic peripheral pulmonary stenosis—low intensity systolic ejection murmur heard best at upper left sternal border, axillae, and back in neonates until 3- to 6-months of age; attributed to relative hypoplasia of branch pulmonary arteries at birth and anatomy of left pulmonary artery
 - d. Venous hum—humming continuous murmur usually heard best at upper right sternal border in sitting position with marked decrease of murmur with change in head position (turn head sideways) or disappearance of murmur in supine position; result of blood flow returning through the SVC to the heart
 - e. Supraclavicular arterial bruit—early systolic murmur heard above clavicles, attributed to turbulence at site of branching of brachiocephalic arteries
 4. Pathologic murmurs are related to anatomical abnormalities within the heart or great vessels
 - a. Diastolic murmurs are always pathologic
 - b. Pathologic murmurs related to heart disease do not change with alterations in position
 - c. For a murmur to be graded IV/VI, there must be a palpable thrill
- Etiology/Incidence of innocent murmurs
 1. Heard in more than 50% of normal children from infancy through adolescence
 2. Still's murmur most common; heard most frequently from 3 to 7 years of age
 3. Increased intensity (incidence) associated with increased cardiac output state (fever, acute illness, anemia, anxiety, exercise); best to reevaluate murmur after illness resolved; frequently murmur is not audible at that time
 - Signs and Symptoms: Asymptomatic if innocent; if pathologic, symptoms related to underlying disease process or defect
 - Differential Diagnosis
 1. Congenital heart disease
 2. Conditions associated with high cardiac output
 - Physical Findings of Innocent Murmurs
 1. No cyanosis or other cardiovascular abnormalities
 2. Normal blood pressure, peripheral pulses in upper and lower extremities
 3. Normal heart sounds, including normal splitting (not fixed) of second heart sound
 4. No clicks
 5. Characteristics
 - a. Usually systolic with exception of venous hum; never diastolic alone
 - b. Usually low intensity (grade 1 to 3); classic musical or “twanging” quality
 - c. Usually short duration, not holosystolic; never associated with precordial thrill
 - d. Well-localized, poorly transmitted except neonatal peripheral pulmonary stenosis (heard at left upper sternal border, axillae, and back)
 - e. Intensity typically varies with position change
 - Diagnostic Tests/Findings
 1. Testing not routine
 2. May be indicated to rule out congenital heart defect
 - a. Electrocardiogram normal
 - b. Echocardiogram if recommended by cardiologist
 - Management/Treatment
 1. Inform parents of murmur
 2. Provide reassurance that child's heart is normal
 3. Inform them that murmur may come and go; may sound louder if child is sick
 4. Child does NOT require SBE prophylaxis
 5. Referral to cardiologist if:
 - a. Symptomatic
 - b. Cardiovascular abnormalities on physical examination
 - c. Uncertainty regarding innocence of murmur; change in murmur intensity
 - d. Persistent parental concern
- DISTURBANCES OF RATE AND RHYTHM**
- Definition: Occur as a result of abnormalities in, or insults to, the cardiac conduction system or heart tissues

- Etiology/Incidence
 1. May be congenital or acquired
 2. Bradyarrhythmias and heart block are uncommon causes of ECG abnormalities in children without congenital heart disease
 - a. Bradycardia—a heart rate below the lower limits of normal for age
 - (1) Athletes frequently have heart rates below normal due to conditioning of heart muscle
 - (2) Symptomatic bradycardia is associated with symptoms of poor perfusion and requires immediate attention
 - (3) Most common prearrest rhythm
 3. Tachyarrhythmias cause nonspecific signs and symptoms if they are extreme rates or persist for an extended period; type and extent of symptoms vary with age
 - a. Tachycardia—a heart rate above the upper limits of normal for age
 - (1) May be normal response to fever or stress
 - (2) Supraventricular tachycardia (SVT) is the most common symptomatic tachycardia in first year of life
 - (3) Supraventricular tachycardia (SVT) is defined as an HR greater than 220 in infants and greater than 180 in children; narrow QRS complex with hidden P waves
 - (4) SVT can be associated with Wolff-Parkinson-White (WPW) syndrome (narrow PR interval, slurring of initial segment of QRS complex creating the “delta wave,” wide QRS complex)
 4. Atrioventricular block—a disturbance of impulse conduction from the atria to the ventricles
 - a. Congenital complete heart block (CHB); 1 in 25,000 live births; associated with maternal systemic lupus erythematosus or other connective tissue diseases
 - b. Acquired heart block caused by damage to conduction system during cardiac surgery; severe myocarditis, acute rheumatic fever, mumps, tumors in conduction system, endocrine/metabolic disorders
 5. Isolated premature atrial contractions (PAC) and premature ventricular contractions (PVC) common in healthy children
 6. Prolonged QT Syndrome—defined as a QT_c interval greater than 0.45 secs
 - a. The QT interval measures the duration of activation and recovery of the ventricles; if this is prolonged, the patient is at risk for early depolarization of ventricles that are not electrically ready and a potentially fatal dysrhythmia could result
 - b. Must correct the QT interval for heart rate; the formula to calculate the QT_c is the QT interval (in secs) / square root of the R-R interval (secs)
 - c. May be hereditary; look for family history of sudden death; there are blood tests that may help confirm diagnosis
 - d. Syncope may be the presenting symptom; all syncope patients should get an EKG
 - 7. Other causes for dysrhythmias
 - a. Drugs (beta agonists, cocaine, antipsychotics, many others)
 - b. Electrolyte imbalance
 - c. Acidosis/hypoxia
 - d. Increased intracranial pressure
 - e. Endocrine disorders (hyper/hypothyroidism)
 - f. Cardiomyopathy, structural heart disease, or surgery
- Signs and Symptoms
 1. Children usually asymptomatic
 2. Symptoms of low cardiac output or congestive heart failure if bradycardia or tachycardia is severe and/or prolonged
 3. Irritability, pallor, poor feeding in infants
 4. Palpitations, syncope, dizziness, chest pain
 5. Seizures
 6. Rare sudden death (with ventricular tachycardia and fibrillation)
 - Differential Diagnosis
 1. Sinus arrhythmia (phasic acceleration and deceleration of heart rate with respiration; normal in children)
 2. Sinus tachycardia and associated causes
 3. Sinus bradycardia and associated causes (normal in athletes)
 4. Conditions associated with heart rate disturbances (see etiology)
 - Physical Findings
 1. Bradycardia, tachycardia, or irregular rhythm
 2. Tachypnea, hepatomegaly, poor perfusion, especially in infants if rate disturbance severe and/or prolonged
 - Diagnostic Tests/Findings
 1. Electrocardiogram
 2. Other tests based on clinical findings and symptoms

- Management/Treatment
 1. Referral to cardiologist for evaluation and treatment if:
 - a. Symptomatic
 - b. Sustained dysrhythmia
 - c. Recurrent dysrhythmia
 2. Treatment of underlying condition if noncardiac cause

■ HYPERTENSION

- Definition: “Average systolic blood pressure (SBP) and/or average diastolic blood pressure (DBP) greater than or equal to the 95th percentile for gender, age, and height, with measurements obtained on at least three occasions” (National High Blood Pressure Education Program [NHBPEP] Working Group on Children and Adolescents)
 1. Prehypertension: Defined as average SBP or DBP levels that are greater than/equal to the 90th percentile but less than the 95th percentile
 2. Children with BP levels greater than 95th percentile in an office or clinic setting that are normotensive outside a clinical setting (“white coat syndrome”) should be followed using their ambulatory BP readings
 3. Children greater than 3 years old should have their BP measured routinely.
 4. Preferred method of BP measurement is auscultation; measures obtained by oscillometric devices (automatic blood pressure machines) should be repeated by auscultation if abnormal
 5. MUST HAVE a correct size BP cuff to obtain an accurate measurement; width of the cuff bladder should be 40% of the arm circumference; if an appropriate cuff size is not available, the next larger size is used
- Etiology/Incidence
 1. Primary hypertension
 - a. No known underlying disease present to cause hypertension
 - b. May be related to factors such as heredity, salt intake, stress, sleep disorders/apnea, and obesity
 - c. May be recognized in childhood; it is characterized by mild hypertension and is often associated with a positive family history of hypertension or cardiovascular disease; these children are frequently overweight
 2. Secondary hypertension—more common in children than adults
 - a. Etiology varies with age
 - (1) Newborn infants—renal artery thrombosis, renal artery stenosis, congenital renal malformations, coarctation of the aorta, bronchopulmonary dysplasia
 - (2) Infancy to 6 years—renal parenchymal diseases, coarctation of the aorta, renal artery stenosis
 - (3) 6- to 10-year-olds—renal artery stenosis, renal parenchymal disease
 - b. Other causes—endocrine disorders, neurogenic tumors, drugs or toxins, increased intracranial pressure
- Prevalence of hypertension in childhood is low (1% to 3%)
- Signs and Symptoms
 1. Primary hypertension—usually no symptoms
 2. Secondary hypertension—symptoms related to underlying disease and may include:
 - a. Congestive heart failure
 - b. Respiratory distress (infants)
 - c. Failure to thrive
 - d. Irritability
 - e. Convulsions
 - f. Feeding problems
 3. Severe hypertension—may complain of headaches, dizziness, visual disturbances, epistaxis (rare); infants may show irritability, failure to thrive, vomiting, or feeding problems
- Differential Diagnosis: Causes of secondary hypertension (see etiology)
- Physical Findings
 1. Elevated blood pressure on at least three occasions (without acute illness)
 2. Related to underlying disease if secondary hypertension—may include edema, pallor, increased sweating, signs of specific syndrome including absent or diminished femoral pulses seen with coarctation of the aorta
- Diagnostic Tests/Findings
 1. Based on history, age, other clinical findings
 2. Complete blood count (to evaluate for anemia)
 3. Urinalysis
 4. BUN, creatinine (to evaluate renal function)
 5. Lipid profile (evaluated as a comorbidity risk factor)
 6. Echocardiogram
 - a. For baseline measurements of left ventricular dimensions and function
 - b. To rule out coarctation of the aorta
 - c. To evaluate for left ventricular hypertrophy as an indicator of end organ damage
 - d. Prior to initiation of drug therapy
 7. Additional tests based on suspected cause

- Management/Treatment
 1. Depends on cause and degree of elevation
 2. General counseling regarding cardiovascular risk factors such as family history, obesity, lack of exercise, smoking
 3. Nonpharmacologic therapy
 - a. Weight reduction if indicated
 - b. Regular aerobic exercise programs (especially in adolescents)
 - c. Dietary modification—sodium reduction
 4. Antihypertensive drug therapy indicated for secondary hypertension and insufficient response to lifestyle modifications for primary hypertension
 - a. Therapy should be initiated with a single drug; acceptable drug classes for use in children include:
 - (1) ACE inhibitors (captopril, enalapril, lisinopril)
 - (2) Angiotensin receptor blockers (losarten)
 - (3) Beta blockers (atenolol, metoprolol)
 - (4) Calcium channel blockers (amlodipine, isradipine, nifedipine)
 - (5) Diuretics (HCTZ, furosemide, spiranolactone)
 - b. Goal of therapy should be reduction of blood pressure to less than 95th percentile
 - c. Severe, symptomatic hypertension should be treated with IV antihypertensive agents in an acute care setting until controlled, then transition patient to PO medications
 5. If secondary hypertension, therapy for underlying disease
 6. Long-term follow-up: referral to subspecialist (cardiologist, nephrologist) for management of underlying disorder and/or for difficulty maintaining normotensive state

■ HYPERCHOLESTEROLEMIA

- Definition: Elevated serum cholesterol level
 1. Total cholesterol of 200 mg/dL or above
 2. Total cholesterol of 170 to 199 mg/dL considered “borderline high”
 3. Low density lipoprotein (LDL) cholesterol of 130 mg/dL or above
- Etiology/Incidence
 1. High tracking correlations for total and LDL cholesterol levels from childhood to adulthood
 2. Potential increased risk for developing atherosclerotic or coronary heart disease as adults, especially in association with other risk factors
 - a. Positive family history of premature coronary heart disease (before 55 years

- of age), peripheral vascular disease, or hypercholesterolemia (above 240 mg/dL)
 - b. Diabetes
 - c. Hypertension
 - d. Obesity
 - e. Smoking
 - f. Physical inactivity
- 3. Among children aged 2 to 11 years in U.S., about 25% with cholesterol levels in borderline high or high range
- 4. Familial hypercholesterolemia is an autosomal dominant condition resulting in deficient or defective LDL receptors, impairing clearance of circulating LDL
- 5. Secondary causes of hypercholesterolemia
 - a. Obesity
 - b. Endocrine and metabolic conditions
 - c. Obstructive liver disease
 - d. Nephrotic syndrome
 - e. Anorexia nervosa
 - f. Collagen disease
 - g. Drugs
 - (1) Corticosteroids
 - (2) Isotretinoin (Accutane)—increases triglycerides
 - (3) Thiazide diuretics
 - (4) Some beta blockers
 - (5) Some oral contraceptives

- Signs and Symptoms: Asymptomatic during childhood
- Differential Diagnosis: Causes of secondary hypercholesterolemia (see etiology)
- Physical Findings
 1. Rare xanthomas with familial hypercholesterolemia and very high cholesterol levels
 2. Signs of risk factors (hypertension, obesity)—The metabolic syndrome is a clustering of risk factors (abnormal waist circumference, lipid levels, BP, and fasting glucose level) for cardiovascular disease and diabetes mellitus that seem to be more prevalent in overweight children and adolescents. “Pathology studies have clearly shown that the presence of an increasing number of risk factors (as seen in Metabolic Syndrome) is associated with increased risk of fatty streaks and fibrous plaques in the aorta and coronary arteries.”
- Diagnostic Tests/Findings
 1. Measurement of total cholesterol in children over 2 years of age with a positive family history or other risk factors; cholesterol

concentrations change with age in children and are particularly variable during puberty

- a. If total cholesterol level acceptable (< 170 mg/dL), repeat measurement within 5 years
 - b. If total cholesterol level borderline (170 to 199 mg/dL), repeat measurement, and if average is borderline or high, do lipoprotein analysis
 - c. If total cholesterol level high (above 200 mg/dL), do lipoprotein analysis
2. American Heart Association recommends the following evaluation standards:
 - a. LDL cholesterol > 130 mg/dL is abnormal
 - b. HDL cholesterol < 35 mg/dL is abnormal
 - c. Triglycerides > 150 mg/dL is abnormal
 3. Additional studies to evaluate causes of secondary hypercholesterolemia, as indicated
- Management/Treatment
 1. Trans fatty acid intake is limited to less than 1% of total calories; these fats tend to increase LDL and do not raise HDL
 2. Intake of fruit juice, sugar sweetened beverages and food, and salt needs to be reduced
 3. In general, children need to consume more fruits, vegetables, fish, whole grains and low-fat dairy products
 4. Dietary therapy guidelines for children over 2 years of age:
 - a. If LDL cholesterol ≥ 110 mg/dL, step 1 diet—no more than 30% of total calories from fat, less than 10% of total calories as saturated fat, and less than 300 mg of cholesterol per day; adequate calories to reach or maintain desirable body weight.
 - b. If LDL cholesterol > 130 mg/dL after 3 months of step 1 diet, step 2 diet is prescribed—saturated fatty acid intake reduced to less than 7% of calories and cholesterol intake to less than 200 mg per day; adequate amounts of nutrients, vitamins, and minerals provided
 - c. Fat and cholesterol restricted diets have been well studied and are safe, but frequently result in only modest improvements in hyperlipidemia
 5. Drug therapy in children aged 10 years and older if after an adequate trial of diet therapy (6 months to 1 year), LDL cholesterol ≥ 190 mg/dL or LDL cholesterol ≥ 160 mg/dL PLUS a positive family history of premature cardiovascular disease OR 2 or more other cardiovascular risk factors are present in the child
 - a. HMG CoA reductase inhibitors (Statins)—work by inhibiting the rate-limiting enzyme HMG CoA reductase for the

endogenous synthesis of cholesterol; ultimately leads to increased clearance of LDL from circulation; pediatric studies have been done; has demonstrated excellent ability to lower LDL with few side effects

- b. Bile acid binding resins—previously advocated as first line treatment but high incidence of GI complaints; poor palatability; low compliance and limited effectiveness have lead to these drugs not being prescribed as first-line therapy
 - c. Cholesterol absorption inhibitors—new class preventing the intestinal absorption of cholesterol; used in conjunction with statins for severe hyperlipidemia; pediatric trials currently in progress
6. Increase exercise—regular physical activity is the best approach; data supports exercise as a factor that has a favorable impact on multiple lipoprotein aspects
 7. Prevention—anticipatory guidance during all well-child maintenance visits to maintain well-balanced diet, encourage daily exercise, and avoid known risk factors

▣ KAWASAKI DISEASE (MUCOCUTANEOUS LYMPH NODE SYNDROME)

- Definition: Acute febrile syndrome associated with generalized vasculitis affecting all blood vessels throughout the body, preferentially involving the coronary arteries
- Etiology/Incidence
 1. Etiology uncertain, but epidemiology and clinical presentation highly suggestive of infectious etiology
 2. Most frequently (80%) affects infants and children under 5 years of age; leading cause of acquired heart disease in U.S.
 3. Male to female ratio 1.5:1
 4. All racial backgrounds affected; highest incidence in children of Asian ancestry
 5. Approximately 20% risk of developing coronary artery abnormalities with decreased risk if intravenous gamma globulin therapy instituted before 10th day of illness
- Signs and Symptoms
 1. Vary in severity and over course of illness
 2. Acute phase—lasts about 10 days; most of diagnostic criteria are noted at some point during this time period
 - a. Preceding or concurrent respiratory symptoms—runny nose, cough, ear infection

- b. Diarrhea, vomiting, or abdominal pain (common)
 - c. Irritability
 - d. Persistent high fever > 5 days
 - e. Reddened eyes
 - f. Red tongue and throat
 - g. Redness and/or swelling of hands and feet
 - h. Rash
 - i. Swollen lymph nodes
 - j. Fast and/or irregular pulse
 - k. Arthritis or arthralgia involving multiple joints
3. Subacute phase—(lasts from approximately day 11–21)
 - a. Decrease in fever
 - b. Skin desquamation (dry, peeling skin on lips, fingers, and toes) begins about day 14; not painful
 - c. Joint pain
 4. Convalescent phase—begins about day 21
 - a. coronary artery aneurysms are often detected by echocardiography at this time
 - b. Acute phase reactants subside
- Differential Diagnosis
 1. Measles
 2. Scarlet fever
 3. Toxin mediated disease related to staphylococcal or streptococcal disease
 4. Drug reactions
 5. Juvenile rheumatoid arthritis
 6. Other febrile viral illness (adenovirus, Epstein-Barr, enterovirus)
 7. Stevens-Johnson syndrome
 8. Rocky Mountain spotted fever
 9. Staphylococcal scalded skin syndrome
 10. Toxic shock syndrome
 - Physical Findings
 1. Diagnostic criteria includes presence of:
 - a. Fever (typically high > 39°C/102°F) for at least 5 days AND four of the five following clinical features:
 - b. Bilateral, painless bulbar conjunctival injection without exudate
 - c. Changes of mucous membrane—erythema, dryness and cracking of lips, strawberry tongue, erythema of oropharyngeal mucosa
 - d. Changes to extremities—acute erythema and/or edema of hands/feet
 - e. Polymorphous nonvesicular exanthem within 4 to 5 days of fever onset
 - f. Cervical nonfluctuant lymphadenopathy; at least one lymph node more than 1.5 cm in diameter; usually unilateral (least common symptom)
 2. Coronary artery abnormalities (usually beyond 10 days of illness onset) with fever and fewer than 5 clinical features is diagnostic
 3. Other physical findings
 - a. Arthritis or arthralgia
 - b. Tachycardia out of proportion to degree of fever; gallop rhythm (signs of myocarditis); new murmur (less common)
 - c. Sterile pyuria
 - d. Aneurysm of peripheral arteries (less common)
 - Diagnostic Tests/Findings
 1. No specific diagnostic test, “diagnosis of exclusion”
 2. Thrombocytosis, may be marked; frequently seen after first week of illness (subacute phase—peaks in 3rd week)
 3. Leukocytosis with left shift
 4. Erythrocyte sedimentation rate elevated
 5. C-reactive protein positive
 6. Mild anemia in acute phase
 7. Hypoalbuminemia (associated with more severe, more prolonged disease)
 8. Electrocardiogram changes are nonspecific and related to secondary myocarditis; may see ST-T wave changes and prolonged PR interval
 9. Echocardiogram—coronary artery dilation, aneurysms, or ectasia; occasional pericardial effusion or decreased contractility; coronary artery dilation may be present as early as the end of the first week of illness
 - Management/Treatment
 1. Immediate referral to tertiary care facility and subspecialist (immunologist, pediatric cardiologist) if suspected
 2. Management goals
 - a. Reducing inflammation in the coronary arterial wall
 - b. Preventing coronary thrombosis
 - c. Long-term therapy for individuals who develop coronary aneurysms is aimed at preventing myocardial ischemia or infarction
 3. Therapy during acute phase
 - a. Intravenous gamma globulin (2 g/kg IV) within 10 days of onset of illness (shown to reduce frequency of coronary aneurysms)
- PLUS**
- b. High-dose aspirin (80 to 100 mg/kg/day) for its anti-inflammatory and antithrombotic effects until patient is afebrile; then reduced to 3 to 5 mg/kg/day for 6 to 8 weeks
 - c. If fever persists > 36 hours after 1st IVIG dose, may repeat same dose

- d. Studies on-going exploring the use of steroids as therapy; usefulness not well established
- 4. If coronary arterial abnormalities detected
 - a. Low-dose aspirin continued indefinitely
 - b. Anticoagulation Therapy—considered if high risk for myocardial infarction
 - (1) Dipyridamole (Plavix)
 - (2) Warfarin
 - c. Long-term follow-up by pediatric cardiologist; may require cardiac catheterization or surgical intervention
- 5. Administration of live virus vaccines (measles and varicella) delayed at least 11 months after intravenous gamma globulin treatment unless risk of exposure is high; may give earlier, but then be reimmunized at 11 months after IVIG administration
- 6. Administration of annual influenza vaccine in patients on long-term aspirin therapy
- 7. Activity restrictions beyond initial 6 to 8 weeks based on severity of coronary artery involvement

■ ACUTE MYOCARDIAL INFLAMMATORY DISEASE

- Definition: Focal or diffuse inflammation of the layers of the heart (pericardium, myocardium, or endocardium)
- Etiology/Incidence
 - 1. Myocarditis and Pericarditis
 - a. Precise etiology usually unknown (idiopathic)
 - b. May be caused by virtually any bacterial, viral, rickettsial, fungal, or parasitic organism—viral infections the most common etiology for both, especially coxsackie B virus (> 50% of myocarditis) and adenovirus
 - 2. Other causes include autoimmune or collagen-vascular diseases, or hypersensitivity drug reactions (rare)
 - 3. Possible genetic predisposition with viral trigger
 - 4. Pericarditis can be postsurgical, post trauma, or associated with cancers.
 - 5. Incidence unknown, as many mild cases may go undetected
 - 6. In patients with myocarditis who present with severe congestive heart failure or shock (20% to 30%), approximately $\frac{1}{3}$ will die or require transplantation
- Signs and Symptoms
 - 1. Great variability—from no distress to severe congestive heart failure or shock
 - 2. Often fever or history of antecedent “flu-like” viral illness for both
 - 3. Suspect myocarditis if onset of congestive heart failure with no obvious structural or functional etiology
 - a. Persistent tachycardia (out of proportion to fever if present)
 - b. Tachypnea, dyspnea
 - c. Easy fatigue, poor feeding in infant
 - d. Gallop rhythm, usually no murmur
 - e. Hepatomegaly
 - f. Poor perfusion
 - 4. Can also present as new onset arrhythmias, syncope, or sudden death
 - 5. Pericarditis presents with usual symptoms of illness (fever, irritability) and:
 - a. chest pain; varies with respirations and position
 - b. pericardial friction rub
 - c. muffled heart sounds possible if effusion is significant
- Differential Diagnosis
 - 1. Carnitine deficiency
 - 2. Idiopathic dilated cardiomyopathy (myocarditis accounts for up to 1/3 of dilated cardiomyopathy cases)
 - 3. Endocardial fibroelastosis
 - 4. Hereditary mitochondrial defects
 - 5. Anomalies of the coronary arteries
 - 6. Metabolic or endocrine disorders (hyperthyroidism)
- Physical Findings: See section on congestive heart failure
- Diagnostic Tests/Findings
 - 1. Erythrocyte sedimentation rate and heart enzymes (CPK, LDH) elevated
 - 2. Viral cultures, antibody titers, and polymerase chain reaction (PCR) may suggest viral etiology
 - 3. Laboratory studies to rule out metabolic causes of cardiomyopathy
 - 4. Echocardiography to assess ventricular function and rule out other cardiac anomalies; document the presence and amount of pericardial fluid
 - 5. Chest radiograph to assess cardiac enlargement which is variable
 - 6. Electrocardiogram may show tachycardia, ST segment and T wave abnormalities, possibly reduced QRS voltage, or dysrhythmias
 - 7. Endomyocardial biopsy for possible confirmation of diagnosis
 - 8. Pericardial fluid sent for cultures, cell count, and differential

- Management/Treatment
 1. Immediate referral to pediatric cardiologist if either disease is suspected, and tertiary-care facility if necessary for acute care
 2. Supportive measures for congestive heart failure
 - a. Inotropic agents, including digoxin, at reduced dosage (may be arrhythmogenic)
 - b. Diuretics
 - c. Afterload reduction (ACE inhibitors such as captopril, lisinopril)
 3. Treatment of dysrhythmias
 4. Use of immunosuppressive medication in myocarditis is supported with some research, but no large, randomized controlled study at this time
 5. Possible administration of intravenous immunoglobulin—effectiveness not proven; risk of anaphylaxis
 6. Pericarditis treatment depends on type of pathogen; drainage of pericardial fluid improves survival of patients with purulent pericarditis
 7. Psychosocial support of child and/or family—usually sudden onset of illness in previously healthy child
 8. Outcome from myocarditis varies from complete resolution and recovery to development of chronic cardiomyopathy or death without cardiac transplantation
 9. Patient's status post pericarditis needs to be monitored for several weeks for reaccumulation of pericardial fluid and long-term for dysrhythmias and constrictive pericarditis

■ RHEUMATIC FEVER/HEART DISEASE

- Definition: A post-infectious inflammatory disease in genetically predisposed individuals to group A β -hemolytic streptococcal pharyngitis. It is a self-limited disease that is diagnosed using the Jones criteria.
- Etiology/Incidence
 1. Follows a group A streptococcal infection of the upper respiratory tract
 2. Probably involves abnormal immune response of certain individuals with genetic predisposition to this complication
 3. Incidence of acute rheumatic fever (ARF) approximately 3% of individuals with untreated or inadequately treated group A streptococcal tonsillopharyngitis (greater risk of recurrence)
 4. Most common in children between 5 and 15 years of age; rarely seen before 3 years in the U.S.; rare in adults

5. Seasonal incidence follows that of streptococcal pharyngitis; peak incidence in winter and early spring
6. Greater frequency of rheumatic heart disease in patients who had severe cardiac involvement during initial attack or recurrence of ARF
 - a. Mitral valve damage and regurgitation most common
 - b. Tricuspid and pulmonary valve involvement rare

- Signs and Symptoms

1. Diagnosis of the initial attack of acute rheumatic fever based on evidence of preceding group A streptococcal pharyngitis PLUS two major manifestations OR one major and two minor manifestations of the following:
2. Major Manifestations
 - a. Carditis (approximately 50% of patients)
 - (1) Valvulitis usually—evidenced by development of new murmur, especially apical systolic murmur of mitral regurgitation
 - (2) Myocarditis rare in absence of valvulitis—evidenced by tachycardia; other signs of congestive heart failure
 - (3) Pericarditis rare in absence of valvulitis—evidenced by distant heart sounds, friction rub, chest pain
 - b. Polyarthrititis (approximately 70% of patients)
 - (1) Several joints may be intermittently involved ranging from vague arthralgia to florid swelling, heat, and redness
 - (2) Most frequently larger joints—knees, hips, ankles, elbows, wrists
 - c. Chorea (Sydenham's chorea)
 - (1) Purposeless, involuntary, rapid movements of trunk and/or extremities
 - (2) Often associated with muscle weakness and emotional lability
 - d. Erythema marginatum (rare)
 - (1) Macular, nonpruritic rash with irregular, geometric morphology; areas have pale centers and rounded margins
 - (2) Lesions most commonly located on trunk and proximal limbs, never on face
 - e. Subcutaneous nodules (rare)
 - (1) Firm, painless nodules over the extensor surfaces of certain joints, particularly elbows, knuckles, knees, ankles, occiput, and vertebrae
 - (2) Skin overlying nodules is not inflamed and moves freely

3. Minor Manifestations
 - a. Arthralgia without objective evidence of inflammation
 - b. Fever usually at least 39°C (102.2°F)
 - c. Elevated acute phase reactants
 - d. Prolonged PR interval on electrocardiogram
- Differential Diagnosis
 1. Juvenile rheumatoid arthritis
 2. Myocardial disease (NOTE: there is no rheumatic heart disease without valvular involvement)
 3. Infective endocarditis
 4. Septic arthritis
 5. Sickle cell disease
- Physical Findings: No single specific finding; see manifestations
- Diagnostic Tests/Findings
 1. No specific diagnostic test
 2. Elevated acute phase reactants
 - a. Erythrocyte sedimentation rate
 - b. C-reactive protein
 - c. Leukocyte count
 3. Elevated or rising streptococcal antibody titer (ASO)
 4. Electrocardiogram may indicate prolonged PR interval (1st degree heart block)
 5. Echocardiogram may show mitral valve damage resulting in valvular regurgitation and/or mitral valve stenosis
- Management/Treatment
 1. Referral to cardiologist if suspected
2. Antibiotic treatment of group A streptococcal infection—Benzathine Penicillin G IM or Penicillin V PO (started even as late as 9 days after acute onset has demonstrated effectiveness)
3. Anti-inflammatory agents for treatment of arthritis and discomfort
 - a. Salicylates or nonsteroidal agents
 - b. Possible steroids if congestive heart failure and/or carditis severe
4. Bed rest for acute carditis not recommended anymore, although some activity restrictions during acute phase may be appropriate (competitive sports)
5. Treatment of congestive heart failure, if present
6. Prevention of further streptococcal infection and recurrence of rheumatic fever; persons with previous GAS pharyngitis are at greater risk of recurrent attack of rheumatic fever; recurrent ARF attacks can be associated with worsening of the severity of current rheumatic heart disease
 - a. Prompt and adequate treatment of streptococcal pharyngitis
 - b. Administration of monthly injections of long-acting benzathine penicillin (most reliable) or daily oral antibiotic regimen (penicillin)
 - c. Antibiotics for endocarditis prophylaxis prior to dental work or surgical procedures are NOT recommended anymore (see **Table 5-3**)
 - d. If NO valvular disease, prophylaxis should continue for 10 years or until patient is 21 years of age, whichever is longer

■ **Table 5-3** Prevention of Bacterial Endocarditis—American Heart Association Guidelines 2007

Antibiotic prophylaxis with dental procedures is recommended only for patients with cardiac conditions associated with the highest risk of adverse outcomes from endocarditis. These include:

1. Prosthetic cardiac valve
2. Previous endocarditis
3. Congenital heart disease only in the following categories:
 - a. Unrepaired cyanotic congenital heart disease, including those with palliative shunts and conduits
 - b. Completely repaired congenital heart disease with prosthetic material or device, whether placed by surgery or catheter intervention, during the first six months after the procedure
 - c. Repaired congenital heart disease with residual defects at the site or adjacent to the site of a prosthetic patch or prosthetic device which would inhibit endothelialization
4. Cardiac transplantation

Antibiotic prophylaxis solely to prevent bacterial endocarditis is no longer recommended for patients who undergo a GI or GU tract procedure, including patients with the highest risk of adverse outcomes.

■ QUESTIONS

Select the best answer

- The most common congenital heart defect in children is:
 - Tricuspid atresia
 - Ventricular septal defect
 - Aortic stenosis
 - Pulmonary atresia
- The mother of a 4-month-old infant reports that he turned “blue” and seemed to have fast, labored breathing after vigorous crying soon after awakening. He “fell asleep” and his color and breathing seemed to improve. On physical examination, the mucous membranes of the lips and mouth appear mildly cyanotic. A systolic murmur is heard best at the left sternal border. Vital signs are normal with normal peripheral pulses. There is no hepatomegaly. A likely diagnosis is:
 - Congestive heart failure
 - Apnea
 - Coarctation of the aorta
 - Cyanotic spell related to tetralogy of Fallot
- Management of the infant with suspected heart disease and a reported cyanotic spell should include:
 - Prompt referral to a cardiologist
 - An apnea monitor
 - Instructing the parent to keep a diary of these episodes
 - Continuous administration of oxygen
- Chest pain in young children is usually:
 - A symptom of congenital heart disease
 - Noncardiac in origin
 - A sign of hypercholesterolemia
 - A symptom of congestive heart failure
- A common cause of congestive heart failure in the first year of life is:
 - Pulmonary stenosis
 - Ventricular septal defect
 - Rheumatic fever
 - Complete heart block
- The least likely physical finding in a 2-month-old with congestive heart failure is:
 - Tachypnea
 - Tachycardia
 - Hepatomegaly
 - Pedal edema
- A vibratory systolic murmur is heard between the lower left sternal and the apex in a healthy 4-year-old at her preschool physical. The cardiovascular exam is otherwise normal. A likely diagnosis is:
 - Venous hum
 - Still’s murmur
 - Transposition of the great arteries
 - Rheumatic heart disease
- Characteristics of a venous hum include:
 - A systolic murmur
 - Radiation over precordium
 - Marked decrease or disappearance of murmur when child is supine
 - Heard best at lower left sternal border
- Which of the following is true regarding innocent murmurs?
 - The murmur is often holosystolic
 - Prompt referral to a cardiologist is indicated
 - A precordial thrill is present
 - The murmur is low intensity, grade 1–3
- SBE prophylaxis is recommended for:
 - All children with congenital heart disease on a daily basis
 - All children with congenital heart disease before dental, GI, and GU procedures
 - Children with repaired congenital heart disease with a residual defect at the repair site
 - Five (5) years after repair of all congenital heart disease
- A 12-year-old girl seen at a routine visit has a blood pressure of 140/90. She denies any symptoms. The initial management would include:
 - Intravenous pyelogram
 - Return for two repeat blood pressure measurements
 - No follow-up needed—blood pressure probably related to anxiety
 - Diuretic therapy
- A 9-year-old boy presents with a fever of 102°F and complaints of leg pains. His mother reports that he had an upper respiratory infection with a sore throat approximately two weeks ago, which subsided without therapy. On physical examination, he has tender, swollen knees bilaterally. His heart rate is 120/min and a blowing systolic murmur is heard at the apex. No murmur was noted at a previous well-child visit. The most likely diagnosis is:
 - Kawasaki disease
 - Rheumatic fever
 - Sickle cell anemia
 - Viral illness

13. The most useful test for evaluation of suspected acute rheumatic fever is:
- Antistreptolysin-O titer
 - Electrocardiogram
 - Hemoglobin electrophoresis
 - Urinalysis
14. The initial attack of acute rheumatic fever is preceded by:
- A viral illness
 - A group A streptococcal infection
 - Exposure to mites
 - Exposure to chicken pox
15. A 3-week-old infant has a one-day history of irritability, pallor, and poor feeding. He is afebrile. On physical examination, his heart rate is 240/min while asleep. The most likely diagnosis is:
- Supraventricular tachycardia
 - Premature ventricular contractions
 - Sinus tachycardia
 - Cyanotic heart defect
16. Congenital complete heart block may be associated with:
- Maternal lupus erythematosus
 - Wolff-Parkinson-White syndrome
 - Maternal myocardial infarction
 - Kawasaki disease
17. The most common cause of myocarditis is:
- Bacterial
 - Viral
 - Drug reaction
 - Radiation therapy
18. Which of the following is not an expected finding in a child with myocarditis?
- Persistent tachycardia
 - History of antecedent "flu-like" illness
 - A gallop rhythm
 - A significant heart murmur
19. Hypercholesterolemia in children over 2 years old is defined as a total cholesterol at or above:
- 100 mg/dL
 - 130 mg/dL
 - 160 mg/dL
 - 200 mg/dL
20. A potential childhood risk factor for development of atherosclerotic or coronary heart disease as adults is:
- Obesity
 - Tachycardia
 - Heart murmur
 - Aerobic exercise
21. Which of the following is not likely to cause secondary hypercholesterolemia?
- Nephrotic syndrome
 - Hypertension
 - Corticosteroids
 - Obstructive liver disease
22. Which of the following is a common cause of acquired coronary artery disease during childhood?
- Rheumatic fever
 - Hypertension
 - Systemic lupus erythematosus
 - Kawasaki disease
23. Kawasaki disease is most common in:
- Neonates
 - Children less than 5 years of age
 - Children over 6 years of age
 - Females
24. A principal clinical feature of Kawasaki disease includes:
- Low grade fever for 24 hours and a pruritic rash
 - Conjunctivitis with exudate and facial rash
 - Arthritis and chorea
 - Fever persisting at least five days and acute erythema and/or edema of hands and feet
25. An essential test in the evaluation of a 2-year-old being managed for Kawasaki disease is:
- An echocardiogram
 - Electrolytes
 - Cholesterol
 - Streptococcal antibody titer

▣ ANSWERS

- | | |
|--------------|--------------|
| 1. b | 14. b |
| 2. d | 15. a |
| 3. a | 16. a |
| 4. b | 17. b |
| 5. b | 18. d |
| 6. d | 19. d |
| 7. b | 20. a |
| 8. c | 21. b |
| 9. d | 22. d |
| 10. c | 23. b |
| 11. b | 24. d |
| 12. b | 25. a |
| 13. a | |

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Lower Respiratory Disorders

Elizabeth Sloand

■ CROUP (LARYNGOTRACHEOBRONCHITIS)

- Definition: An acute inflammatory disease of the upper airway and larynx caused by a viral infection
- Etiology/Incidence: Parainfluenza virus is the most common organism, followed by respiratory syncytial virus (RSV) and influenza A and B
 1. Most common in children 6 months to 5 years of age; 60% younger than 24 months
 2. Males more often affected than females
 3. Peak incidence during fall and winter
- Signs and Symptoms
 1. URI prodrome; rhinitis for 3 to 4 days, low-grade fever, sore throat
 2. Rapid onset of characteristic “barky” cough, inspiratory stridor, hoarseness
 3. Fever usually absent or low-grade; occasionally high
 4. Symptoms worse at night; symptoms increase with agitation or crying
 5. Can be mild; moderate if stridor persists when at rest
- Differential Diagnosis
 1. Bacterial tracheitis
 2. Epiglottitis
 3. Laryngotracheomalacia
 4. Foreign body aspiration
 5. Extrinsic compression of airway from trauma, tumor, abscess, or congenital malformations
- Physical Findings
 1. Inspiratory stridor
 2. Brassy, barky cough
 3. Dyspnea; expiratory stridor if severe
 4. Chest sounds are usually clear
 5. Low-grade fever; may be high grade
 6. Usually no involvement of lower respiratory tract
- Diagnostic Tests/Findings
 1. Croup is typically a clinical diagnosis; diagnostic tests usually not done
 2. If severe or atypical presentation:
 - a. Pulse oximetry—hypoxia indicates severe disease
 - b. Radiographic image of airway—classic tapering of the subglottic trachea (“steeples sign”)
 - c. White blood cell count—normal or elevated (not usually done)
- Management/Treatment
 1. Mild disease—no dyspnea, hypoxia, or dehydration
 - a. Supportive outpatient care—adequate oral hydration, antipyretics, calm noninvasive care
 - b. Family education regarding worsening respiratory distress
 2. diphtheria
 3. Severe asthma
 4. Retropharyngeal abscess

- c. There is no evidence to support the use of mist therapy; it is often still used since it is harmless and some find it soothing to inflamed mucosa (Shah, 2007)
- 2. Moderate to severe disease—stridor at rest, dyspnea, hypoxemia, or dehydration
 - a. Airway maintenance is the first priority
 - b. Hospitalize for supportive care, oxygen supplementation, medications, and intravenous fluids if indicated
 - c. Fewer than 1% of patients require intubation
- 3. Medications
 - a. Nebulized racemic or L-epinephrine—observe for 3 hours after a dose for rebound respiratory distress
 - b. Corticosteroids will lessen the severity and duration of symptoms
 - (1) Oral dexamethasone—a single dose of 0.15–0.6 mg/kg
 - (2) Intramuscular or IV dexamethasone in a single dose of 0.6 mg/kg
 - (3) Budesonide may also be effective
 - c. Antibiotics are not indicated unless a bacterial infection is present

■ FOREIGN BODY ASPIRATION

- Definition: Inhalation of a foreign body that lodges in the upper trachea or lower airways, resulting in total or partial airway obstruction with local injury and inflammation
- Etiology/Incidence
 - 1. Food or objects with pliable, slick, or cylindrical surface; peanuts, other nuts or seeds, hot dogs, candy, grapes and other raw fruits and vegetables, latex balloons, and small toys or toy parts; nuts are the most common (Midulla, 2005)
 - 2. 73% are < 3 years of age; peak incidence is during second year of life
- Signs and Symptoms
 - 1. Initially, sudden, violent cough with gagging, choking, and subsequent stridor, wheezing, and possible cyanosis
 - 2. Later, may be asymptomatic after object lodges and symptoms decrease; this occurs if object is not obstructing or irritating airway
- Differential Diagnosis
 - 1. Acute phase—laryngotracheitis, epiglottitis, laryngeal edema, pertussis, status asthmaticus, croup, retropharyngeal abscess
 - 2. Chronic phase—asthma, pneumonia, bronchitis, tuberculosis, and tracheal stenosis
- Physical Findings
 - 1. Presentation dependent on substance aspirated as well as size and location of foreign body
 - 2. Unilateral wheezing and localized decreased breath sounds on side of aspiration; may also be bilateral
 - 3. Persistent cough, voice changes, stridor, dyspnea, sputum production, and emesis may be present
 - 4. If foreign body persists, bronchiectasis and abscess formation is likely; may present with recurrent pneumonia and/or hemoptysis
- Diagnostic Tests/Findings
 - 1. Pulse oximetry—decreased O₂ saturations with significant obstruction
 - 2. Chest radiograph—only 10% to 25% of aspirated objects are radiopaque; localized air trapping, atelectasis, and unilateral changes in aeration from obstruction
 - 3. Expiratory chest films (cooperative patients), lateral soft tissue neck films (younger patients), decubitus, or fluoroscopy (imaging throughout the complete respiratory cycle)—obstructive emphysema (failure to deflate) or atelectasis typical; right main stem bronchus main site for foreign bodies
 - 4. If high index of suspicion (witnessed choking and/or abnormal films), rigid bronchoscopy is indicated for diagnostic and therapeutic purposes; if diagnosis uncertain, flexible bronchoscopy may aid diagnosis, but rigid bronchoscopy will still be needed if foreign body found
- Management/Treatment
 - 1. Institute cardiopulmonary resuscitation if needed
 - 2. Immediate transport to hospital for evaluation and removal
 - 3. Use of pulmonary clearance (chest physiotherapy) may be contraindicated if suspicion of foreign body is high; may cause object to migrate to large airways and cause severe obstruction
 - 4. Refer to an expert pediatric endoscopy team for rigid bronchoscopy evaluation and removal (if convincing history of foreign body aspiration, regardless of radiographic findings)
 - 5. Humidification, bronchodilators, and anti-inflammatory medications may be useful after removal
 - 6. Antibiotics if evidence of pneumonia or bronchitis

7. Chest radiograph in six to eight weeks to evaluate resolution of previous findings, or sooner if child is not improving
8. Prevention with parental education
 - a. Remove small items from environment of small child
 - b. Sit while eating and provide adult supervision
 - c. Cut food in small pieces for young children
 - d. Avoid commonly aspirated foods and objects in children under three years of age

■ BRONCHITIS

- Definition
 1. Bronchitis is a nonspecific inflammation of the larger lower airways, associated with several childhood conditions; cough is usually part of the clinical picture
 2. Chronic bronchitis symptoms persist for more than two weeks; chronic bronchitis is rarely an isolated entity in children, but rather a symptom of some other condition
- Etiology/Incidence
 1. Most commonly viral—parainfluenza, RSV, and rhinovirus
 2. Bacterial causes—*Mycoplasma pneumoniae*, *Bordetella pertussis*, *Chlamydia pneumoniae*, and *Corynebacterium diphtheria*
 3. Acute bronchitis occurs most frequently in the winter and early spring
- Signs and Symptoms
 1. Initial phase includes symptoms of an upper respiratory illness, such as rhinitis, nasopharyngitis, and conjunctivitis.
 2. Cough is the hallmark symptom; is initially dry and brassy but may become productive as illness progresses
 3. Sputum may become purulent, which reflects leukocyte migration and not necessarily bacterial involvement
 4. Swallowing sputum may result in nausea
 5. Retrosternal pain as cough persists
- Differential Diagnosis
 1. Pneumonia
 2. Asthma
 3. Allergic disease/posterior nasal drainage
 4. Gastroesophageal reflux and/or aspiration
 5. Exposure to irritants such as cigarette smoke
 6. Cystic fibrosis
 7. Immune deficiency
 8. Chronic sinusitis

9. Immotile cilia syndrome
10. Foreign body aspiration
11. Respiratory tract anomalies
12. Tobacco or marijuana use in adolescents

- Physical Findings
 1. Brassy cough
 2. Coarse, bronchial breath sounds in periphery of lungs
 3. Fever may be low-grade or absent
- Diagnostic Tests/Findings
 1. Diagnosis primarily based on history and physical
 2. Chest radiograph may be normal or show peribronchial thickening
 3. Pulmonary function tests (PFT) may be normal or may indicate an obstructive pattern; PFT may or may not improve after bronchodilator
- Management/Treatment
 1. Primarily supportive; none specific
 2. Acute bronchitis—Increase fluid intake; avoid expectorants, antihistamines, and cough suppressants
 3. Chronic bronchitis is not a distinct entity in children; investigate underlying systemic or pulmonary problem
 4. Bronchodilators if accompanied by wheezing
 5. Inhaled steroids may be indicated for chronic bronchitis
 6. Antibiotics are not usually helpful for viral illnesses; however, antibiotics (with efficacy against mycoplasma and chlamydia species) are useful if bacterial etiology is suspected and cough is > 10 to 14 days duration; children with chronic pulmonary disease other than asthma may benefit from antibiotics for acute exacerbations; consider pertussis if immunizations incomplete
 7. Prevention
 - a. Avoidance of irritants
 - b. Frequent hand washing with careful disposal of nasal and oral drainage
 - c. Avoid overcrowding

■ BRONCHIOLITIS

- Definition: An acute viral infection of the smaller airways; the most common serious acute respiratory illness of young children and infants; characterized by acute inflammation, edema, and necrosis of epithelial cells of the small airways, increased mucous production, and bronchospasm

- Etiology/Incidence
 1. Primarily caused by RSV
 2. Parainfluenza, human metapneumovirus, adenovirus, rhinovirus, influenza, mycoplasma are less common
 3. Older family members may be the infection source for young children, with older children and adults having a much milder illness
 4. Those more likely requiring hospitalization include premature or young infants and those with cardiorespiratory disease (especially bronchopulmonary dysplasia) or immunodeficiency
 5. Most commonly occurs in midwinter to early spring
 6. More severe disease in boys, infants who have not been breastfed, and with lower socioeconomic status
- Signs and Symptoms
 1. Initial symptoms—rhinorrhea, congestion, cough, decreased appetite and low-grade fever
 2. Progresses to increased work of breathing, tachypnea, decreased appetite or poor suck; change in level of consciousness or activity level
 3. Oftentimes, history of exposure to an older child or adult with a mild respiratory illness
 4. May present as apnea, lethargy, irritability, or poor feeding in very young or premature infants
- Differential Diagnosis
 1. Bacterial or chlamydial pneumonia
 2. Aspiration pneumonia
 3. Asthma and allergy
 4. Foreign body aspiration
 5. Cystic fibrosis
 6. Congenital malformations of the respiratory tract
- Physical Findings
 1. Wheezing is the most prominent physical finding; crackles; very faint breath sounds suggest more severe disease with more significant obstruction
 2. Tachypnea with shallow breathing, retractions, nasal flaring, prolonged expiratory phase, paroxysmal wheezy cough
 3. Rhinorrhea, otitis, conjunctivitis, and/or pharyngitis
 4. Afebrile, or low-grade fever
 5. Palpable liver and spleen due to hyperinflation of lungs
- Diagnostic Tests/Findings
 1. Diagnosis primarily based on history and physical; labs and diagnostics not routinely recommended (AAP, 2006)
 2. Chest radiograph—focal infiltrates, hyperinflation, peribronchial thickening, air trapping, patchy or subsegmental atelectasis
 3. Pulse oximetry—hypoxemia with significant disease
 4. White blood cell count with differential—may or may not be increased
 5. Viral nasal wash with rapid diagnostic techniques—may be positive for RSV
- Management/Treatment
 1. Careful monitoring of respiratory status, use oxygen therapy if hypoxemic
 2. Mechanical ventilation rarely necessary in previously healthy infants, but may be required in high-risk groups
 3. Fluid and nutritional support
 4. Bronchodilators—not routinely recommended; can be used in select infants; discontinue if not beneficial
 5. Corticosteroids—not routinely recommended; use in those with underlying chronic conditions
 6. Ribavirin use is generally not recommended; use based on clinical condition and physician experience
 7. Antibiotics not routinely used unless bacterial infection is suspected
 8. Prevention
 - a. Palivizumab is indicated for children younger than 2 years of age with chronic lung disease of prematurity, history of prematurity (< 35 weeks gestation), or congenital heart disease; Dose: 15 mg/kg IM, given in five monthly doses, beginning in November/December (Red Book, 2009)
 - b. Strict contact precautions (gown, gloves) in hospitalized children
 - c. Home measures—decrease exposure to contagious settings, avoid tobacco smoke exposure, and careful hand washing
 - d. An RSV vaccine is currently in development

▣ PNEUMONIA

- Definition: An inflammation of the pulmonary parenchyma
 1. Infectious pneumonia—infection which usually involves small airways in children, but may also infect the larger airways and/or the alveoli

2. Aspiration pneumonia—caused by the ingestion of food, saliva, gastric contents, or other substances into air passages
- Etiology/Incidence
 1. Common viral agents—RSV, parainfluenza, influenza A and B, and adenovirus; usually involves the small airways
 2. Neonates: group B strep, enterococcus, *Listeria monocytogenes*, *E. coli*, ureaplasma urealyticum
 3. 1 to 3 months: most common bacterial causes include *S. aureus*, streptococcus pneumoniae, *H. flu*, *Chlamydia trachomatis*, *Bordetella pertussis*, ureaplasma
 4. 4 months to 4 years: *S. pneumoniae*, *S. aureus*, *H. flu*
 5. 5 years to adolescence: *Streptococcus pneumoniae* is primary cause of bacterial pneumonia; *Mycoplasma pneumoniae*, *Mycoplasma tuberculosis*
 6. Immunocompromised or malnourished children—consider opportunistic organisms, such as *Pneumocystis carinii*, yeasts
 7. Many other less common pathogens can be considered
 8. Aspiration pneumonia occurs most frequently in children with upper airway abnormalities or neurologic deficits, severe gastroesophageal reflux and other esophageal problems; may also occur subsequent to the inhalation of smoke or hydrocarbons; following near drowning or ethanol ingestion by adolescents
 9. Annual incidence 3% to 5%; highest incidence in children < 5 years
 10. Incidence of *Haemophilus influenzae pneumoniae* and pneumococcal pneumonia have dramatically decreased since introduction of Hib and heptavalent pneumococcal conjugate vaccines, respectively
 - Signs and Symptoms
 1. Cough occasionally productive or associated with emesis
 2. Tachypnea is a key finding; also retractions, expiratory grunting, nasal flaring, cyanosis, and rash
 3. Fever
 4. Neck or chest pain (upper lobe), or abdominal pain (lower lobe); nausea/vomiting
 5. Wheezing (frequent in viral etiology)
 6. May have change in level of consciousness, activity, feeding
 7. Bacterial pneumonia—usually an abrupt onset of fever, chills, cough, and chest pain
 - Differential Diagnosis
 1. Bronchiolitis, URI, croup
 2. Foreign body aspiration
 3. Asthma
 4. Cystic fibrosis
 5. Sepsis, meningitis, pertussis
 6. Tuberculosis
 7. Immunodeficiency
 8. Reflux, swallowing dysfunction
 - Physical Findings
 1. Fever with bacterial pneumonia
 2. Tachypnea is most consistent finding; sub-sternal, supraclavicular, and intercostal retractions; grunting, nasal flaring; pallor/cyanosis/decreased oxygen saturation
 3. Rales/crackles, wheezing, crackles, diminished or tubular breath sounds
 4. If pleural effusion present, may have decreased breath sounds with dullness to percussion
 - Diagnostic Tests/Findings—in an uncomplicated patient who is stable, no diagnostic tests are needed; if not stable or with fever > 39°C, chest radiography is advised
 1. Chest radiograph—severity of disease may not directly correlate with radiography findings; not reliable for distinguishing between bacterial and viral disease
 - a. Atelectasis—opacification, intrathoracic structures will shift toward the atelectatic area; hemidiaphragm on affected side may be elevated and intercostal spaces on that side may be narrow
 - b. Viral pneumonia—may show hyperinflation and bilateral interstitial infiltrates and peribronchial cuffing
 - c. Bacterial pneumonias—may show patchy infiltrates in infants; lobar consolidation is most common with *Streptococcus pneumoniae* or *Haemophilus influenzae*; hilar adenopathy is most common with *Haemophilus influenzae* or *Staphylococcus aureus*; pleural effusion may be present
 - d. Acute aspiration pneumonia usually develops in portion of lung that is dependent at time of aspiration; otherwise, radiograph typically shows diffuse or localized mottled infiltrates with or without atelectasis
 2. White blood cell count—bacterial pneumonia is associated with elevated WBCs (15,000–40,000/mm³); may or may not be elevated with viral pneumonia
 3. Viral cultures of nasopharyngeal secretions; rapid identification with immunofluorescent assay

4. Blood cultures—if child is toxic, is an infant, or if bacterial pneumonia is suspected (frequently negative); positive 30% with pneumococcal pneumonia
 5. Sputum cultures—warranted if cough is productive; rarely beneficial in younger children
 6. Positive cold agglutinin screen or titer > 1:64 is suggestive of *Mycoplasma pneumoniae*; positive PCR test or seroconversion in an IgG assay can diagnose *Mycoplasma pneumoniae*. (Usually not done due to usual clinical diagnosis and effective treatment)
 7. PPD if TB is suspected
 8. Pulse oximetry—decreased O₂ saturations with severe disease
- Management/Treatment
 1. Most often viral (> 80%) and self limited
 2. Antimicrobial treatment based on etiology and age of the child
 - a. Hospitalized: birth to 3-week-old child—IV ampicillin and third generation cephalosporin, X 10 to 21 days; 3-week to 4-month-old child—IV cephalosporin with or without a macrolide, X 10 days; 4-month to 4-year-old—IV cephalosporin or high dose ampicillin X 7 to 10 days; 5 years or older—IV cephalosporin with or without azithromycin X 7 to 10 days
 - b. Outpatient: 4-month to 5-year-old—amoxicillin 100 mg/kg/day; 5 years or older—Azithromycin 12 mg/kg/day or amoxicillin 80 to 100 mg/kg/day
 - c. All ages consider *Staph aureus* if rapidly toxic, > 50% with effusions
 - d. For infants consider *B. pertussis* if immunizations incomplete or *C. trachomatis* if mother infected
 - e. Consider anaerobes if aspiration pneumonia; treat with clindamycin
 - f. Bronchodilators and chest physiotherapy may improve airway clearance
 - g. Influenza and/or pneumococcal vaccine for high-risk groups
 - h. Other supportive therapy may include additional fluids and/or oxygen
 - i. Pneumonia in immunocompromised hosts can progress rapidly; should be managed in a monitored setting, with aggressive diagnostic measures and appropriate broad-spectrum antimicrobial coverage
 - j. Refer to pulmonary specialist if recurrent disease without known cause

■ CYSTIC FIBROSIS (CF)

- Definition: Autosomal recessive disorder of cystic fibrosis transmembrane regulation (CFTR), causing defective epithelial ion transport, which results in dehydrated, viscous secretions which obstruct the exocrine ducts (with subsequent destruction and scarring) in the respiratory, hepatobiliary, gastrointestinal, and reproductive tracts
- Etiology/Incidence
 1. Caused by mutations in a single gene on the long arm of chromosome 7 which directs the production of CFTR
 - a. CFTR is the principal chloride channel of epithelial cells and controls other ion transport
 - b. Defective CFTR results in increased sodium reabsorption, decreased chloride secretion, and dehydrated, highly viscous secretions in all exocrine ducts which causes disease
 - c. There are over 1500 mutations of the CFTR gene causing CF although the functional importance of only a small number are known
 2. Incidence—most common fatal genetic disorder affecting Caucasian population; less frequent among other racial groups
 3. Median life expectancy—37 years (in 2006) with respiratory failure leading cause of death; life expectancy slowly increasing as treatments improve
- Signs and Symptoms
 1. Extremely viscid meconium (newborns), with delayed passage
 2. Poor growth despite normal to increased appetite
 3. Recurrent and chronic respiratory infections (sinuses and lower airways); dyspnea on exertion which progresses to dyspnea at rest as disease progresses
 4. Large, bulky, foul-smelling, greasy stools
 5. Frequent flatulence or abdominal pain
 6. Chronic cough, varies in character, but usually productive, blood-streaked mucus not uncommon; major hemoptysis can be life threatening
 7. Recurrent or persistent wheezing
 8. Salty-tasting skin
 9. Distal small bowel obstruction
 10. Recurrent pancreatitis
 11. CF-related diabetes mellitus (30% of those > 25 years)
 12. Salt loss syndromes

13. Heat prostration with hypernatremia dehydration
 14. Male infertility (98% of men sterile) and women have reduced fertility
- Differential Diagnosis
 1. Asthma
 2. Recurrent pneumonia, bronchitis
 3. Immunologic deficiencies, metabolic alkalosis
 4. Celiac disease, protein losing enteropathy, α_1 -antitrypsin deficiency (rare)
 5. Airway abnormalities, e.g., airway stenosis, vascular ring, etc.
 6. Gastroesophageal reflux, with or without aspiration
 7. Other causes of malabsorption and/or failure to thrive
 - Physical Findings
 1. Wheezing and air trapping with increased anteroposterior diameter of the chest (barrel chest) as disease progresses
 2. Crackles
 3. Increased work of breathing with accessory muscle use
 4. Nasal polyps, chronic sinusitis
 5. Failure to thrive from maldigestion and fat malabsorption (90% of patients)
 6. Abdominal pain and distention, unexplained pancreatitis or cirrhosis, hepatosplenomegaly, cholelithiasis
 7. Rectal prolapsed
 8. Digital clubbing
 9. Delayed puberty
 10. Meconium illius (15% of newborns)
 - Diagnostic Tests/Findings
 1. Pilocarpine iontophoresis sweat test—two tests with a sweat chloride in excess of 60 mmol/L; test should be performed by a Cystic Fibrosis Foundation-approved laboratory
 2. Genetic testing for mutation analysis; certain countries and states include genetic analysis in newborn screening; prenatal screening available
 3. Other findings
 - a. Elevated immunoreactive trypsin (IRT) in newborn; elevated IRT must be confirmed with sweat test
 - b. Milder, atypical presentations are being identified by expanded diagnostic criteria
 - c. Chest radiograph—hyperinflation, increased peribronchial markings, atelectasis, bronchiectasis
 - d. CT scan—early bronchiectasis
 - e. Pulmonary function tests—demonstrates obstructive pattern; decreased flow rates and vital capacity as disease progresses
 - f. Sputum culture—typical CF pathogens: *Staphylococcus aureus*, *Haemophilus influenzae*, *Pseudomonas aeruginosa*, *Burkholderia cepacia*
 - g. Oximetry—decreased oxygen saturation with exacerbation and worsening disease
 - h. Hyponatremia, hypoproteinemia, hypochloremic alkalosis
 - i. Elevated liver enzymes (AST or ALT)
 - j. Fat soluble (A, E, and K) vitamin deficiencies
 - k. Glucose testing
 - l. Sinus CT—chronic inflammation/disease
- Management/Treatment
 1. Referral to cystic fibrosis care center for long-term care
Cystic Fibrosis Foundation
6931 Arlington Road
Bethesda, Maryland 20814
(301) 951-4422
Fax: (301) 951-6378
(800) FIGHT CF (800-344-4823) for list of approved centers
www.cff.org
 2. Interdisciplinary management involving intensive education, airway clearance techniques, replacement of pancreatic enzymes, nutritional support, prevention and aggressive treatment of infection, and psychosocial support
 3. Routinely given medications
 - a. Inhaled mucolytic agent
 - b. Recombinant human DNase (Pulmozyme)
 - c. Inhaled tobramycin
 - d. Oral azithromycin for chronic *Pseudomonas aeruginosa*
 4. Antibiotic therapy based on sputum culture and sensitivity
 - a. Given early in course of disease to delay onset of chronic colonization with *P. aeruginosa* (the primary organism found in the airways of CF children and adults; once established, it is nearly impossible to eradicate)
 - b. Once colonized, given to slow decline in pulmonary function
 - c. Intravenous courses given for pulmonary exacerbations to restore pulmonary function and reduce symptoms
 5. Isolation—standard precautions for all patients; contact and droplet precautions when indicated, minimize contact with other CF patients to avoid spread of infections

(particularly if infected with *Burkholderia cepacia*, a virulent pathogen that has innate antibiotic resistance, causes rapid decline in lung function and is associated with mortality in CF patients)

6. Role of primary care provider—annual influenza vaccine, more liberal use of antibiotics for respiratory infections, be alert for CF complications

■ ASTHMA

- Definition: Chronic lung disease characterized by:
 1. Airway inflammation, the underlying pathologic process, which contributes to:
 - a. Airway hyper-responsiveness to a variety of stimuli
 - b. Variable airway obstruction that is partially or completely reversible, either spontaneously or with treatment
 2. Immunohistopathologic features include:
 - a. inflammatory cell infiltration with neutrophils, eosinophils, lymphocytes
 - b. mast cell activation and epithelial cell injury
 3. Persistent airway inflammation can lead to airway wall remodeling, irreversible changes, and progressive loss of pulmonary function
- Etiology/Incidence
 1. Airway inflammation plays a central role
 - a. Inflammation causes airway narrowing and increased airway secretions
 - b. Inflammation contributes to airway hyper-responsiveness
 - c. Broad variety of factors trigger inflammation
 - d. Causes recurrent acute episodes
 2. Atopy, the genetic predisposition for development of an IgE-mediated response to common allergens, is the strongest predisposing factor for developing asthma
 3. Triggers—factors that provoke airway inflammation and acute episodes
 - a. Viral respiratory infections—one of the most common causes of asthma exacerbations in young children and may contribute to the development of asthma
 - b. Indoor allergens—dust mites, roaches, mold, animal saliva/dander
 - c. Outdoor allergens—seasonal trees, grasses, weeds, pollens, molds
 - d. Weather/humidity changes
 - e. Exercise induced
 - f. Gastroesophageal reflux
 - g. Irritants—tobacco smoke, pollution, strong odors

- h. Food allergies—nuts, shellfish, sulfites (shrimp, dried fruit, beer or wine)
 - i. Medications—aspirin, beta blockers, non-steroidal anti-inflammatory drugs
 - j. Psychosocial factors such as crying, laughing, anxiety attacks
4. Approximately 8.9% of children, 6.5 million in the U.S. affected; incidence and mortality increasing worldwide
 - a. Rising incidence in childhood and all age groups, especially among males, lower socioeconomic groups, urban African-Americans and Hispanics, and those with family history of asthma or allergies
 - b. A leading cause of hospitalization and school absenteeism
 5. Underdiagnosis and inappropriate treatment are major contributors to morbidity and mortality

- Signs and Symptoms
 1. Recurrent episodes of cough, wheezing, mucus production, chest tightness, breathlessness, and decreased endurance
 2. Symptoms often display an initial response (bronchospasm) to a trigger (early-phase asthmatic response), then a late (inflammatory) phase response approximately 4 to 12 hours after the initial response that is more severe and prolonged than the earlier response and can last hours to several weeks
 3. Symptoms often worse at night, early morning, and during or after exercise
 4. Variability—some may have severe, life-threatening exacerbations separated by long periods of normal lung function and no symptoms
 5. A subset present with chronic (usually nighttime) cough without wheezing or exercise intolerance (cough-variant asthma)
- Differential Diagnosis
 1. Upper or lower respiratory tract infections
 2. Foreign body aspiration
 3. Cystic fibrosis
 4. Cardiac or anatomical defects
 5. Vocal cord dysfunction
 6. Conditions that may coexist with asthma and complicate diagnosis include gastroesophageal reflux and obstructive sleep apnea
- Physical Findings: Pulmonary exam is normal when asymptomatic; when symptomatic:
 1. Cough and shortness of breath
 2. Diffuse wheezes (initially heard at the end of the expiratory phase) with decreased air flow and prolonged expiratory phase

3. Respiratory distress, retractions, increased hypoxia, and decreased breath sounds as severity increases
 4. Allergic appearance—allergic “shiners,” Dennie’s lines, nasal crease, nasal congestion and mouth breathing, and boggy turbinates and nasal edema; nasal polyps (consider CF)
 5. Skin—atopic dermatitis
 6. Concurrent respiratory infection—viral respiratory illness, sinusitis
 7. Barrel chest—chronic hyperinflation
- Diagnostic Tests/Findings
 1. Thorough history is vital to diagnosis and management, with special emphasis on triggers, severity of previous episodes, response to medications, and family history of asthma or allergy
 2. Pulmonary Function Testing (Spirometry)—the gold standard in diagnosing asthma in children older than 4 to 5 years; demonstrates obstruction and assess reversibility
 - a. Lower airway obstruction indicated by a reduction in the values for both FEV₁ (the forced expiratory in 1 second) and the FEV₁/FVC (forced volume vital capacity [or FEV₁/FEV₆]) relative to reference or predicted values
 - b. Reversibility—FEV₁ increases at least 12% from baseline OR FEV₁ increases of at least 10% of predicted after using a short-acting inhaled beta₂ agonist
 3. Chest radiograph
 - a. Bilateral hyperinflation with peribronchial thickening
 - b. Atelectasis (can be misread as pneumonia)
 - c. May be normal
 - d. May be unnecessary unless uncertain diagnosis or if poor response to treatment
 4. Evaluation of other factors contributing to asthma severity
 - a. Allergy evaluation and treatment as indicated to improve allergy/asthma control
 - (1) Complete blood count—eosinophilia
 - (2) Immunoglobulin E—elevated, values age dependent
 - (3) Radioallergosorbent test (RAST)—serum test which identifies possible allergic responses to various environmental substances
 - (4) Skin testing
 - b. Nasal and sinus evaluation—identify and treat allergic rhinitis, sinusitis; consider sweat test if CF is suspected
 - c. Gastroesophageal reflux assessment
5. Pulse oximetry—evaluate hypoxemia during an acute episode
- Management/Treatment—as recommended by National Heart, Lung, and Blood Institute, *Expert panel report 3: Guidelines for the diagnosis and management of asthma. National Asthma Education and Prevention Program (EPR-3)*; Asthma treatment should be based on this comprehensive plan and document, available at <http://www.nhlbi.nih.gov/guidelines/asthma/asthgdln.pdf>
 1. Goals in primary care—reduce current impairment and reduce future risk; child should live as normal a life as possible
 2. Four components of asthma management in guidelines:
 - a. Accurate measures of assessment and monitoring
 - b. Pharmacologic therapy
 - c. Education for a partnership in asthma care
 - d. Control of environmental factors and comorbid conditions that affect asthma
 3. Initially, assess asthma for severity to initiate therapy, and subsequently, assess control for monitoring and adjusting therapy
 4. Asthma severity is determined *before* therapy; if already on medications, determined by type of medications needed for control
 - a. Classifications divided into four groups: intermittent, mild persistent, moderate persistent, and severe persistent, and consider both impairment (burden of symptoms) and risk (morbidity potential).
 - b. Guidelines for assessment and treatment are divided into three age groups: 0 to 4 years old, 5 to 11 years old, and 12 years old and older
 - c. Classification based on:
 - (1) Frequency and timing of symptoms
 - (2) Interference with daily activities
 - (3) PFTs
 - (4) Need for short-acting medications
 - d. If a child’s assessment falls between two levels of severity, they should be treated for the highest level (most aggressive)
 5. Medication treatment is based on severity and control, using a step-wise approach created by the EPR-3
 - a. Ranges from step 1 (intermittent asthma) to step 6 (severe persistent)
 - b. Step-wise approach is intended to assist, not substitute, for clinical decision making and individualized care

6. Medications are in two categories: long-term controller and quick-relief
 - a. Long-term control medicines—taken daily
 - (1) Inhaled corticosteroids (ICS) are the preferred medication for all children with persistent asthma (beclomethasone, budesonide, flunisolide, fluticasone, triamcinolone); delivered by nebulizer or inhaler; dose of drug matched to level of disease severity
 - (2) Leukotriene modifiers (montelukast and zafirlukast)
 - (3) Long-acting beta₂ agonists (LABA)—salmeterol and formoterol, inhaled
 - (a) Use in combination with ICS, but only use when ICS are not effective alone; do not use as monotherapy
 - (b) Black box warning due to association with severe asthma attacks and death; close monitoring advised
 - (4) Omalizumab, an immunomodulator that prevents binding of IgE to mast cells, used for steps 5 and 6 in controlled clinical settings due to side effects
 - (5) Cromolyn sodium or nedocromil (inhaled)—useful for exercise induced bronchospasm
 - (6) Methylxanthines—used rarely
 - (7) Oral corticosteroids in short courses for control of symptoms; also used as controller with step 6 severity (if severe, chronic asthma, not to exceed 60 mg/day)
 - b. Quick relief medications—taken as needed to provide prompt reversal of acute airflow obstruction and relief of bronchoconstriction; not for daily long-term use
 - (1) Inhaled short-acting beta₂ agonists (albuterol, levalbuteral)
 - (2) Anticholinergics (ipratropium bromide, inhaled)
 - (3) Short course systemic corticosteroids (oral or parenteral)
7. Assess for control and adjust treatment
 - a. Close follow-up; return visits scheduled every 1 to 6 months
 - b. Step therapy up or down according to level of control and severity
 - c. Spirometry can assist in follow-up visits
8. Educate for a partnership in care of child with asthma
 - a. Identify and address family and child's concerns
 - b. Establish open lines of communication; be mindful of cultural differences
 - c. At every visit, review the written asthma action plan that has clear instructions of medications, doses, signs/symptoms of problems, and instructions for increasing therapy or contacting provider
 - d. Review child's technique with devices; peak flow meter, inhaler, and spacer
9. Control of environmental factors and comorbid conditions
 - a. Those with persistent asthma should have skin testing or in vitro testing to identify specific allergens
 - b. Identify and reduce child-specific triggers such as house dust mites, cockroaches, pet dander, indoor and outdoor plants and trees, and molds
 - c. Employ a broad approach with family to reduce allergen exposure in the environment
 - d. Make strong recommendations to eliminate environmental tobacco smoke (ETS)
 - (1) Identify all tobacco smokers in child's environment and advise them to quit
 - (2) Assist by referral to appropriate smoking cessation programs and advising nicotine replacement therapy

■ BRONCHOPULMONARY DYSPLASIA (BPD)

- Definition: Any child requiring supplemental oxygen at 36 weeks or greater (postconceptual age) with radiographic changes of chronic lung disease
- Etiology/Incidence
 1. Disease of infants < 1000 g at birth and < 28 weeks gestation
 2. Most have normal lung function at birth but develop respiratory failure within the first weeks of life
 3. Develops in babies who are treated with oxygen and positive pressure ventilation
 4. Majority of those who develop BPD do so following respiratory distress syndrome; other causes include persistent pulmonary hypertension, meconium aspiration, severe lower respiratory infection, maternal infection (amnionitis), or chronic aspiration
 5. Leading cause of chronic lung disease in infants
 6. Multifactorial—lung immaturity, oxygen toxicity, barotrauma from mechanical ventilation, infections, malnutrition, or fluid overload
 7. Mild to severe disease possible (decreased lung compliance and increased airway resistance)

8. 5,000 to 10,000 infants per year develop BPD in the U.S.; white male infants at higher risk
- Signs and Symptoms
 1. Acute respiratory distress in first week of life; may have cough and wheeze
 2. Poor growth and poor feeding skills
 3. Fussy with decreased endurance
 - Differential Diagnosis
 1. Meconium aspiration syndrome
 2. Congenital infection
 3. Congenital cardiac or pulmonary defects that result in supplemental oxygen requirement
 4. Upper airway obstruction
 5. Lower airway abnormalities (structural/anatomic, extrinsic compression, tracheomalacia)
 6. Pulmonary hypertension
 7. Metabolic disorders
 8. Respiratory infections/immunodeficiency
 9. Chronic gastroesophageal reflux
 10. Chronic or recurrent aspiration
 - Physical Findings—vary with severity of disease
 1. Tachypnea with retractions
 2. Diffuse inspiratory or expiratory crackles and/or wheezes
 3. Pale with cyanotic episodes
 4. Findings consistent with cardiac insufficiency or fluid overload
 5. Fluid sensitivity
 6. Associated findings consistent with sequelae of prematurity and/or chronic lung disease
 - Diagnostic Tests/Findings
 1. Oximetry on room air—hypoxemia
 2. Growth curve—often FTT, even after corrected for prematurity
 3. Serum electrolytes
 - a. Carbon dioxide retention
 - b. Chloride and potassium depletion, metabolic alkalosis—with diuretic use
 4. Arterial blood gases (on room air)—to monitor in acute and chronic stages
 5. Chest radiograph—abnormalities may include hyperinflation, emphysema, cyst formation, fibrosis, CV changes
 6. Electrocardiogram and echocardiogram to monitor cardiac status
 - Management/Treatment
 1. Maintain adequate oxygenation (92% or greater) to prevent cor pulmonale and promote growth
 - a. Provide supplemental oxygen—wean as tolerated, expect gradual improvement with growth
 2. Adequate nutrition and fluids
 - a. Nutritional supplementation and hypercaloric formulas to provide additional calories; nutrition referral
 - b. Occupational therapy intervention for feeding skills
 - c. Gastrostomy or supplemental tube feedings as needed
 - d. Balance nutrition volume with fluid restriction when fluid sensitive
 - e. Treat gastroesophageal reflux as needed
 3. Medications
 - a. Bronchodilators—inhaled, may decrease airway resistance
 - b. Diuretics—as needed for acute fluid overload; avoid chronic use if possible
 - c. In older BPD children, inhaled β agonists with inhaled steroids can be used for symptomatic relief
 4. Protective care to avoid lung reinjury
 - a. Immunizations, including influenza
 - b. Smoke free environment
 - c. RSV prevention—administer Palivizumab or RSV intravenous immunoglobulin
 - d. Avoid high density daycare settings
 - e. Good hand washing and other infection control precautions
 - f. Avoid elective procedures, especially during winter months
 5. Family education, support, and follow-up
 - a. Close follow-up (every 1 to 3 months) to determine adequate oxygenation and growth
 - b. Higher incidence of rehospitalization in early years due to exacerbations
 6. Monitor for associated findings, manage/refer as needed—airway disorders, hypertension, gastroesophageal reflux, heart failure, neurodevelopmental problems, ophthalmology or renal complications
 7. High risk for neurologic, developmental, or academic concerns throughout childhood
 8. Referrals
 - a. Early intervention for developmental assistance and monitoring
 - b. Financial assistance and counseling
 - c. Home nursing and equipment companies
 - d. Family counseling and support groups
 - e. Community rescue services
 - f. Case management
 9. Prevention—reduce incidence of premature births
 - b. Desaturation commonly occurs during sleeping and feeding

■ APNEA

- Definition: Respiratory pause lasting > 20 seconds or associated with cyanosis, bradycardia, marked pallor, or hypotonia; a disorder of respiratory control; may be central, obstructive, or mixed
 1. Types
 - a. Central apnea—caused by decreased CNS stimuli to respiratory muscles; cessation of airflow without accompanying respiratory effort
 - b. Obstructive sleep apnea (OSA)—most common form of apnea; cessation of airflow despite accompanying respiratory effort
 - c. Mixed apnea—evidence of both central and obstructive apnea
 - d. Apnea of infancy—pathologic apnea in infants that are > 37 weeks gestation at onset; always demands diagnostic evaluation
 - e. Apnea of prematurity—periodic breathing with pathologic apnea in a premature infant < 37 weeks gestation; occurs in the absence of identifiable predisposing disease; common and may be due to prematurity or an associated illness
 - f. Periodic breathing—three or more respiratory pauses of > 3 seconds duration with < 20 seconds of respiration between pauses; common and physiologic in preterm infants; pathologic if accompanied by cardiac or respiratory instability
 - g. Apparent Life Threatening Event (ALTE)—an episode that an observer believes is life threatening; includes some combination of apnea, change in skin color, marked change in muscle tone, choking or gagging, and requires significant intervention to restore normal breathing
- Etiology/Incidence
 1. Apneas vary significantly depending on underlying etiology
 2. Gestational age is the most significant determinant of respiratory control; younger gestational age corresponds to more apnea
 3. ALTE—most frequent associated problems are GI (50%), neurologic (30%), respiratory (20%), CV (5%), metabolic and endocrine (< 5%); more than 50% remain unexplained and then are termed “apnea of infancy;” no causal link with SIDS
 4. Obstructive sleep apnea syndrome (OSAS)
 - a. Prevalence of 1% to 4%
 - b. 2 to 8 years peak age (peak adenotonsillar hypertrophy)
 - c. Increased risk with craniofacial anomalies, obesity, Down syndrome
- Signs and Symptoms
 1. During event, respiratory pause accompanied by marked pallor, cyanosis, hypotonia, and bradycardia
 2. OSAS—most common presentation is snoring; typically loud, habitual snoring; apnea followed by gasping, choking, and arousal; restless sleep and diaphoresis
 3. OSAS daytime symptoms—behavioral changes, poor school performance, daytime sleepiness, hyperactivity, growth delay
- Differential Diagnosis of ALTE—very broad range, including:
 1. GER
 2. Sepsis/meningitis
 3. Respiratory syncytial virus infection
 4. Seizures
 5. Metabolic defects
 6. Cardiac defects
 7. Munchausen syndrome by proxy (MSP)
- Physical Findings
 1. Often normal at time of examination
 2. OSAS—mouth breathing; upper airway narrowed/obstructed from inflammation or structural defects; allergic stigmata; congestive heart failure; obese, normal weight, or failure to thrive; adenotonsillar hypertrophy
 3. Lungs usually clear
 4. May have physical findings of cardiac insufficiency or fluid overload with longstanding, severe obstruction
- Diagnostic Tests/Findings
 1. Complete history—emphasize circumstances surrounding the event (ALTE), medical and family history
 2. Complete physical examination with detailed neurodevelopmental assessment
 3. Based on history, examination, and age of child
 4. Arterial blood gas—indicate severity of event and current stability
 5. Pulse oximetry—likely normal, may show hypoxemia
 6. Electroencephalogram—seizure disorder
 7. Viral respiratory panel—RSV infection
 8. Complete blood count, blood culture—indicative of sepsis, polycythemia (chronic hypoxemia), anemia
 9. Electrolytes—elevated bicarbonate level suggests compensation for hypoventilation

10. Fluid balance profile—metabolic status, elevated bicarbonate (chronic respiratory insufficiency)
 11. Lumbar puncture—meningitis
 12. Electrocardiogram—cardiac arrhythmias
 13. Radiographs—chest to evaluate structure and anatomy, heart size; lateral neck to evaluate size of adenoids
 14. Sleep study (overnight polysomnogram)—definitive test to identify significant apneas and differentiate type of apneas
- Management/Treatment (based on history and physical findings)
 1. Institute treatment and referrals based on diagnostic findings, history, and physical examination
 2. If cause unclear, decision is more complex, but should include:
 - a. Frequent follow up visits with serial measurements of:
 - (1) Height, weight, and head circumference
 - (2) Developmental assessment
 - (3) Emotional support and parental counseling
 3. ALTE—Admit to hospital for close observation and monitoring
 4. Infants—Apnea monitor recommendations with a specific clinical indication
 - a. Use monitor with an event recorder
 - b. Monitor only detects episodes of central apnea
 - c. Monitoring does not prevent all deaths
 - d. Monitoring is stressful (false alarms, child safety, travel cumbersome, and financial burden)
 - e. Requires close supervision and plan for monitor termination
 - f. CPR instruction for family and other caregivers
 5. OSAS
 - a. Referrals as indicated for adenotonsillectomy, weight loss, or continuous positive airway pressure device with sleep
 - b. Repeat sleep study after adenotonsillectomy (may have persistent obstruction despite resolution of snoring)

■ TUBERCULOSIS (TB)

- Definition: A chronic and serious granulomatous infection which may cause pulmonary, extrapulmonary, or disseminated disease

- Etiology/Incidence
 1. Caused by *Mycobacterium tuberculosis*
 2. Primary mode of transmission is inhalation of aerosol droplets through person-to-person contact; almost all cases of pediatric TB are acquired through close and prolonged contact with an untreated infected adolescent or adult
 3. Tuberculosis is classified as primary infection or tuberculosis disease
 - a. Latent tuberculosis infection (LTBI)—positive tuberculin skin test without clinical, radiographic, or laboratory evidence of active disease
 - b. Tuberculosis disease—when child has signs or symptoms of disease or there is radiographic evidence of disease; clinical manifestations occur 1 to 6 months after infection
 - (1) Pulmonary—the lung is the primary site of infection; involves hilar, subcarinal, or paratracheal adenopathy, usually with chest radiographic findings of patchy infiltrates; chest radiographic evidence may be so subtle that chest CT is needed for diagnosis
 - (2) Extrapulmonary—miliary tuberculosis may involve any organ of the body, most frequently seen in lymph nodes and central nervous system, and can include meningitis
 4. TB rates for all ages in U.S. are highest in urban, low-income areas, and in nonwhite and foreign-born groups
 5. Increased TB risk associated with:
 - a. Contacts of persons with confirmed or suspected infectious TB
 - b. Foreign-born persons from high-risk countries in Asia, Africa, Latin America, and the Middle East; children who travel to these countries; significant contact with persons from these countries
 - c. Homeless, migrant, institutionalized, or prison populations
 - d. Native Americans
 - e. Other—immunodeficiency (HIV) or immunosuppression, IV drug use, diabetes, chronic renal failure, malnourished
- Signs and Symptoms
 1. Usually asymptomatic
 2. Cough, wheezing, dyspnea
 3. Abdominal pain, diarrhea, poor weight gain or weight loss, anorexia, or poor suck
 4. Fever, lethargy, night sweats, chills
 5. May present with clinical symptoms of meningitis

6. Late clinical presentations include symptoms of TB of middle ear and mastoid, bones, joints, skin, and kidneys
 7. Drug resistant TB is clinically indistinguishable from drug-susceptible disease
- Differential Diagnosis
 1. Histoplasmosis
 2. Atypical mycobacteria
 3. Pneumonia
 4. Lung abscess
 5. Foreign body aspiration
 6. Sarcoidosis
 7. Neoplasm
 8. Crohn's disease
 9. Meningitis
 - Physical Findings
 1. Usually asymptomatic
 2. Dry, hacking, or brassy, paroxysmal cough
 3. Localized wheezing, crackles, rales
 4. Lymphadenopathy—cervical or axillary
 5. Poor weight gain, possibly growth delay
 6. Low-grade fever, night sweats, chills
 7. Splenomegaly, hepatomegaly
 - Diagnostic Tests/Findings
 1. Tuberculin skin test (TST)—Mantoux test recommended; multiple puncture test lacks adequate specificity and sensitivity
 - a. All children should have a routine assessment of their risk of exposure to TB; risk assessment should be done every 6 months until 2 years old; after 2 years, perform annual risk assessment; skin test all children with increased risk
 - b. Skin test immediately any child identified as a contact of a person with confirmed or suspected infectious TB, any child with clinical or radiographic findings suggestive of TB, any child immigrating from an endemic area (Asia, Middle East, Africa, Latin America, countries of the former Soviet Union), including international adoptees, those traveling in endemic areas or having significant contact with persons from endemic areas
 - c. Skin test annually any child with HIV, an incarcerated adolescent, children with increased risk of progression of LTBI to tuberculosis disease
 - d. Skin test every 2 to 3 years any child exposed to high-risk persons (refer to #5 etiology/incidence section)
 - e. Skin test before initiation of immunosuppressive therapy
 - f. A positive TST indicates likely infection—a reaction of 15 mm or greater is considered positive in any population; reactions of 5 to 14 mm may be considered positive in certain high risk groups
 - 2. Chest radiograph findings vary from normal to lobar or segmental parenchymal lesion, or collapsed consolidation, lymphadenopathy, pleural effusion, or miliary disease (characterized by snowflake appearance)
 - 3. Recent availability of interferon-gamma release assays (IGRAs) that test for the presence of *M. tuberculosis* have become available in the U.S. in the form of QuantiFERON-TB Gold; IGRA testing is more specific than TST
 - a. Recommendation for IGRAs—immunocompetent children > 5 years, in place of TST to confirm TB; fewer false-positive test results
 - b. Positive result from an IGRA indicates *M. tuberculosis* infection
 - c. Negative result—not necessarily absence of infection
 - d. Useful to determine whether a BCG-immunized child with a reactive TST more likely has LTBI or false-positive TST due to BCG
 - e. Not recommended for children < 5 years or immunocompromised (Red Book online, 2009)
- Management/Treatment
 1. Diagnosing a child with TB infection or disease represents recent transmission of TB in the community; every effort should be made to identify and treat the source case and others infected by the source case; all cases should be reported to local health department; epidemiological investigation, isolation, compliance with therapy, and evaluation of the resolution of disease are crucial in controlling spread of disease; prescribing provider and public health authorities are both responsible for treatment completion
 2. Referral to appropriate specialist
 3. Guidelines for chemotherapy vary widely depending on the sensitivity of the organism, the condition of the patient, and the location of disease; refer to most recent edition of AAP *Red Book* for a complete list of drug dosages and side effects as well as therapeutic regimens; commonly used medications are ethambutol, isoniazid, rifampin, pyrazinamide
 4. For LTBI, 9 months treatment with isoniazid, once a day, is regimen of choice; prolonged therapy for immunocompromised conditions

5. For newborns exposed to household contact infected with TB, recommendations vary based on categorization of infection of household contact; refer to most recent edition of *AAP Red Book*
6. Directly observed therapy and patient-centered case management are recommended for treatment of people with tuberculosis disease; management plan should be tailored to promote adherence to the drug regimen
7. Children younger than 10 with primary pulmonary disease are usually not contagious; older children and adults suspected of having TB require TB isolation until proved noncontagious
8. Bacillus Calmette-Guérin (BCG) vaccine—live vaccine from attenuated strains of *Mycobacterium bovis*; recommended only in limited and select circumstances in U.S.; recommended internationally by World Health Organization for administration at birth to help prevent disseminated and other life-threatening diseases caused by *Mycobacterium tuberculosis*; indications, adverse reactions, contraindications, and interpretations of Mantoux skin tests in persons who have received BCG described in *AAP Red Book*
9. Chest radiographs after 2 months of therapy to evaluate medication response

■ QUESTIONS

Select the best answer

1. Shelby, a 4-week-old, presents to your office in mid-January with a one-week history of nasal congestion and occasional cough. On the evening prior to this visit Shelby developed a temperature of 102°F, refused to breastfeed, had paroxysmal coughing, and noisy, labored breathing. On exam, you note an ill-appearing infant who is lethargic with tachypnea and intercostal retractions. Shelby does not attend daycare but has a 3-year-old sibling who is in daycare and who recently had a “cold.” Considering the clinical presentation, what is the most likely cause of Shelby’s illness?
 - a. *Mycoplasma pneumoniae*
 - b. RSV bronchiolitis
 - c. Aspiration pneumonia
 - d. Streptococcal infection of the pharynx
2. In the above scenario, which of the following would be the treatment of choice?
 - a. Antihistamine, decongestant, and cough suppressant
 - b. Oral antibiotics and follow-up chest radiograph in two weeks
 - c. Bronchoscopy with lavage, chest physiotherapy, and respiratory isolation
 - d. Hospitalization, bronchodilators, supplemental oxygen, and nutritional support
3. Of the following children, which one should not have tuberculin skin testing?
 - a. Richard, a 14-year-old, whose uncle was recently granted parole after five years in prison and is currently living with Richard’s family
 - b. Theresa, a 2-year-old who was infected with RSV three months ago and is currently asymptomatic
 - c. Han, a 3-month-old whose family emigrated to the U.S. from Cambodia one month ago
 - d. Chris, an 18-month-old whose mother is infected with HIV
4. Which of the following clinical presentations least warrants sweat chloride testing?
 - a. 10-year-old female sibling of a patient newly diagnosed with cystic fibrosis; sibling is without pulmonary problems and growth parameters are at 50% for age
 - b. 2-year-old male with recurrent pneumonia and growth parameters at 5% for age
 - c. 4-year-old female with nocturnal cough, which resolves after treatment with bronchodilators and short-term steroids; growth parameters at 10% for age
 - d. 7-year-old female with nasal polyps, mildly hyperexpanded lungs, growth parameters at 25% for age
5. Of the following diagnostic findings, which one should be referred to a specialist immediately?
 - a. Suspected foreign body aspiration
 - b. Sweat chloride results of 30 mEq/L
 - c. Pulmonary function tests of 85% predicted
 - d. Chest radiograph with hyperexpansion
6. What is the most common agent for nonviral pneumonia from older preschool to young adulthood?
 - a. *Mycoplasma/Chlamydia aureus*
 - b. *Staphylococcus aureus*
 - c. *Ureaplasma*
 - d. *Haemophilus influenzae*
7. Which one of the following diagnoses would not be part of the differential for recurrent lobar pneumonia in a 2-year-old?
 - a. Cystic fibrosis
 - b. Foreign body aspiration
 - c. Atelectasis
 - d. Bronchitis

8. The most common clinical presentation of pneumonia includes:
 - a. Cough, fever, tachypnea, and abdominal pain
 - b. Hemoptysis, putrid breath, and weight loss
 - c. Sudden chest pain, cyanosis
 - d. Retractions, stridor
9. In addition to airway hyper-responsiveness and reversible airway obstruction, asthma is a chronic lung disease characterized by:
 - a. Bronchiectasis
 - b. Inflammation
 - c. Pleural effusion
 - d. Pulmonary edema
10. The most common trigger for an acute asthma episode in the very young child is:
 - a. Respiratory infections
 - b. Exercise
 - c. Tobacco smoke
 - d. Outdoor allergens
11. Luke has mild persistent asthma. Appropriate daily medication should include:
 - a. An inhaled low-dose corticosteroid
 - b. Short-acting β_2 agonists
 - c. An oral systemic corticosteroid
 - d. A cough suppressant
12. Which of the following is not a goal of appropriate asthma management?
 - a. Limited activity and exercise
 - b. Prevent recurrent exacerbations
 - c. Prevent chronic troublesome symptoms
 - d. Maintain near normal pulmonary functions
13. Deon is a 4-year-old male with a history of atopic dermatitis and recurrent pneumonias, according to his mother. He presents with a persistent nighttime cough. His most likely diagnosis is:
 - a. Asthma
 - b. Foreign body aspiration
 - c. Croup
 - d. Cystic fibrosis
14. The most typical chest radiographic finding consistent with the diagnosis of asthma is:
 - a. Normal chest film
 - b. Diffuse airway edema
 - c. Right upper lobe infiltrate
 - d. Hyperinflation
15. When providing asthma education regarding the use of a long-acting β_2 agonist, it is important to stress:
 - a. It should not be used as a quick-relief medication
 - b. May be given every 30 minutes times three for rescue therapy
 - c. May be most beneficial for exercise-induced asthma
 - d. Should never be taken while also using inhaled corticosteroids
16. Claire is an 8-year-old with moderate persistent asthma who is still having a daily cough. She reports three times a day use of a short-acting inhaled β_2 agonist and cromolyn sodium at her clinic visit. Your management plan should be altered to include:
 - a. Broad-spectrum antibiotics and recheck in two weeks
 - b. Addition of systemic corticosteroids for five days
 - c. Replace cromolyn sodium with inhaled corticosteroids
 - d. Addition of an inhaled anticholinergic
17. Ben is a 10-year-old who has recently been diagnosed with mild intermittent asthma. Which of the following is not a routine part of his clinic management?
 - a. Spirometry evaluation
 - b. Metered dose inhaler technique demonstration
 - c. Environmental triggers and control methods review
 - d. School excuse to not participate in physical education activities
18. Major contributors to asthma morbidity and mortality are:
 - a. Underdiagnosis and inappropriate treatment
 - b. An increase in indoor allergens
 - c. Overuse of anti-inflammatory medications
 - d. An increase in air pollution
19. The primary treatment for bronchopulmonary dysplasia is:
 - a. Pancreatic enzymes
 - b. Surgical repair
 - c. Adequate oxygenation
 - d. Chest physiotherapy
20. The single most predictive factor in the development of bronchopulmonary dysplasia is:
 - a. Birth weight
 - b. Maternal age
 - c. Maternal educational level
 - d. Respiratory infections

21. The classic radiographic finding in croup is:
- Hyperinflation
 - Perihilar lymphadenopathy
 - Thumb sign
 - Steeple sign
22. Unilateral wheezing is a finding suggestive of:
- Croup
 - Asthma
 - Foreign body aspiration
 - Cystic fibrosis
23. Which of the following is not characteristic of an apparent life-threatening event (ALTE):
- Change in muscle tone
 - Fever
 - Change in skin color
 - Apnea
24. Following an ALTE, management and treatment are based on findings from:
- A thorough history and physical exam
 - An electroencephalogram
 - A chest radiograph
 - A sleep study
25. The predominant characteristic of a young infant with bronchopulmonary dysplasia is:
- Prolonged fevers
 - Hypoxemia on room air
 - Recurrent pneumonias
 - Chronic hypoinflation

■ ANSWERS

- | | |
|--------------|--------------|
| 1. b | 14. d |
| 2. d | 15. a |
| 3. b | 16. c |
| 4. c | 17. d |
| 5. a | 18. a |
| 6. a | 19. c |
| 7. d | 20. a |
| 8. a | 21. d |
| 9. b | 22. c |
| 10. a | 23. b |
| 11. a | 24. a |
| 12. a | 25. b |
| 13. a | |

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Dermatologic Conditions

Jeanne Steman Findlay

■ NEWBORN EXANTHEMA

Cutis Marmorata

- Definition: Transient mottling of the neonate's skin with a lacy, bluish appearance
- Etiology/Incidence
 1. Physiologic response of uneven blood flow which results in constriction of small blood vessels while others dilate
 2. Often precipitated by exposure to cold
 3. More common in premature infants
 4. Persistence after neonatal period found in Down syndrome
- Signs and Symptoms: Generalized lacy, reddish-blue appearance to the skin
- Differential Diagnosis
 1. Cutis marmorata telangiectatica congenita
 2. Cyanosis
 3. Erythema toxicum neonatorum
- Physical Findings: Generalized reddish-blue, reticulated pattern to most of body surface
- Diagnostic Tests/Findings: None
 1. 50% of these patients have one or more congenital skin conditions which include glaucoma, hemangiomas, or vascular malformations

- Management/Treatment
 1. Keep neonate at stable temperature
 2. Reduce exposure to cold environment
 3. Don't overdress or keep environment overly warm

Erythema Toxicum Neonatorum

- Definition: Transient, benign, self-limited skin rash with lesions of varied morphology; erythematous macules; wheals, vesicles, and pustules
- Etiology/Incidence:
 1. Unknown cause
 2. Occurs in 50% to 60% of neonates
 3. More common in full-term and post-term neonates
 4. More common in neonates with birth weight > 2500 g
 5. Onset usually within first 24 to 48 hours of life, but occasionally present at birth
- Signs and Symptoms
 1. Yellow-white lesions on reddish-pink base; may be blotchy
 2. Extent of rash varies from minimal to most of body surface; palms and soles are usually spared
- Differential Diagnosis
 1. Congenital candidiasis
 2. Incontinentia pigmenti
 3. Miliaria rubra
 4. Sterile transient neonatal pustular melanosis

5. Urticaria pigmentosa
 6. Bacterial infestation—change to “infection”
 7. Folliculitis
 8. Herpes Simplex
- Physical Findings
 1. Lesions of varied morphology—erythematous macules 2 to 3 cm in diameter appear first, followed by wheals, vesicles, and rarely pustules
 2. Lesions usually arise from erythematous base with macular erythema fading within 2 to 3 days
 3. Occurs predominately on the trunk, however, may occur anywhere on body except soles and palms
 4. Number of lesions varies from few to many
 5. Spontaneous resolution in 5 to 7 days
 - Diagnostic Tests/Findings: Wright’s stained smear of pustules identifies predominance of 90% eosinophils rather than neutrophils, which rules out neonatal pustular melanosis; consider bacterial and fungal cultures
 - Management/Treatment
 1. Obtain detailed history of onset, duration, and progression
 2. Describe and monitor lesions in terms of morphology/structure, size, shape, number, color, location, distribution
 3. No treatment necessary
 4. Educate regarding characteristics of condition and expected resolution
 5. Refer to dermatologist for evaluation if condition does not improve

Milia

- Definition: Benign and common condition of small, yellow-white, 1–2 mm sized inclusion cysts filled with cheesy keratinous material on face of newborn
- Etiology/Incidence
 1. Caused by superficial keratinous material accumulated within developing pilosebaceous follicle
 2. Occurs in 50% of newborns
- Signs and Symptoms: Numerous small, yellow-white raised lesions on face of newborn
- Differential Diagnosis
 1. Sebaceous hyperplasia
 2. Calcinosi cutis (especially in Down syndrome infants)
 3. Acne vulgaris
 4. Syringoma

- Physical Findings
 1. Numerous firm, pearly yellow-white, 1 to 2 mm inclusion papular cysts on the cheeks, forehead, and nose; predominately on face; may be found on other body surfaces
 2. Oral counterpart are yellow, papular lesions on hard palate known as Epstein’s pearls
 3. Condition resolves spontaneously without treatment within a few weeks as lesions exfoliate
- Diagnostic Tests/Findings: None
- Management/Treatment
 1. Describe and monitor lesions in terms of morphology/structure, size, shape, number, color, location, distribution
 2. No treatment necessary
 3. Educate regarding characteristics of condition and expected resolution

■ VASCULAR LESIONS

Salmon Patch (Nevus Simplex)

- Definition: Benign, flat, light red to orange vascular birthmark on head and face
- Etiology/Incidence
 1. Caused by overgrowth of blood vessels within dermis skin layer
 2. Seen in approximately 40% to 50% of newborns
 3. More common in girls
- Signs and Symptoms: Flat, light red to orange lesions on face and head of newborn
- Differential Diagnosis
 1. Port-wine stain (Nevus flammeus)
 2. Hemangioma
 3. Contact irritation or chronic rubbing
 4. Child abuse
- Physical Findings
 1. Single or multiple irregular, light red to orange macular lesions on eyelids, nape of neck, glabella, and/or occiput; vary in size
 2. Lesions gradually fade and disappear spontaneously with time
 - a. Eyelid lesions fade first, resolving completely within 3 to 6 months
 - b. Nape of neck lesions fade but may persist into adulthood
 - c. Other lesions resolve completely by 7 years of age

- Diagnostic Tests/Findings: None
- Management/Treatment
 1. Describe and monitor lesions in terms of morphology/structure, size, shape, number, color, location, distribution
 2. No treatment necessary
 3. Educate regarding characteristics of condition and expected resolution

Port-wine Stain (previously termed Nevus Flammeus)

- Definition: Benign, permanent, flat, dark red to purple vascular lesion, predominantly on head and face
- Etiology/Incidence
 1. Caused by proliferation of dilated capillaries in the dermis
 2. Lesions may be associated with other conditions
 - a. Lesions covering entire half of face or bilateral; may be associated with Sturge-Weber syndrome
 - b. Lesions on extremities may be associated with hypertrophy of soft tissue and bone
 - c. Lesions on the back, especially crossing the midline, may be associated with defects in the spinal cord and vertebrae
 3. Seen in approximately 0.4% of newborns
- Differential Diagnosis
 1. Nevus Simplex
 2. Hemangioma
 3. Child abuse
- Physical Findings
 1. Irregular dark red or purple macular lesions occurring on any body surface, predominately on face and head
 2. Size varies from less than 1 cm to more than 20 cm
 3. May initially appear pink in infancy and gradually become darker
 4. Lesion never fades and becomes thickened and raised in adulthood
- Diagnostic Tests/Findings: If there is uncertainty about the diagnosis, pediatric ophthalmology examination and ultrasoundography < 5 months old
- Management/Treatment
 1. Describe and monitor lesions in terms of morphology/structure, size, shape, number, color, location, distribution

2. Refer for dermatologist evaluation to rule out Sturge-Weber syndrome and other associated conditions
3. Treatment of choice: is b, with a and c as adjuncts
 - a. Refer for dermatologist evaluation for consideration of pulsed dye laser treatment, which is recommended to start as early as possible in infancy; definitely before one year of age
 - b. May be camouflaged later in childhood with water resistant cosmetics
 - c. Counseling as needed for related psychological concerns
4. Educate regarding characteristics of condition

Capillary Hemangioma (Strawberry Nevus)

- Definition: Bright red or blue-red nodular tumors of varying sizes and shape with a rubbery and rough surface predominately on head and face
- Etiology/Incidence
 1. Caused by proliferation of capillary endothelial cells which may be superficial or deep
 2. Seen in approximately 2.5% of newborns, although may not be present at birth
 3. More common in girls
 4. More common in light-skinned and premature infants
- Signs and Symptoms: Red or blue-red lesions on skin surface
- Differential Diagnosis
 1. Venous malformation
 2. Cystic hygroma
 3. Neonatal hemangiomatosis
 4. Blue rubber bleb nevus syndrome
- Physical Findings
 1. Often is not present at birth, however, area of eventual lesion is blanched or slightly colored
 2. Size varies from less than 1 cm to over 4 cm
 3. Pattern of growth and resolution
 - a. Grows quickly within 2 to 4 weeks to a red or blue-red, protuberant, rubbery nodule or plaque
 - b. Most growth the first six months
 - c. Gradual reduction in proliferation usually begins between 9 to 12 months with gray areas developing, followed by flattening from center to periphery
 - d. A flat or involuted area of hyperpigmentation often remains following dissolution of the lesion

4. Complications may occur resulting from location and depth of lesion (size does not determine risk of complication)
 - a. Lesions involving eye area and orbit may cause visual disturbances
 - b. Lesions of head and neck may be associated with subglottic hemangiomas causing airway obstruction
 - c. Lesions may cause cardiovascular disturbances through compression
 - d. Lesions may ulcerate as they involute
 5. Complication of thrombocytopenia may occur resulting from trapped platelets within lesion
 6. Lesions resolve spontaneously and completely disappear with age
 - a. 50% are cleared by 5 years of age
 - b. 90% are cleared by 10 years of age
 - c. Remainder clear during adolescence
- Diagnostic Tests/Findings: None
 - Management/Treatment
 1. Describe and monitor lesions in terms of morphology/structure, size, shape, number, color, location, distribution
 2. Refer for dermatologist evaluation to rule out involvement with vital organs
 3. Treatment depends on location and complications; if ulcerates, pulsed dye laser may be needed; oral prednisone was the treatment of choice until recently with oral propranolol protocols being established to replace oral steroids; surgical intervention is rarely indicated
 4. Educate regarding characteristics of condition and expected resolution

■ MELANOCYTE CELL AND PIGMENTATION CONDITIONS

Café au lait Spots

- Definition: Light to medium brown pigmented macular lesions of varying sizes and shapes found anywhere on the body; the color of coffee with milk, from which the name is derived
- Etiology/Incidence
 1. Caused by increased pigmentation activity of melanocyte cells
 2. Overall incidence is higher in dark-skinned populations than light-skinned
 3. Lesions larger than 1.5 cm occur in 10% of light-skinned population and 20% of darker-skinned populations
 4. Lesions are usually present at birth however, may develop at any age

5. Lesions are present throughout life, however, color intensity may fade
6. Six or more lesions and/or lesions larger than 1.5 cm in diameter may be associated with neurofibromatosis or Albright's syndrome (refer to Neurological Disorders Chapter 11)

- Signs and Symptoms: Flat light brown lesions on skin; may be deeper in color in dark-skinned populations
- Differential Diagnosis: None
- Physical Findings
 1. Macular light to medium brown lesions on any skin surface
 2. Size varies from less than $\frac{1}{2}$ cm to 20 cm in diameter
 3. May be single or multiple
 4. Vary in shape, frequently oval
 5. Six or more lesions and/or lesions larger than 1.5 cm may be associated with neurofibromatosis or Albright's syndrome
- Diagnostic Tests/Findings: None
- Management/Treatment
 1. Describe and monitor lesions in terms of morphology/structure, size, shape, number, color, location, distribution
 2. If suspected that lesions may be associated with any other condition, refer to dermatologist for further evaluation
 3. No treatment necessary
 4. Educate regarding characteristics of condition

Mongolian Spots

- Definition: Blue-black and gray macular lesions of irregular shape and varying sizes; usually on sacrococcygeal region, buttocks, and lumbar areas but may also involve extremities, upper back and shoulders
- Etiology/Incidence
 1. Lesions consist of migrating spindle-shaped pigmented/melanocyte cells deep within dermis layer
 2. Occurs in 90% of darker-skinned infants; 5% of light-skinned infants
- Signs and Symptoms: Blue-black or gray lesions of irregular shape and varying size on lower aspect of back
- Differential Diagnosis: Child abuse

- Physical Findings
 1. Blue-black or gray macular lesions of irregular shapes
 2. Vary in size from < 2 cm to > 10 cm
 3. Located on dorsal body surface, predominantly on sacrococcygeal area of buttocks and lumbar areas, but also on upper back, shoulders, and extremities
 4. Lesions not seen on palms or soles
 5. Lesions resolve spontaneously without treatment
 - a. Most fade completely during childhood and adolescence
 - b. Some may still be evident in adulthood
- Diagnostic Tests/Findings: None
- Management/Treatment
 1. Describe and monitor lesions in terms of morphology/structure, size, shape, number, color, location, and distribution
 2. No treatment necessary
 3. Educate regarding characteristics of condition and expected resolution

Malignant Melanoma

- Definition: Lethal form of skin cancer involving melanocyte cells; may occur on any skin surface
- Etiology/Incidence
 1. Caused by abnormal growth within melanocyte cells
 2. Severe sunburn or excessive exposure to the sun before the age of 10 years predisposes developing melanoma later in childhood or in adult life
 - a. Sun damaged skin cells may be dormant for years
 - b. Melanocyte cells provide mechanism to activate malignant process
 3. Melanoma cells spread through the lymphatic system and invade other skin surfaces and organs
 - a. 90% survival rate with localized condition
 - b. 20% survival with metastasis
 4. Increasing incidence in general population
 5. In the U.S. it is more common in females from birth to 40 years of age and light-skinned individuals
 6. Increased incidence with family history
 7. More lethal and faster growing than basal cell or squamous cell cancers
- Signs and Symptoms
 1. Localized change in skin color or increase in size of existing nevus

2. May have itching with bleeding and tenderness

- Differential Diagnosis: Other skin cancers
- Physical Findings
 1. Asymmetrical lesion with irregular, ragged and blurred borders
 2. Uneven color with shades of blue, black, brown, tan, and red; all colors may exist within same lesion
 3. More common on arms and lower legs of females and on chest of males
 4. Single or multiple (clusters) lesions may be found in distant areas with metastasis
 5. Bleeding and ulceration—usually late signs
- Diagnostic Tests/Findings: Skin biopsy confirms diagnosis
- Management/Treatment
 1. Obtain detailed history of onset, duration, and progression
 2. Refer to dermatologist for evaluation immediately if suspected; surgical excision is indicated
 3. Educate regarding characteristics of condition, treatment, and expected prognosis
 4. Educate regarding specific preventive measures
 - a. Protect skin from exposure to sunlight
 - (1) Cover-up clothing and hats
 - (2) Sunglasses
 - (3) Water resistant sunblocks that protect against UVB and UVA ultraviolet light with > 30 sun protection factor (SPF)
 - b. Avoid exposure to sunlight especially during 10 a.m. to 3 p.m.
 - c. Avoid sun lamps and tanning booths
 - d. Teach the ABCDEs of pigmented lesions: Asymmetry, Borders, Color, Diameter, and Evolution of lesions, along with checking for the “ugly duckling” lesion that doesn’t resemble any other pigmented skin lesions; recommend monthly mole checks at home for high risk individuals

Albinism

- Definition: Inherited congenital defect of total or partial lack of pigmentation in which affected body parts lack normal color; condition is present at birth; there are 4 main types:
 1. Total form—affects entire skin, hair, and retina
 2. Partial or localized forms—confined to specific area of skin, hair (forelock of hair), or eyes (pupil or retina)

- Etiology/Incidence
 1. Metabolic process within melanocyte cells required for melanin production is impaired—melanin, giving skin its distinctive color, is not secreted
 2. Incidence equal between males and females with type 2 more common in African-American population and type 1 only confirmed in African Americans
- Signs and Symptoms
 1. Milky-white skin (localized or generalized)
 2. Light sensitivity
- Differential Diagnosis
 1. Homocystinuria
 2. Phenylketonuria
- Physical Findings
 1. Skin is milky-white, hair is white or yellow, iris is usually blue, pupil usually appears red and becomes darker in adulthood
 2. Skin is sensitive to light and sunburns easily
 3. Other symptoms not involving skin include decreased visual acuity, photosensitivity
- Diagnostic Tests/Findings: None
- Management/Treatment
 1. Describe skin and areas of hypopigmentation and monitor routinely for any skin changes that may occur including development of lesions
 2. Educate regarding need to protect from exposure to sunlight
 - a. Cover-up clothing and hats
 - b. Sunglasses
 - c. Water resistant sunblocks that protect against UVB and UVA ultraviolet light with > 30 SPF
 3. Educate regarding characteristics and prognosis of condition
 4. Counsel as indicated regarding:
 - a. Related psychologic effects
 - b. Genetic counseling related to potential inheritance factors
 5. Refer to dermatologist for evaluation if skin changes occur
 6. Refer to ophthalmologist for evaluation of vision and eye involvement

Vitiligo

- Definition: Acquired autoimmune condition involving patches of depigmentation on skin surfaces and in mouth and genitalia

1. Segmented form—unilateral involving two dermatomes
 2. Generalized form—involves more than two dermatomes, often has bilateral distribution
- Etiology/Incidence
 1. Unknown cause
 - a. Affected areas of hypopigmentation have loss or destruction of melanocyte cells
 - b. May be associated with autoimmune conditions—diabetes mellitus, Addison's disease, or thyroiditis
 2. Occurs in approximately 4% of all ethnic populations
 3. Onset is usually before 20 years of age
 - Signs and Symptoms: Milky-white patches on skin
 - Differential Diagnosis
 1. Post inflammatory hypopigmentation
 2. Pityriasis alba
 3. Idiopathic Guttate Hypomelanosis
 4. Pityriasis versicolor
 5. Nevus Anemicus
 - Physical Findings
 1. Milky-white macular patches of depigmentation with sharply demarcated borders occur in unilateral or bilateral pattern on skin of normal texture
 2. Shape varies from round, oval, to irregular
 3. Size varies from less than 2 cm to well over 20 cm
 4. Varies in number from one to many
 5. Condition is often permanent without repigmentation
 - Diagnostic Tests/Findings: None
 - Management/Treatment
 1. Obtain detailed history of onset, duration, severity and progression, and possible precipitating factors
 2. Describe skin and areas of depigmentation and monitor for any skin changes that may occur including development of lesions
 3. Protect skin from exposure to sunlight especially during 10 a.m. to 3 p.m.
 - a. Use cover-up clothing, hats, and sunglasses
 - b. Apply water resistant sunblocks that protect against UVB and UVA ultraviolet light with > 30 SPF
 4. Refer for dermatologist evaluation and treatment to stimulate repigmentation
 - a. Topical steroid applications and controlled ultraviolet light exposure

- b. Repigmentation efforts have varying degrees of success especially for involved areas on extremities
- c. Excimer laser FDA approved for treatment of vitiligo
- 5. Educate regarding characteristics and expected prognoses
- 6. Recommend camouflage with water resistant cosmetics for adolescent
- 7. Counsel regarding
 - a. Related psychological impact of condition
 - b. Serious need for protection to reduce risk for skin cancer and sunburn
- 8. Refer for dermatologist evaluation if complications develop

Pityriasis Alba

- Definition: Acquired condition of scaly hypopigmented, finely scaled macular lesions of varying sizes and shapes with indistinct borders occurring predominately on cheeks
- Etiology/Incidence
 1. Unknown cause
 2. May be associated with overdrying of skin causing inflammation and hypopigmentation
 3. Occurs most often in children ages 3 to 12 years
 4. More apparent in dark-skinned populations
- Signs and Symptoms
 1. Finely scaled white patches most commonly seen on cheeks
 2. May be pruritic
 3. May appear mildly erythematous
- Differential Diagnosis
 1. Psoriasis
 2. Tinea corporis
 3. Vitiligo
- Physical Findings
 1. Scaly hypopigmented lesions of varying sizes or shapes with nondistinct borders occurring predominately on cheeks, less commonly on other skin surfaces
 2. Some lesions may be slightly erythematous
 3. Number of lesions varies from one to many
 4. Exposure to sunlight may exacerbate lesions, making them more pronounced
 5. Repigmentation occurs as condition resolves spontaneously in 3 to 4 months
- Diagnostic Tests/Findings: KOH preparation to rule out tinea corporis

- Management/Treatment
 1. Obtain detailed history of onset, duration, severity and progression of symptoms, and possible precipitating factors
 2. Describe and monitor lesions in terms of morphology/structure, size, shape, number, color, location, distribution
 3. Educate child and parents regarding need to protect skin from exposure to sunlight, especially during 10 a.m. to 3 p.m.
 - a. Use cover-up clothing, hats, and sunglasses
 - b. Apply water resistant sunblocks that protect against UVB and UVA ultraviolet light with > 30 SPF
 4. Use bland moisturizer to reduce overdrying
 5. Educate regarding characteristics and expected prognoses
 6. Recommend camouflage with water resistant cosmetics for adolescent
 7. Refer for dermatologist evaluation if condition does not improve

▣ PAPULOSQUAMOUS CONDITIONS

Pityriasis Rosea

- Definition: Acquired common mild inflammatory condition characterized by scaly, hypopigmented, and hyperpigmented lesions predominately on the trunk, upper arms, and upper thighs
- Etiology/Incidence
 1. Unknown cause
 2. Possible viral association
 3. Occurs more often in fall and spring months
 4. Occurs especially in older children of all ethnic groups
- Signs and Symptoms
 1. Scaly pink marks on skin in light-skinned individuals; appears hyperpigmented on darker skin
 2. Periodic pruritus of varying degrees of severity especially at onset
 3. Possible prodrome of malaise and low grade fever before onset of rash
- Differential Diagnosis
 1. Pityriasis alba
 2. Seborrheic dermatitis
 3. Secondary syphilis
 4. Tinea corporis
 5. Guttate psoriasis

- Physical Findings
 1. Scaly, hyperpigmented pink to salmon to violaceous lesions with progressive pattern
 - a. “Herald” patch of 1 cm to 5 cm on trunk or buttocks, usually occurs 5 to 10 days before generalized rash
 - b. Round and oval scaly, macular lesions develop over two-week period on skin lines and in parallel fashion suggestive of a Christmas tree pattern
 - c. Individual lesions clear in central to peripheral pattern
 2. On darker-skinned populations, lesions are more predominant on neck, axillary and inguinal regions
 3. Condition is self-limiting and resolves spontaneously in 3 to 4 months
- Diagnostic Tests/Findings
 1. KOH test to rule out tinea corporis
 2. VDRL to rule out secondary syphilis, especially in sexually active individuals
- Management/Treatment
 1. Obtain detailed history of onset, duration, severity and progression of symptoms, and possible precipitating factors
 2. Describe and monitor lesions in terms of morphology/structure, size, shape, number, color, location, distribution
 3. Educate regarding characteristics of condition and prognosis
 4. Use symptomatic treatment for pruritus
 - a. Topical calamine lotion on lesions
 - b. Oral antipruritic agents for severe pruritus, e.g., diphenhydramine
 - c. Cool bath or compresses on lesions
 - d. Low potency steroid creams
 5. Educate regarding medication dosage, signs of irritation, sensitivity
 6. Use controlled and limited sunlight exposure to shorten resolution time
 7. Refer for dermatologist evaluation if condition worsens or does not resolve

Psoriasis

- Definition: Acquired chronic, relapsing inflammatory condition characterized by erythematous plaques with silver-gray-white scales
 1. Psoriasis vulgaris—large plaques occurring predominately on elbows and knees
 2. Psoriasis guttate—small patches occurring predominately on trunk, upper arms, and thighs
- Etiology/Incidence
 1. Specific cause is unknown
 2. Associated with overproduction and too rapid migration of epithelial cells to skin surface; cells migrate in 3 to 4 days in comparison to usual 28 days
 - a. Psoriasis vulgaris—often associated with constant rubbing, or with trauma to the affected area known as Koebner’s response
 - b. Psoriasis guttate—often follows streptococcal infection
 3. Occurs in over 33% of children
 4. Up to 20% of people with psoriasis have psoriatic arthritis
 5. More common in light-skinned than dark-skinned populations
 6. Positive family history in approximately $\frac{1}{3}$ of cases strongly suggestive of a genetic connection
- Signs and Symptoms
 1. Silvery, gray-white scaling of skin, mainly on trunk or extremities, especially elbows and knees; less commonly on scalp and face
 2. Bleeding may occur if scales are picked at or removed
 3. Nails may be dystrophic with thickening with pits and ridges
- Differential Diagnosis
 1. Atopic dermatitis
 2. Drug eruptions
 3. Pityriasis rosea
 4. Seborrhea
 5. Secondary syphilis
 6. Tinea corporis
- Physical Findings
 1. Psoriasis vulgaris—large 5 to 10 cm plaques with thick silvery-white scales located on elbows and knees
 2. Psoriasis guttate—small 3 to 10 mm multiple teardrop, round or oval papules and patches which become covered by a silvery-gray-white scale on trunk and proximal extremities
 3. Bleeding occurs when scale is removed
 4. Nail plates may be thicker and show signs of pits, ridges, “oil spots” which are yellow discolorations of the nail plate; onycholysis: not all nail plates are involved
- Diagnostic Tests/Findings
 1. VDRL to rule out secondary syphilis
 2. KOH to rule out fungal infections

- Management/Treatment
 1. Obtain detailed history of onset, duration, severity, and progression of symptoms, and possible precipitating factors
 2. Describe and monitor lesions in terms of morphology/structure, size, shape, number, color, location, distribution
 3. Reduce hypertrophy of lesion
 - a. Use controlled and limited sunlight exposure
 - b. Apply topical steroids, e.g., hydrocortisone, triamcinolone
 - c. Apply mineral oil and moisturizers at least BID to decrease drying
 4. Educate regarding medication dosage, signs of irritation, sensitivity
 5. Educate regarding characteristics of condition and prognosis
 6. Refer for dermatologist evaluation if condition does not improve

■ DERMATITIS CONDITIONS

Atopic Dermatitis

- Definition: Common skin disorder with lesions of varied morphology commonly known as eczema; it is called “the itch that rashes”
 1. Acute form—occurs predominately in infants
 2. Chronic form—occurs predominately in children and adolescents
- Etiology/Incidence
 1. Specific cause is unknown
 2. May be associated with a disorder of immunity in some cases due to elevated levels of IgE; it is primarily a disease with an altered skin barrier function
 3. Positive family history may be predisposing factor in some cases
 4. Occurs in approximately 10% to 15% of children
 5. Up to 50% of affected infants develop asthma and/or other respiratory manifestations, e.g., allergic rhinitis, hay fever, and progress to chronic form
 6. Up to 25% of children and adolescents continue to have symptoms throughout adulthood
- Signs and Symptoms
 1. Skin changes are acute and chronic with xerosis (dry skin)
 - a. Infant—erythematous, itchy, easily irritated scaly patches
 - b. Older children—more focal pruritic patches in the antecubital and popliteal creases
 - c. Chronic skin changes include hyperpigmentation and lichenification
 2. Pruritus for both, worsens with sweating and temperature extremes
- Differential Diagnosis
 1. Contact dermatitis
 2. Psoriasis
 3. Seborrheic dermatitis
 4. Scabies
 5. Tinea corporis
 6. Impetigo or other secondary bacterial infection
- Physical Findings
 1. Acute form in infants usually develops between ages 2 weeks to 6 months with 50% cases resolving by 3 years and remainder progressing to chronic form
 - a. Lesions appear as erythematous, scaly patches of skin on face, head, trunk, and extensor surfaces
 - b. Lesions of varied morphology, e.g., xerotic, scaly, erythematous papules, sometimes with excoriations, oozing and crusting are present in various locations
 2. Chronic form develops with poor skin management and personal and family history of atopy; may continue into adulthood
 - a. Skin is hyperpigmented, leathery, and lichenified in the flexor surfaces of the neck, antecubital areas, wrists, popliteal area, ankle, fingers, and toes
 - b. Scratch marks on affected areas
 3. Other findings include:
 - a. Circles under eyes “allergic shiners”
 - b. Facial pallor
 - c. Nasal crease on top nose from frequent rubbing
 - d. Dry scalp
 - e. Prominent Dennie’s creases
 4. Pustules may be present as sign of secondary bacterial infection
- Diagnostic Tests/Findings
 1. No specific test confirms diagnosis—serum level of IgE may support diagnosis in some cases
 2. Skin scraping to rule out scabies
- Management/Treatment
 1. Obtain detailed history of onset, duration, severity/progression of symptoms, and possible precipitating factors
 2. Describe and monitor lesions in terms of morphology/structure, size, shape, number, color, location, distribution

3. Treat secondary infections if present
 - a. Oral antibiotics, e.g., bactrim, cefaclor, cefadroxil, cephalexin, clindamycin
 - b. Topical antibiotics for localized infection—mupirocin or altobax; others may lead to sensitivity reactions
4. Reduce and prevent nocturnal pruritus with oral antipruritics, e.g., hydroxyzine, diphenhydramine; nonsedating antihistamines during the day only if comorbid environmental or seasonal allergies suspected
5. Use topical steroids to reduce inflammation, immune response, and pruritus, e.g., hydrocortisone, triamcinolone
6. Rehydrate skin with daily lukewarm baths
 - a. Wet compresses applied over bland emollients helpful to manage chronic eczema or during eczema flares
 - b. Avoid skin-drying agents such as harsh soaps, perfumes, lotions
 - c. Apply cream emollients and lubricants at least BID, e.g., petroleum jelly
7. Educate regarding medication dosage, signs of irritation, sensitivity
8. Use mild soaps for general bathing and hygiene habits
9. Eliminate exposure to all substances and agents that may dry or irritate the skin and exacerbate condition; individually determined
 - a. Soaps, perfumes, hand and body lotions, makeup, household cleaning agents, bleach, chlorine, turpentine
 - b. Materials and fabrics such as wool, feathers, polyesters, stuffed animals and other fabric toys
 - c. If there is a positive family history or correlation with increased skin symptoms, eliminate suspected food products such as cow's milk, eggs, nuts, citrus fruits
 - d. If there is a positive family history of correlation with increased skin symptoms, minimize exposure to pets and other animals
 - e. Dust and dust mites
10. Monitor environment
 - a. Maintain cool temperature to reduce sweating
 - b. Increase humidity during cold winter months
11. Educate regarding characteristics of condition and expected prognosis
12. Refer for dermatologist evaluation if condition does not resolve

Contact Dermatitis (Allergic Contact Dermatitis)

- Definition: Allergic response to local contact with an allergen manifested by development of skin eruptions at site of contact
- Etiology/Incidence
 1. Caused by hypersensitivity to an allergen
 - a. Initial contact—allergic response usually delayed for several days
 - b. Re-exposure—allergic response usually occurs within 24 hours due to prior sensitization
 2. Numerous substances are associated with producing hypersensitivity reactions in sensitive individuals with the most common including:
 - a. Perfumes, soaps, cosmetics, fabric dyes
 - b. Topical medications, e.g., neomycin
 - c. Animal products—animal dander, feathers, fur, wool, leather
 - d. Plastics, synthetics—latex, rubber
 - e. Plants—poison sumac/ivy/oak
 - f. Metals—jewelry, clothing snaps, and belt buckles; especially nickel
- Signs and Symptoms
 1. Erythema and edema at site of contact
 2. Pruritus with varying degrees of intensity
 3. Vesicle and bulla formation
- Differential Diagnosis
 1. Bacterial infection
 2. Candida
 3. Diaper dermatitis
 4. Seborrhea dermatitis
 5. Impetigo
 6. Herpes simplex
- Physical Findings
 1. Erythema and edema with development of lesions of varying morphology—papules, vesicles, and denudation
 2. Lesions confined to area of direct contact with allergen
 3. Pruritus with varying degrees of intensity
 4. Excoriation/scratch marks and bleeding
 5. Chronic exposure may produce areas of hyperpigmentation and lichenification
- Diagnostic Tests/Findings: Skin testing to determine allergen hypersensitivities after acute stage
- Management/Treatment
 1. Obtain detailed history of onset, duration, severity and progression of symptoms, and possible precipitating factors

2. Describe and monitor lesions in terms of morphology/structure, size, shape, number, color, location, distribution
3. Avoid contact with allergen if sensitivity is known; if unknown, consider skin testing after acute phase to determine allergen
4. Cool compresses of Burrow's solution to affected areas
5. Steroids to reduce inflammation, immune response, and pruritus
 - a. Apply topical steroids to affected areas, e.g., hydrocortisone, triamcinolone
 - b. Oral steroids for severe cases, e.g., hydrocortisone
6. Oral antihistamines for pruritus, e.g., hydroxyzine, diphenhydramine
7. Oral antibiotics if secondary infection present, e.g., erythromycin, dicloxacillin
8. Educate regarding medication dosage, signs of irritation, sensitivity
9. Educate regarding characteristics of condition and expected resolution
10. Refer to dermatologist for:
 - a. Evaluation if condition does not show improvement in 2 days
 - b. Consideration of skin testing for hypersensitivities after acute episode to identify specific allergens

Contact Irritant Dermatitis (Diaper Dermatitis)

- Definition: Common disorder of genital-perineal area due to skin breakdown; characterized by erythema, scale, and other skin lesions such as vesicles
- Etiology/Incidence
 1. Breakdown of skin associated with:
 - a. Exposure to chemical irritants in soaps, bleach, water softeners, skin lotions, diaper cleansing tissues
 - b. Excessive contact with urine, feces; lax hygiene habits (primary irritant)
 2. Occurs in over 95% of all infants
 3. Peak incidence is 9 to 12 months of age
 4. Monilial rash caused by *Candida albicans*
 5. May persist until completion of toilet training
- Signs and Symptoms
 1. Redness, sores in diaper area, blisters
 2. Fiery red rash with satellite lesions on lower abdomen or upper thighs
 3. May have general irritability and/or crying, especially after elimination
- Differential Diagnosis
 1. Atopic dermatitis
 2. Seborrheic dermatitis

3. Intertrigo
4. Allergic/contact dermatitis
5. Psoriasis
6. Secondary bacterial infection
7. Child abuse

- Physical Findings
 1. Erythema with varying degrees of severity which may be generalized to entire area or localized to small area
 2. Lesions of varied morphology may develop—papules, vesicles, crusts, erosions, and ulcerations
 3. Pustules may be present signaling secondary bacterial infection
 4. Monilial rash—fiery red papular lesions within folds and on genitals; may also be pustular; may have associated oral thrush
 5. Poor genital hygiene may be present in some children
- Diagnostic Tests/Findings: No specific test confirms diagnosis
- Management/Treatment
 1. Obtain detailed history of onset, duration, severity and progression of symptoms, and possible precipitating factors
 2. Describe and monitor lesions in terms of morphology/structure, size, shape, number, color, location, distribution
 3. Treat secondary bacterial infection if present with topical antibiotics, e.g., mupirocin
 4. Treat present diaper dermatitis
 - a. Mild erythema—emollients to affected areas with each diaper change, e.g., petroleum jelly, zinc oxide
 - b. Erythema with papules—topical steroids, e.g., hydrocortisone, triamcinolone
 - c. Severe erythema and edema with papules, vesicles, and ulcerations—wet dressings may be soothing, e.g., Burrow's compresses; topical antibiotics may be indicated
 - d. Monilial rash—topical nystatin, clotrimazole, ketoconazole; oral nystatin for thrush
 - e. Avoid occlusive diapers and plastic pants
 - f. Expose diaper area to air as often as possible
 - g. Use appropriate preventive measures
 5. Educate regarding medication dosage, signs of irritation, sensitivity
 6. Preventive measures
 - a. Expose diaper area to air several times each day
 - b. Increase oral fluids to make urine less irritating
 - (1) Water for infant under 12 months
 - (2) Cranberry juice for older child

- c. Change diaper immediately after soiling
 - d. Wash diaper area with nonirritating agents after each diaper change, e.g., mild soap and water
 - e. Avoid occlusive diapers and plastic pants
 - f. Diaper selection and care
 - (1) Home laundry—mild soap and double rinse using 1 ounce of vinegar per gallon of water in last rinse
 - (2) Disposable diapers—select alternate brand if sensitivity occurs
 - g. “Diaper-wipes,” may need to be avoided because of irritation or sensitivity
 - h. Use lubricating ointment when diaper area skin at each diaper change, e.g., petroleum jelly
7. Refer for dermatologist evaluation if no improvement within 2 to 3 days or if condition worsens

Seborrhea Dermatitis

- Definition: Inflammatory condition usually on sebum-rich areas such as the scalp and face
 - 1. Newborn and young infant—cradle cap
 - 2. Adolescents—dandruff
- Etiology/Incidence
 - 1. Associated with overproduction of sebum in areas abundant with sebaceous glands
 - 2. Increase in sebaceous gland activity may be connected with hormonal stimulation at times when hormonal influence is highest
 - 3. Occurs more often in spring and summer months
- Signs and Symptoms
 - 1. Newborns and infants—areas of erythema under yellow crusts and greasy scales on scalp, face, neck folds, and axillary creases
 - 2. Adolescents—white flakes and greasy scaling on scalp, forehead, eyebrows, and face; often pruritic
- Differential Diagnosis
 - 1. Atopic dermatitis
 - 2. Bacterial infection
 - 3. Candidiasis
 - 4. Irritant contact dermatitis
 - 5. Psoriasis
- Physical Findings
 - 1. Newborns and infants—areas of underlying erythema with yellow crusts and greasy scaling on scalp and face; in more severe cases lesions may be present on trunk and in diaper area
 - 2. Adolescents—white flakes and greasy scaling on scalp, forehead, eyebrows, and face;

severity varies from simple dandruff to extensive, giving appearance of psoriasis; mild underlying erythema may be present

- Diagnostic Tests/Findings: No tests necessary to confirm diagnosis
- Management/Treatment
 - 1. Obtain detailed history of onset, duration, severity and progression of symptoms, and possible precipitating factors
 - 2. Describe and monitor lesions in terms of morphology/structure, size, shape, number, color, location, distribution
 - 3. Treat existing condition
 - a. For infants, shampoo and wash affected areas with a non-perfumed baby shampoo or baby wash; for adolescents, use antiseborrheic soaps and shampoos
 - b. Mineral oil with brushing to loosen crusts prior to washing
 - c. Topical steroid lotions for extreme cases to reduce inflammation, e.g., hydrocortisone
 - 4. Educate regarding medication dosage, signs of irritation, sensitivity
 - 5. Educate regarding characteristics of condition and expected prognosis
 - 6. Refer for dermatologist evaluation if condition persists without improvement

■ BURN CONDITIONS

Burns

- Definition: Injury of skin from exposure to hot surfaces and agents
 - 1. Classified according to depth of injury to skin layers
 - a. First degree/superficial burns—involve epidermis layer only
 - b. Second degree/partial thickness burns—involve epidermis and part of dermis which may be superficial dermis or deep dermis
 - c. Third degree/full thickness burns—involve epidermis, dermis, and dermal appendages
 - 2. Classified also according to extent of affected area
 - a. Minor burns—less than 10% of body surface if burn is superficial and less than 2% if burn is partial or full thickness
 - b. Major burns—10% or more of body surface if burn is superficial and 2% or more if burn is partial or full thickness
 - c. Major burns—hands, feet, face, eyes, ears, and perineal burns are always considered major burns, regardless of extent of body surface affected

- Etiology/Incidence
 1. Caused by external exposure to hot chemicals, electrical and thermal substances, and materials including the sun, electrical cords and outlets, irons, flames, fireworks, hot water and foods, cigarettes, light bulbs
 2. Affected cells in epidermis, dermis, or subcutaneous skin layers are injured and no longer capable of providing protective, electrolyte storage, sensory, and other functions of normal skin cells
 3. Third leading cause of death in children and adolescents
 - a. More common in toddlers and males
 - b. Commonly occurs in kitchen in late afternoon during dinner preparation
 - c. Approximately 10% of burns are thought to be intentional in infant, toddler, and young child
- Signs and Symptoms: According to degree, appearance, and healing time
 1. Superficial—red, swollen, and dry areas with tenderness
 2. Partial thickness and superficial burns—red, swollen, moist, and blistered areas with tenderness
 3. Partial thickness and deep burns—white, dry areas with loss of sensation
 4. Full thickness burns—white, brown, black, swollen dry areas with loss of sensation
- Differential Diagnosis
 1. Child abuse
 2. Staphylococcal scalded skin syndrome
- Physical Findings
 1. Superficial burns—erythema, mild edema, dryness, tenderness, and general discomfort of affected areas
 2. Partial thickness and superficial burns—erythema, edema, moist, few vesicles/blisters may develop, sensitive to touch and air
 3. Partial thickness and deep burns—white, dry, decreased sensitivity to touch, pain, temperature, and may blanch with pressure
 4. Full thickness burns—white, brown, to black; swollen, dry; lack full touch, pain, temperature sensitivity
 5. Physical findings associated with secondary bacterial infection may be present
- Diagnostic Tests/Findings
 1. Electrolyte studies especially if burn is extensive
 2. Culture to determine causal agent if secondary bacterial infection is present
- Management/Treatment
 1. Obtain detailed history of onset, duration, severity and progression of symptoms
 2. Describe and monitor burn area in terms of morphology/structure, extent of burn area, location, distribution
 3. Inpatient hospital management for all children with major burns, suspected abuse, esophageal and airway burns, and/or injuries such as fractures
 4. Outpatient management for children in stable environment with minor burns
 - a. Partial thickness burn if, 10% of body surface area (BSA) or full thickness burn is, 2% BSA
 - b. Monitor daily healing process by documenting changes
 - c. Cool compresses to affected areas
 - d. Medication for pain control, e.g., acetaminophen, ibuprofen
 - e. Topical antimicrobial agents to prevent infection on open blistered areas, e.g., silver sulfadiazine (except on face due to potential for hyperpigmentation), mupirocin
 - f. Do not excise vesicles/blisters
 - g. Fluids to reduce possibility of dehydration, e.g., water, juices
 - h. Topical emollients to repair and maintain skin barrier, e.g., petroleum jelly
 5. Educate regarding need to protect skin from exposure to sunlight especially during 10 a.m. to 3 p.m.
 - a. Cover-up clothing, hats, and sunglasses
 - b. Water resistant sunblocks that protect against UVB and UVA ultraviolet light with > 30 SPF
 - c. Educate regarding myths about getting a “base tan”
 6. Educate regarding medication dosage, signs of irritation, sensitivity
 7. Educate regarding characteristics of condition and prognosis
 8. Educate regarding measures to prevent further burn episodes and injuries
 9. Refer for dermatologist evaluation if condition does not show improvement

Sunburns

- Definition: Thermal burn due to excessive sunlight exposure
- Etiology/Incidence
 1. Exposed skin results in altered cell function and properties
 - a. Inflammatory skin response with increased blood flow
 - b. Increased melanin production

2. Fair-skinned populations are most sensitive
 3. Other factors involving sensitivity include high altitude, nearness to equator, and exposure to sun during hours of 10 a.m. and 3 p.m. when UVB waves are strongest
- Signs and Symptoms
 1. Redness, swelling, blisters, and tenderness of sun-exposed areas
 2. Fatigue, chills, and headache after sun exposure
 - Differential Diagnosis
 1. Child abuse
 2. Photosensitivity from medications
 3. Systemic viral exanthema
 4. Systemic drug reaction
 - Physical Findings
 1. Dependent on degree of exposure and injury; develops within several minutes to several hours after exposure
 - a. First-degree burns—erythema and tenderness
 - b. Second-degree burns—increased intensity of erythema and tenderness with edema, some vesicles/blisters
 - c. Third-degree burns—increased intensity of erythema, tenderness, edema, and vesicles/blisters
 2. Systemic symptoms of malaise, fever, headache may be evident especially in younger child with second- and third-degree burns
 3. Epidermis cells scale and desquamate within 3 to 7 days after injury
 4. Exposed areas may become hyperpigmented with development of freckles and moles
 - Diagnostic Tests/Findings: None used to confirm diagnosis
 - Management/Treatment
 1. Obtain detailed history of onset, duration, severity and progression of symptoms, and precipitating factors
 2. Describe and monitor location, color, degree of burn, and symptoms
 3. Treat existing condition
 - a. Remove from sunlight exposure
 - b. Cool water or saline compresses to affected areas
 - c. Do not use warm or hot showers/baths
 - d. Increase oral fluids to prevent dehydration
 - e. Oral pain medications, e.g., acetaminophen, ibuprofen
 - f. Topical emollients for dry skin, e.g., petroleum jelly

4. Educate regarding medication dosage, signs of irritation, sensitivity
5. Educate regarding measures of prevention
 - a. Risk factors of sun exposure
 - (1) Teach early signs of skin cancer
 - (2) Teach regarding individuals most vulnerable to sun exposure
 - b. Use sun screens and blocks with 30 or greater SPF
 - (1) Apply at least 20 minutes before exposure
 - (2) Apply frequently if sustained exposure—every hour
 - (3) Use waterproof agents when in water
 - (4) Discontinue if sensitivity is suspected
 - (5) Avoid use in infants < 6 months
 - c. Use cover-up clothing and hats designed to block UVB waves
6. Refer for dermatologist evaluation if condition does not improve or becomes worse

■ BACTERIAL CONDITIONS

Cellulitis

- Definition: Localized acute infection often precipitated by an insect bite (spider, mosquito, flea) or trauma that penetrates the protective skin barrier
- Etiology/Incidence: Caused when surface streptococci, *Haemophilus influenzae*, or *Staphylococcus aureus* bacteria invade all skin layers—epidermis, dermis, and subcutis, after a break in the skin has occurred
- Signs and Symptoms
 1. Irregular-shaped areas of skin with redness, swelling
 2. Warm and tender to touch
 3. Fever, chills, and malaise may be present
- Differential Diagnosis
 1. Impetigo
 2. Furuncle
 3. Pyoderma gangrenosum
- Physical Findings
 1. Erythema and edema with ill-defined, irregular borders
 2. Tenderness and warmth
 3. Regional lymphadenopathy may be present
 4. Fever, chills, and malaise indicates systemic involvement
 5. Facial, periorbital, or orbital involvement is vulnerable to development of more severe conditions

- Diagnostic Tests/Findings: Blood culture to confirm causal agent
- Management/Treatment
 1. Detailed history of onset, duration, severity and progression of symptoms, and precipitating factors
 2. Describe and monitor
 - a. Affected skin areas in terms of morphology/structure, size, shape, color, location, distribution
 - b. Systemic signs and symptoms of fever, chills, and malaise
 3. Hospitalization for severe cases and those involving face and eyes
 4. Treat with intramuscular, intravenous, and/or oral antibiotics according to severity of condition, organism, and site of involvement
 - a. If streptococcus suspected—cefazolin, nafcillin
 - b. If *Haemophilus influenzae* suspected—amoxicillin
 - c. If *Staphylococcus aureus* suspected—dicloxacillin
 - d. If MRSA—bactrim or clindamycin
 5. Educate regarding medication dosage, signs of irritation, sensitivity
 6. Educate regarding characteristics of condition and expected prognosis
 7. Refer for dermatologist evaluation if condition shows no improvement

Impetigo

- Definition: Localized bacterial infection of skin often precipitated by insect bites (spider, mosquito, flea) or other trauma that breaks protective skin barrier; predominately involves face and less commonly other body surfaces including perineum
- Etiology/Incidence
 1. *Staphylococcus aureus* and streptococci bacteria invade epidermis after break in skin
 2. Children < 6 years old have a higher incidence than adults
 3. Bullous impetigo most common in neonates and infants; nonbullous impetigo most common in 2 to 5 year olds
 4. Highly communicable with incubation period of 1 to 10 days
 5. Autoinoculable
- Signs and Symptoms
 1. Itching and tenderness may be present
 2. Areas of erythematous swollen skin, blisters, and/or moist, honey-colored crusts

- Differential Diagnosis
 1. Eczema
 2. Herpes simplex
 3. Scabies
- Physical Findings
 1. Two major forms
 - a. Nonbullous—underlying erythema with vesicles that erupt, resulting in honey/serous colored crusts with erosion of epidermis
 - b. Bullous—underlying erythema with pustules and vesicles that erupt, resulting in smooth shiny appearance
 2. Regional adenopathy with tenderness
- Diagnostic Tests/Findings: Culture will confirm diagnosis and causative organism
- Management/Treatment
 1. Obtain detailed history of onset, duration, severity and progression of symptoms, and precipitating factors
 2. Describe and monitor lesions in terms of morphology/structure, size, location, distribution
 3. Apply compresses of Burrow's solution several times daily to aid in cleaning and removing crusts
 4. Apply topical antibiotics to areas of involvement, e.g., mupirocin
 5. Prescribe oral antibiotics according to specific bacterial cause
 - a. For staphylococci—dicloxacillin
 - b. For streptococci—penicillin or erythromycin
 - c. For MRSA, bactrim or clindamycin
 6. Educate regarding medication dosage, signs of irritation, sensitivity
 7. Educate regarding characteristics of condition, treatment regime, prognosis, and good hygiene for prevention
 8. Exclude from school and other public programs until treated for 48 hours due to high communicability
 9. Refer for dermatologist evaluation if condition does not improve

Staphylococcal Scalded Skin Syndrome

- Definition: Toxin-mediated systemic bacterial infection with skin manifestations
- Etiology/Incidence
 1. Caused by effects of toxin produced by *Staphylococcus aureus* bacteria
 2. Occurs any season
 3. More common in neonates and infants than older children
 4. Incubation is variable, commonly 3 to 10 days

- Signs and Symptoms
 1. May present with abrupt onset of fever, irritability, and general malaise
 2. Bright, red, painful rash; more pronounced around eyes, mouth, neck, underarms, elbow, groin, and knees
 3. Pain on pressure
 4. Blistering and/or scaling of skin
- Differential Diagnosis
 1. Streptococcal scarlet fever
 2. Kawasaki disease
 3. Stevens-Johnson syndrome
 4. Toxic epidermal necrolysis
 5. Burns
 6. Child abuse
 7. Drug toxicity
- Physical Findings
 1. Abrupt onset of fever and general malaise
 2. General exanthema with erythema and swelling; more pronounced in perioral, periorbital areas, flexure surfaces of neck, axilla, antecubital, groin, and popliteal areas
 3. Light pressure causes extreme pain and exfoliation of top epidermal layers
 4. After peeling, skin appears glistening and scalded
 5. Vesicles/bullae may occur in more toxic cases
- Diagnostic Tests/Findings
 1. Blood culture to confirm *Staphylococcus aureus*
 2. Culture secretions to confirm *Staphylococcus aureus*
- Management/Treatment
 1. Obtain detailed history of onset, duration, severity and progression of symptoms, and precipitating factors
 2. Describe and monitor in terms of morphology/structure, size, shape, number, color, location, distribution
 3. Hospitalization is indicated for all neonates; treat more severe cases with IV antibiotics and monitor fluid and electrolytes
 4. Outpatient management may be considered with less toxic cases if environment is stable
 - a. Oral antistaphylococcal antibiotics, e.g., dicloxacillin
 - b. Oral antipyretics and analgesics for fever and pain control, e.g., acetaminophen, ibuprofen
 - c. Increase fluids to maintain hydration and prevent dehydration, e.g., water, juices
 5. Educate regarding medication dosage, signs of irritation, sensitivity

6. Educate regarding characteristics of condition, treatment, and prognosis
7. Refer for dermatologist evaluation if condition does not improve

■ BACTERIAL CONDITIONS INVOLVING PILOSEBACEOUS UNIT

Acne Vulgaris

- Definition: Common, inflammatory, chronic skin disorder involving the pilosebaceous follicle unit
 1. Occurs predominately on the face, neck, chest, and upper back skin surfaces; less commonly in other areas
 2. Often occurs in cyclic periods of exacerbation and remission
- Etiology/Incidence
 1. Specific cause is unknown
 - a. Associated with breakdown of follicle wall
 - b. Cells combine with sebum and plug follicle
 - c. Enzymes from *Corynebacterium acnes* mix with trapped debris causing edema and irritation
 2. Proven factors which may contribute to acne development
 - a. Increased androgenic hormonal influence
 - b. Positive family history
 - c. Stress
 3. Unproven factors with questionable and unsubstantiated contribution
 - a. Food—nuts, eggs, cheese, chocolate, milk
 - b. Poor hygiene
 4. Affects more than 70% of adolescents with varying degrees of severity
 - a. Onset parallels puberty
 - b. More common in females
 - c. More males develop severe acne
 - d. More females experience continuation of acne into adult years
- Signs and Symptoms
 1. Open and closed comedones (“blackheads” and “whiteheads”)
 2. Soreness at site of lesions
 3. Post inflammatory hyperpigmentation and scars at site of previous lesions
- Differential Diagnosis
 1. Folliculitis
 2. Rosacea
 3. Tuberous sclerosis
 4. Perioral dermatitis
 5. Contact dermatitis
 6. Urticaria
 7. Allergic drug reaction

- Physical Findings
 1. Lesions of varying morphology
 - a. Mild acne—lesions are scattered covering small areas
 - (1) Open comedones/blackheads—lesions filled with dry oxidized sebum; brown in color
 - (2) Closed comedones/whiteheads—lesions filled with follicle cells and sebum
 - b. Moderate acne—lesions are more numerous covering large areas
 - (1) All lesions of mild acne
 - (2) Pustules—lesions filled with follicle cells, sebum, and white blood cells
 - c. Severe acne—lesions are much more numerous covering larger areas
 - (1) All lesions of mild and moderate acne
 - (2) Erythema with papules and pustules
 - (3) Nodules and cysts—deep dermal lesions filled with follicle debris, often with communicating tracks to other cysts
 2. Increased oiliness of hair and skin
 3. Scarring especially when:
 - a. Lesions at any stage have been manipulated and squeezed
 - b. Cysts have erupted deep within the dermis
 4. Signs of related psychological distress/depression may be present
- Diagnostic Tests/Findings: None; clinically determined diagnosis
- Management/Treatment
 1. Obtain detailed history of onset, duration, severity and progression of symptoms, and possible precipitating factors
 2. Describe in terms of morphology/structure, size, shape, number, color, location, distribution
 3. Wash and dry face and affected areas with mild nonoil base soap
 4. Use topical exfoliates and comedolytic preparations
 - a. Mild acne—topical benzoyl peroxide
 - b. Moderate acne—topical tretinoin, topical benzoyl peroxide
 - c. Severe acne—topical tretinoin, topical or oral antibiotics, oral tretinoin
 5. Use topical antibiotics for moderate to severe acne, e.g., clindamycin
 6. Add oral antibiotics for persistent and unresponsive cases of moderate and severe acne
 - a. Tetracycline, doxycycline, minocycline, tetracycline
 - b. Oral clindamycin contraindicated due to adverse GI side effects
 7. Consider using isotretinoin for unresponsive, persistent severe acne
 - a. Contraindicated in pregnancy; teratogenic
 - b. For sexually active females, birth control measures required
 - c. Federally mandated enrollment in iPLEDGE program requires informed signed consent, monthly laboratory studies, and monthly office visits
 8. Educate regarding medication dosage, signs of irritation, sensitivity
 9. Consider counseling for signs of psychological distress and depression
 10. Educate regarding characteristics of condition, treatment regime, and expected prognosis
 - a. Condition may become worse with treatment before improvement
 - b. Treatment will improve but not cure most cases; may take months
 - c. Treatment must be consistent to be effective
 11. Monitor progress every 4 to 6 weeks initially; less often as indicated when improvement is evident
 12. Refer for dermatologist evaluation if condition does not meet prognostic expectations

Folliculitis and Furuncles

- Definition: Infectious condition involving pilosebaceous follicle occurring on any skin surface where hair follicles are present but predominately on face, neck, scalp, and buttocks
 1. Folliculitis—superficial involvement of upper follicle
 2. Furuncle or boil—deeper involvement of follicle and dermal appendages
- Etiology/Incidence
 1. Caused most often by *Staphylococcus aureus*; less commonly by streptococcus bacteria
 2. Also seen with some tinea infections
 3. More common in males
- Signs and Symptoms
 1. Areas of tenderness, erythema, and swelling
 2. Nodules may be present with deep-seated furuncles
 3. Tenderness and warmth at site
- Differential Diagnosis
 1. Candida
 2. Impetigo

- Physical Findings
 1. Localized areas of erythema and edema with papular or pustular lesions on face, scalp, neck, buttocks, and other areas
 2. Nodules are present with deep-seated furuncles
 3. Tenderness and warmth may be present
 4. Regional adenopathy may be present
- Diagnostic Tests/Findings: Culture confirms specific bacterial agent
- Management/Treatment
 1. Obtain detailed history of onset, duration, severity and progression of symptoms, and possible precipitating factors
 2. Describe and monitor lesions in terms of morphology/structure, size, shape, number, color, location, distribution
 3. Wash with antimicrobial soap and apply warm moist compresses to affected areas
 4. Topical antibiotics, e.g., mupirocin
 5. Oral antibiotics
 - a. For staphylococci—use dicloxacillin
 - b. For streptococci infection—penicillin or cephalosporin; erythromycin for penicillin allergic patients
 - c. For MRSA, bactrim or clindamycin
 6. Educate regarding medication dosage, signs of irritation, sensitivity
 7. Educate regarding characteristics of condition, treatment regime, prognosis, and good hygiene measures
 8. Refer for dermatologist evaluation if condition does not follow prognostic expectation

■ VIRAL CONDITIONS

Herpes Simplex/Common Cold Sore

- Definition: Contagious infection, predominately of lips and oral mucosa, commonly known as fever blisters
 1. Initial infectious state—more severe, lasts longer, and is more painful
 2. Dormant state—virus lives on ending of selected nerves, asymptomatic
 3. Secondary infectious state—activated at times of increased stress, illness, fatigue, sun exposure, menses, dental procedures
- Etiology/Incidence
 1. Herpes simplex virus type 1—most common cause
 2. Herpes simplex virus type 2—considered in situations of oral sex
 3. Incubation varies, commonly 2 to 12 days

- Signs and Symptoms
 1. Erythema with grouped vesicles and crusting on lips
 2. Erythema and swelling with painful white ulcerated patches inside mouth
 3. Fever, generalized malaise, and sore throat may occur
 4. Mild itching, tingling, pain, and burning may precede blisters
- Differential Diagnosis
 1. Erythema multiforme
 2. Hand-foot-mouth disease
 3. Candidiasis
 4. Localized bacterial infection
 5. Sexual abuse
- Physical Findings
 1. Lip lesions—grouped or singular vesicles on an erythematous base erupt and form crusts; usually can be found on mucocutaneous border of lips
 2. Oral cavity lesions—erythema and edema of mucous membranes with singular or multiple vesicles and white ulcerations; may include tongue, palate, and gums
 3. Regional adenopathy may be present
 4. Halitosis may be present with oral lesions
 5. Lesions are present 10 to 14 days, gradually resolving
 6. Secondary infection may be present—most caused by staphylococcus bacteria
- Diagnostic Tests/Findings
 1. Tzanck smear confirms presence of multinuclear giant cells indicative of herpes
 2. Culture to confirm causal agent
- Management/Treatment
 1. Obtain detailed history of onset, duration, severity and progression of symptoms, and precipitating factors
 2. Describe and monitor lesions in terms of morphology/structure, size, shape, number, color, location, distribution
 3. Treat lip lesions
 - a. Burrow's compresses to alleviate discomfort
 - b. Topical antiviral applications, e.g., acyclovir, for recurrent disease
 4. Treat oral lesions
 - a. Avoid spicy and acid foods
 - b. Cool, bland fluids, especially when lesions are most painful
 - c. Anesthetic mouth rinses, e.g., lidocaine (with caution), or diphenhydramine

5. Oral antiviral medication with recurrent disease at first sign of prodrome (skin tingling), e.g., acyclovir, famciclovir, valacyclovir
6. Oral antibiotic to treat secondary bacterial infection, e.g., dicloxacillin, cefadroxil, cephalexin
7. Educate regarding medication dosage, signs of irritation, sensitivity
8. Educate regarding cause, characteristics of condition, communicability, and prognosis
9. Educate regarding preventive measures
 - a. Avoid direct exposure of others to lesions (kissing)
 - b. Wash hands before and after applying topical medications or touching lesions
 - c. Avoid sharing personal items—cosmetics, cups, eating utensils
10. Refer for dermatologist evaluation if condition does not improve

Molluscum Contagiosum

- Definition: Common infectious, self-limiting skin condition characterized by waxy, firm papules which may occur on any skin surface; predominantly on face, axillae, abdomen, and arms
- Etiology/Incidence
 1. Caused by a poxvirus
 2. Most common in children and adolescents
 3. Common in children with atopic dermatitis, HIV or AIDS
 4. Incubation is usually 2 to 8 weeks but may be up to 6 months
 5. Period of communicability uncertain
 - a. May persist as long as lesions are present
 - b. Spread by direct contact and through autoinoculations
- Signs and Symptoms
 1. Mild itching may be present
 2. Few or multiple small, firm, raised, pinkish-white or skin colored lesions
- Differential Diagnosis
 1. Warts
 2. Closed comedones
 3. Milia
 4. Juvenile xanthogranuloma
 5. Condylomata acuminata
- Physical Findings
 1. Papular pink-white or skin colored lesions of 1 to 5 mm in size, usually on face, neck, axillae, abdomen, and arms
 2. Occasionally lesions grow to 1 to 2 cm
- 3. Secondary bacterial infection may be present
- 4. Lesions may become umbilicated (central pitting, dimpled, or depressed)
- 5. Lesions are self-limiting but may be present for 2 to 3 years if left untreated
- 6. May occur in genital area in sexually active and sexually abused; more typically they are transmitted by a caregiver
- Diagnostic Tests/Findings
 1. Usually not necessary
 2. Wright or Giemsa stain of papule core will show characteristic intracytoplasmic inclusions
- Management/Treatment
 1. Obtain a detailed history of onset, duration, severity and progression of symptoms, and precipitating factors
 2. Describe and monitor lesions in terms of morphology/structure, size, shape, number, color, location, distribution
 3. Rule out child abuse if lesions in genital area
 4. Treatment options—with the exception of observation, none are cures
 - a. In otherwise healthy individuals, lesions typically resolve spontaneously without treatment over time
 - b. Curettage removal of lesions provides more expedient resolution as this is one method of stimulating an immune response; not recommended for facial lesions due to potential scarring
 - c. Topical application of keratolytics; not recommended for lesions near eyes
 - (1) Tretinoin cream
 - (2) Cartharidin
 - d. Topical antibiotics for secondary bacterial infection, e.g., mupirocin
 5. Education regarding medication dosages, signs of irritation, sensitivity
 6. Education regarding cause, characteristics of lesions, communicability, and prognosis
 7. Education regarding preventive measures
 - a. Avoid direct exposure of others to lesions
 - b. Wash hands before and after application of topical medications and/or touching lesions
 - c. Avoid sharing personal items—cosmetics, towels, cups, eating utensils, and siblings sharing baths if one of them has molluscum lesions
 8. Refer for dermatologist evaluation if condition does not resolve with selected treatment

Verruca vulgaris (Warts)

- Definition: Common self-limiting skin lesions characterized by firm, well-circumscribed, smooth to irregular, singular or multiple hyperkeratotic papules; predominately on fingers, palms, and soles of feet; commonly known as warts
- Etiology/Incidence
 1. Human papillomaviruses with more than 50 identified types
 2. Virus enters skin through minor trauma
 3. Occurs in 10% of children and adolescents with school-age children having the highest incidence
 4. Incidence may be increased with ongoing exposure to moisture
 5. Period of incubation varies widely, and estimated from 2 months to 2 years
 6. Period of communicability unknown
 - a. May persist as long as lesions are present
 - b. Spread by direct and indirect contact and through autoinoculations
- Signs and Symptoms
 1. Raised gray, brownish to skin colored, smooth to rough, singular or multiple lesions on hands
 2. Painful flat ingrown lesions on soles
 3. Bleeding may occur with trauma or picking
- Differential Diagnosis
 1. Molluscum contagiosum
 2. Calluses
- Physical Findings
 1. Common verruca—gray, brown, or skin colored; rough, singular or multiple papular lesions; most common on hands and fingers
 2. Flat verruca—skin colored, smooth, round, multiple lesions, slightly elevated; most common on the face and extremities
 3. Plantar verruca—skin colored, irregular, single or multiple lesions that appear flush with sole of foot and grow inward
 4. May occur in genital area of sexually active and sexually abused
 5. Lesions are self-limiting, usually 6 to 9 months but due to reinfection through autoinoculation, condition may persist for several years
- Diagnostic Tests/Findings: Excision and histological examination may confirm diagnosis
- Management/Treatment
 1. Obtain a detailed history of onset, duration, severity and progression of symptoms, and possible precipitating factors

2. Describe and monitor lesions in terms of morphology/structure, size, shape, number, color, location, distribution
3. Rule out child abuse if genital lesions are present
4. Consider treatment options knowing that with the exception of observation, none of the treatment options are cures; some of the treatment options are painful and some can scar
 - a. No treatment is usually necessary due to self-limiting condition
 - (1) Ideal treatment has not been established
 - (2) Frequently recur regardless of treatment choice until individual immune system creates immunity to the virus
 - b. Topical applications of keratolytics, e.g., OTC wart preparations, cantharidin, tretinoin
 - c. Applications of waterproof plastic tapes treated with keratolytics
 - d. Excision of lesions except those on face due to potential scarring
5. Topical antibiotics to treat secondary bacterial infection, e.g., mupirocin
6. Educate regarding medication dosages, signs of irritation, sensitivity
7. Educate regarding cause, characteristics of condition, communicability, and prognosis
8. Consider congenital or acquired immunodeficiency if no resolution and/or widespread
9. Refer for dermatologist evaluation if condition does not improve or bleeds with light trauma

■ FUNGAL INFECTIONS

Tinea Capitis (Ringworm of the Scalp)

- Definition: Superficial dermatophyte fungal skin infection of the scalp
- Etiology/Incidence
 1. Caused predominately by *Trichophyton tonsurans* (90%); also by *Microsporum canis*, *Microsporum audouinii*, and *Trichophyton mentagrophytes* (less common)
 2. Dermatophytes attach to epidermis skin layer of host's scalp and multiply within stratum corneum; do not involve lower layers of epidermis or dermis
 3. Spreads through direct and indirect contact with infected individuals, animals, caps, combs, brushes, glasses, and other personal articles
 4. *Microsporum canis* may be transmitted through contact with infected dogs or cats
 5. Occurs more often in hot humid climates

6. More common in darker-skinned individuals; boys more than girls
 7. Incubation period is unknown, possibly 10 to 14 days
 8. Communicability occurs as long as lesions with dermatophytes are present
- Signs and Symptoms
 1. Itching with varying degrees of severity
 2. Slightly raised round or angular scaly areas
 3. Sometimes yellow honeycomb crusts
 4. Broken hairs and alopecia may be present
 - Differential Diagnosis
 1. Impetigo
 2. Eczema
 3. Seborrhea dermatitis
 4. Psoriasis
 5. Trichotillomania
 6. Alopecia areata
 - Physical Findings
 1. Several presentations may occur singularly or at the same time
 - a. Scaly patches of varying sizes with or without alopecia and pruritis
 - b. Pustules, papules with areas of honeycomb crusts
 - c. Tender erythematous areas with broken hairs at scalp level leaving a “black-dot” appearance
 2. Regional adenopathy may be present, especially occipital nodes
 - Diagnostic Tests/Findings
 1. Wood’s lamp will fluoresce the *Microsporum canis* only and is of limited use in confirming tinea capitis
 2. KOH scraping from the areas of scalp with alopecia, “black dots,” or broken hairs will confirm hyphae and spores of dermatophytes
 - Management/Treatment
 1. Obtain detailed history of onset, duration, severity and progression of symptoms, and precipitating factors
 2. Describe and monitor lesions in terms of morphology/structure, size, shape, number, color, location, and distribution
 3. Treat with oral antifungal medication
 - a. Griseofulvin; ultramicrosize formulation has best absorption
 - b. Treat for 8 weeks
 - c. Topical antifungal medications are ineffective

4. Shampoo 2 to 3 times weekly with selenium sulfide or ketoconazole to reduce spore count and infectivity
5. Although condition is communicable, exclusion from school and other groups is not indicated unless treatment is refused or not followed
6. Educate regarding medication dosage, signs of irritation, sensitivity
7. Educate regarding characteristics of condition, treatment regime, and prognosis
8. Educate regarding communicability and prevention
 - a. Avoid sharing personal items of caps, combs, brushes, towels, pillows, glasses, razors; wash these items frequently
 - b. Wash hair immediately after barbershop or salon haircut
 - c. Maintain personal hygiene, wash hands before/after treatment
 - d. Avoid touching or scratching affected areas
9. Refer for dermatologist evaluation if condition does not improve

Tinea Corporis (Ringworm of the Body)

- Definition: Superficial dermatophyte fungal skin infection of less-hairy surfaces of body and face; commonly known as “ring worm” due to pattern of healing centrally while spreading peripherally
- Etiology/Incidence
 1. Primary source—*Trichophyton rubrum*, *Trichophyton mentagrophytes*, as well as *Microsporum canis*, and *Epidermophyton floccosum*
 2. Dermatophytes attach to epidermis skin layer of host and multiply within stratum corneum; do not involve lower layers of the epidermis or dermis
 3. Spreads through direct and indirect contact with infected individuals, animals, shower stalls, benches, and other articles
 4. *Microsporum canis* may be transmitted through contact with infected dogs or cats
 5. Occurs more often in hot humid climates
 6. Incubation period is unknown, possibly 4 to 14 days
 7. Communicability occurs as long as lesions with dermatophytes are present
- Signs and Symptoms
 1. Mild itching at site of affected areas
 2. Slightly raised, round or angular scaly areas with pink borders

- Differential Diagnosis
 1. Contact dermatitis
 2. Eczema
 3. Psoriasis
 4. Pityriasis rosea
 5. Pityriasis versicolor
 6. Granuloma annulare
- Physical Findings
 1. Typical lesions are scaly plaques of varying sizes from less than 5 mm to more than 3 cm with mild erythematous active borders
 2. Lesions spread peripherally as they heal centrally
 3. Lesions may be singular or several; numerous lesions are uncommon
- Diagnostic Tests/Findings
 1. Wood's lamp will fluoresce the *Microsporum canis*
 2. KOH scraping of lesion border—confirms hyphae and spores
 3. Dermatophyte test medium (DTM)—confirm diagnosis
- Management/Treatment
 1. Obtain a detailed history of onset, duration, severity and progression of symptoms, and precipitating factors
 2. Describe and monitor lesions in terms of morphology/structure, size, shape, number, color, location, and distribution
 3. Treat with topical antifungal medications
 - a. Clotrimazole, miconazole, econazole, terbinafine, tolnaftate, naftifine, ciclopirox, ketoconazole
 - b. May require treatment up to 8 weeks before resolution
 4. Treat with oral antifungal medication for extensive, recurrent, and unresponsive conditions, e.g., griseofulvin
 5. Educate regarding medication dosage, signs of irritation, sensitivity
 6. Educate regarding characteristics of condition, treatment regime, and prognosis
 7. Educate regarding communicability and prevention
 - a. Avoid sharing personal items of clothing, towels, pillows, razors, and wash these items frequently
 - b. Maintain personal hygiene and wash hands before and after applying treatment
 - c. Avoid touching or scratching affected areas
 - d. Avoid or shower after using public pools
 - e. Wash clothing touching affected areas after each use

8. Refer for dermatologist evaluation if condition does not improve

Tinea Cruris (Jock Itch)

- Definition: Superficial dermatophyte fungal skin infection of the groin, upper thighs, and/or inguinal folds; commonly called “jock itch”
- Etiology/Incidence
 1. Caused by *Epidermophyton floccosum*, *Trichophyton rubrum*, and *Trichophyton mentagrophytes*
 2. Dermatophytes attach to epidermis skin layer of host and multiply within stratum corneum; lower layers of epidermis or dermis are not involved
 3. Occurs more often during hot humid weather with increased sweating
 4. More common in adolescents, athletes, obese children, and males
 5. Spreads through direct and indirect contact with infected individuals, including sexual contact
 6. Incubation period is unknown, possibly 4 to 14 days
 7. Communicability occurs as long as lesions with dermatophytes are present
- Signs and Symptoms
 1. Pain and tenderness with varying degrees of severity
 2. Itching with varying degrees of severity reported, especially during healing
 3. Erythematous, hyperpigmented, slightly raised scaly patches with defined borders
 4. Blisters may also be present
- Differential Diagnosis
 1. Contact dermatitis
 2. Eczema
 3. Intertrigo
 4. Psoriasis
 5. Seborrheic dermatitis
 6. Acanthosis nigricans
- Physical Findings
 1. Erythematous, scaly red to brown lesions of varying sizes with well-defined raised borders
 - a. Small vesicles, central clearing, and peripheral spreading may or may not be present
 - b. Affected areas may be singular or multiple
 - c. In chronic cases, lichenification may be present

2. All areas of the groin may be affected including scrotum, gluteal folds, buttocks, inner aspect of thighs
 3. Painful to touch and with movement
 4. Often concurrent with tinea pedis
- Diagnostic Tests/Findings
 1. KOH scraping of lesion border—confirms hyphae and spores
 2. DTM—confirms diagnosis
 - Management/Treatment
 1. Obtain a detailed history of onset, duration, severity and progression of symptoms, and precipitating factors
 2. Describe and monitor lesions in terms of morphology/structure, size, shape, number, color, location, and distribution
 3. Treat with topical antifungal medications
 - a. Clotrimazole, haloprogin, miconazole, terbinafine, tolnaftate, ciclopirox, econazole, ketoconazole, naftifine, oxiconazole, sulconazole
 - b. May require treatment up to 4 to 6 weeks before resolution
 4. Treat with oral antifungal medication for extensive, recurrent, and/or unresponsive conditions, e.g., griseofulvin
 5. Educate regarding medication dosage, signs of irritation, sensitivity
 6. Educate regarding characteristics of condition, treatment regime, and prognosis
 7. Educate regarding communicability and prevention
 - a. Avoid sharing undergarments—pants, jock straps
 - b. Wash personal undergarments frequently
 - c. Maintain good daily personal hygiene and dry well after bathing
 - d. Wash hands before and after applying topical treatment
 - e. Avoid touching or scratching affected areas
 - f. Avoid or shower after using public pools
 - g. Launder clothing touching affected areas after each use
 - h. Don't wear tight clothes next to affected area, including jeans and undergarments
 - i. Use cotton undergarments and change daily
 8. Refer for dermatologist evaluation if condition does not improve

Tinea Pedis (Athlete's Feet or Ringworm of the Feet)

- Definition: Superficial dermatophyte fungal skin infection of toes and feet
- Etiology/Incidence
 1. Caused by *Trichophyton rubrum*, *Trichophyton mentagrophytes*, and *Epidermophyton floccosum* fungal dermatophytes
 2. Dermatophytes attach to epidermis skin layer of host and multiply within stratum corneum; do not involve lower layers of epidermis or dermis
 3. Occurs more often during hot, humid weather with increased sweating
 4. Occurs worldwide; more common in adolescents, athletes, and males
 5. Spreads through direct and indirect contact with infected individuals, public baths, swimming pools, and locker rooms
 6. Incubation period is unknown
 7. Communicability occurs as long as lesions with dermatophytes are present
- Signs and Symptoms
 1. Pruritus of affected areas
 2. Erythematous, scaly, and occasionally blistered areas anywhere on foot; cracks and scaling between toes
 3. Stinging or pain if cracks between toes
- Differential Diagnosis
 1. Atopic dermatitis
 2. Contact dermatitis
 3. Candidiasis
 4. Eczema
- Physical Findings
 1. Erythematous scaly patches of varying sizes
 - a. Small vesicles, central clearing, and peripheral spreading may or may not be present
 - b. Affected areas may be anywhere on foot, most commonly on lateral and plantar portions
 2. Lesions on or between toes are scaly with mild erythema
 - a. Interdigital fissures are present
 - b. One or multiple toes may be involved, most commonly between third and fourth toes
 3. Dystrophy of toenails may be present with yellow discoloration of the nail matrix and periungual debris

- Diagnostic Tests/Findings
 1. KOH scraping of lesion border confirms hyphae and spores
 2. DTM of skin scraping or nail clippings confirms diagnosis
- Management/Treatment
 1. Obtain a detailed history of onset, duration, severity and progression of symptoms, and precipitating factors
 2. Describe and monitor lesions in terms of morphology/structure, size, shape, number, color, location, and distribution
 3. Treat with topical antifungal medications
 - a. Clotrimazole, haloprogin, miconazole, econazole, ciclopirox, terbinafine, tolnaftate, ketoconazole, naftifine, oxiconazole, sulconazole
 - b. May require treatment 8 to 12 weeks before resolution
 4. Treat vesicular and fissured lesions with compresses of Burow's solution
 5. Use absorbent antifungal powder
 6. Treat with oral antifungal medication for extensive, recurrent, and unresponsive conditions, e.g., griseofulvin
 7. Educate regarding medication dosage and signs of irritation and sensitivity
 8. Educate regarding characteristics of condition, treatment regime, and prognosis
 9. Educate regarding communicability and prevention
 - a. Avoid sharing personal items of shoes, socks, and towels
 - b. Wash personal items frequently
 - c. Maintain good daily personal hygiene and dry well after bathing
 - d. Wash hands before and after applying topical treatment
 - e. Avoid touching or scratching affected areas
 - f. Avoid using public pools or shower after each use
 - g. Launder clothing touching affected areas after each use
 - h. Don't wear tight and closed shoes
 - i. Use cotton socks instead of nylon or polyester
 10. Refer for dermatologist evaluation if condition does not improve

■ INSECT CONDITIONS

Common Insect Bites

- Definition: Wound inflicted by bite of a blood-sucking arthropod
- Etiology/Incidence
 1. Caused when mosquitos, fleas, chiggers, and bedbugs feed on human blood
 - a. Are attracted to host's moisture, odor, and warmth
 - b. Serve as vectors for diseases such as malaria
 2. Pet dogs and cats act as hosts for some fleas that are also attracted to humans
 3. More bites occur:
 - a. In warm and humid weather
 - b. Around stagnant water
 - c. In outside grassy and sandy areas
 - d. On uncovered body areas
 4. Itching caused by sensitivity to insect's saliva
- Signs and Symptoms
 1. Itching is major symptom—may persist 5 to 7 days after exposure
 2. Pain—variable
 3. Single or multiple pink/red raised lesions on legs, abdomen, and exposed areas of upper body
- Differential Diagnosis
 1. Folliculitis
 2. Insect sting
 3. Spider bite
 4. Scabies
- Physical Findings
 1. Single or multiple erythematous papules and wheals on lower extremities, abdomen, and exposed upper body parts
 2. Lesions from bed bug and chigger bites are smaller, more erythematous, and more numerous
 3. Vesicles may develop, signaling greater sensitivity
 4. Excoriation may be present with intense pruritus
 5. Pustules may develop, indicating secondary bacterial infection
- Diagnostic Tests/Findings: Culture of pustules confirms causal organism of secondary infection
- Management/Treatment
 1. Obtain detailed history of bite, progression of symptoms, and precipitating factors
 2. Describe and monitor lesions in terms of morphology/structure, size, shape, number, color, location, and distribution
 3. Provide symptomatic treatment of pruritus
 - a. Cool compresses
 - b. Topical histamines, e.g., hydroxyzine, diphenhydramine

- c. Oral antihistamines if topical treatment is ineffective, e.g., hydroxyzine, diphenhydramine
- 4. Topical steroids to reduce inflammation and immune response, e.g., hydrocortisone, triamcinolone
- 5. Treat secondary bacterial infection
 - a. Use topical antibiotics, e.g., mupirocin
 - b. Treat with oral antibiotics if extensive, recurrent, or unresponsive, e.g., cefadroxil, cephalexin
- 6. Educate regarding characteristics of condition, treatment regime, and prognosis
- 7. Educate regarding medication dosage, signs of irritation, sensitivity
- 8. Educate regarding prevention
 - a. Outside environmental controls—clearing areas, pesticide spraying, removing stagnant water
 - b. Inside environmental controls—routine cleaning, vacuuming
 - c. Bathe flea-infested pets
 - d. Wear cover-up clothing
 - e. Wear insect repellants
 - f. Avoid wearing fragrances that may attract
 - g. Avoid scratching to prevent infection
- 9. Refer for dermatologist evaluation if condition does not resolve

Spider Bites

- Definition: Wound inflicted by spider, characterized by both local and systemic manifestations
- Etiology/Incidence
 1. Most spider bites are harmless, causing small localized reaction at site of bite
 2. In U.S., bites from two nonaggressive venomous spiders produce severe toxic reactions in some individuals
 - a. Black widow
 - (1) Mature female is shiny, black, gray, or brown with an orange hourglass marking on the ventral surface
 - (2) Overall size is 2.5 to 4.5 cm including legs
 - (3) Male is smaller with fangs that cannot penetrate human skin
 - (4) Most common in Ohio, South, Southwest, and West coast
 - (5) Likes dry, warm, dark areas; found in grass, wood piles, gardens, sheds, basements, closets, and trunks
 - (6) Spin irregular asymmetrical web to catch flies and other prey

- b. Brown recluse
 - (1) Mature spider is gray, or varying shades of red to pale brown, with a violin-shaped marking on cephalothorax
 - (2) Overall size is 1.5 to 2.5 cm, including legs
 - (3) Most common in the Midwest and South
 - (4) Likes trunks, carpets, old shoes, old clothes, closets, crates, shelves
- 3. Most bites are in self-defense when spider feels threatened
- 4. Most bites occur in warmer months
- 5. Infants and small children are most vulnerable to developing serious reactions

- Signs and Symptoms

1. Black widow
 - a. Initial sensation of pinch or sting is often unnoticed
 - b. Later within 1 hour of the bite
 - (1) Dull burning or pain at site of bite
 - (2) Two red puncture marks surrounded by white area with bluish-red border
 - (3) Muscle cramps and sweating
 - (4) Muscle spasms can spread to rest of body
 - (5) In severe cases can progress to shock, coma, and death
2. Brown recluse
 - a. Initial sensation of bite is most often unnoticed or moderately painful
 - b. Later within 2 to 7 hours:
 - (1) Mild localized tingling
 - (2) Redness or blanching
 - c. After 48 to 72 hours:
 - (1) Blister surrounded by blue-gray area
 - (2) Flu-like symptoms may be experienced
3. Reactions from bites of both spiders may become more intense and last for days with more serious life-threatening signs/symptoms developing in a few cases

- Differential Diagnosis—Black widow and Brown recluse

1. Other insect bites (both)
2. Tetanus (black widow)
3. Appendicitis (black widow)
4. Diabetic ulcers (brown recluse)
5. Stevens-Johnson syndrome (brown recluse)

- Physical Findings—specifics vary by type of spider

1. Black widow spider bite
 - a. Symptoms begin within one hour
 - b. Dull burning or pain at site

- c. Two red puncture marks surrounded by a blanched area with bluish erythematous border
 - d. Muscle spasms, hypertension, tachycardia, diaphoresis
2. Brown recluse spider bite
 - a. Initial symptoms begin within 2 to 7 hours
 - (1) Mild, localized tingling
 - (2) Erythema or blanching at site
 - b. Delayed symptoms after 48 to 72 hours
 - (1) Hemorrhagic vesicle surrounded by bluish, gray areas of developing necrosis
 - (2) Flu-like symptoms
 3. Both (black widow and brown recluse)
 - a. Reactions may last for days to weeks
 - b. Potential to become serious and life-threatening with major renal, respiratory, cardiovascular, and neurological system involvement
- Diagnostic Tests/Findings
 1. No tests confirm specific diagnosis
 2. Dead spider specimen may help to confirm specific species
 - Management/Treatment
 1. Obtain detailed history of onset, duration, severity, progression of symptoms, and precipitating factors
 2. Describe and monitor symptoms and lesions in terms of morphology/structure, size, shape, number, color, location, and distribution
 3. If bite from black widow or brown recluse spider is suspected:
 - a. Apply cold compresses to site of bite
 - b. Refer immediately for dermatologist evaluation and hospitalization due to potential risk of severe reaction
 4. If bite from another less harmful spider is suspected:
 - a. Apply cool compresses to site of bite
 - b. Use oral antihistamines to reduce severe pruritus, e.g., hydroxyzine, diphenhydramine
 - c. Monitor for hypersensitivity reaction
 5. Educate regarding characteristics of condition, treatment regime, and prognosis
 6. Educate regarding medication dosage, signs of irritation, sensitivity
 7. Educate regarding prevention
 - a. Outside environmental controls—clearing areas, pesticide spraying
 - b. Inside environmental controls—routine cleaning, vacuuming
 - c. Avoid and/or be observant around areas of natural habitat

- d. Wear protective clothing and hats
- e. Wear gloves when cleaning closets and trunks
- f. Inspect clothing and shoes prior to wearing

Insect Stings

- Definition: Wound inflicted by sting of an insect, characterized by systemic and/or local manifestations
- Etiology/Incidence
 1. Caused by bees, hornets, wasps, yellow jackets, and fire ants
 2. Hypersensitivity to venom develops after initial exposure with more severe reactions upon subsequent exposures
 - a. Mild reactions occur in 90% of children
 - b. Anaphylaxis occurs in approximately 7% of general population
 3. Most stings occur in self-defense when insect feels threatened
 4. Most stings occur in warmer months
 5. Multiple stings may occur when around nests or swarms of insects
- Signs and Symptoms
 1. Usual reaction after initial exposure lasts up to 24 hours
 - a. Pain with varying degrees of severity
 - b. Redness and swelling at site of sting
 2. More pronounced reaction after re-exposure
 - a. Nausea and abdominal pain
 - b. Sneezing and coughing
 - c. Itching
 - d. Larger area of redness and swelling
 3. Anaphylactic reaction may occur after initial or re-exposure
 - a. Early signs within minutes of exposure
 - (1) Dizziness
 - (2) Swelling of lips and throat
 - (3) Difficulty breathing
 - (4) Difficulty swallowing
 - b. Later signs
 - (1) Weakness and collapse
 - (2) Confusion
 - (3) Coma
- Differential Diagnosis
 1. Spider bites
 2. Other insect bites
- Physical Findings
 1. Usual reaction after initial exposure may last up to 24 hours
 - a. Pain with varying degrees of severity
 - b. Erythema and edema surrounding central punctum at site of sting

2. Thin white to gray stinger may project from center
 3. More pronounced reaction may last several days especially after re-exposure
 - a. Nausea, abdominal pain
 - b. Sneezing, coughing
 - c. Pruritus
 - d. Larger area of redness, swelling
 4. Anaphylactic reaction may occur after initial or re-exposure
 - a. Could result in ultimate collapse and death
 - b. Early signs within minutes of exposure
 - (1) Dizziness
 - (2) Swelling of lips and throat
 - (3) Difficulty breathing
 - (4) Difficulty swallowing
 - c. Later signs
 - (1) Weakness and collapse
 - (2) Confusion
 - (3) Coma
 - (4) Stridor
- Diagnostic Tests/Findings: None
 - Management/Treatment
 1. Obtain detailed history of onset, duration, severity and progression of symptoms
 2. Describe and monitor area in terms of size, color, and location
 3. If known sensitivity, life-threatening and/or severe reaction:
 - a. Administer epinephrine as indicated
 - b. Apply cool compresses
 - c. Refer immediately for dermatologist evaluation and hospitalization
 4. If mild reaction:
 - a. Remove stinger by flicking off, do not squeeze
 - b. Apply cool compresses to site of sting
 - c. Use oral antihistamines to reduce severe pruritus, e.g., hydroxyzine, diphenhydramine
 - d. Monitor for hypersensitivity reaction
 5. Provide emotional support as needed to child and family
 6. Educate regarding characteristics of condition, treatment regime, and prognosis
 7. Educate regarding medication dosage, signs of irritation, sensitivity
 8. Educate regarding prevention
 - a. Outside environmental controls—clearing areas, pesticide spraying
 - b. Inside environmental controls—routine cleaning, vacuuming
 - c. Avoid and/or be observant around areas of natural habitat

- d. Wear protective clothing, hats and gloves
- e. Avoid wearing bright clothing when hiking around natural habitat
- f. Avoid wearing perfumes when around natural habitat
- g. If known sensitivity, wear medical alert tag and carry epinephrine kit

■ INSECT INFESTATIONS

Scabies Infestation

- Definition: Highly contagious condition caused by parasitic mite infestation
- Etiology/Incidence
 1. Caused by the *Sarcoptes scabiei* (itch mite); gravid female mite burrows into stratum corneum to lay ova, which hatch in 4 to 14 days
 2. Incubation period of 4 to 6 weeks with initial exposure; 1 to 5 days with re-exposure causing intense itching
 3. Worldwide distribution in all population groups regardless of hygiene
 4. Major infestations have occurred in cyclic patterns of every 15 to 30 years
 5. Spreads through direct contact with infected person or indirect contact with clothing, bed linens, and other personal items
 6. Communicability is present until all mites, larva, and ova are destroyed on body surface and in surrounding environment
- Signs and Symptoms
 1. Irritability in infants
 2. Intense pruritis, especially at night in older children and adolescents
 3. Red bumps, blisters, pustules, and small burrow marks which may be obliterated by scratch marks
- Differential Diagnosis
 1. Insect bites
 2. Impetigo
 3. Secondary bacterial infection
- Physical Findings
 1. Intense itching
 2. Fine gray- to skin-colored superficial 2 to 8 mm linear curved burrows with small papule at proximal end; burrows may be obliterated by scratch and excoriation marks due to scratching
 3. Infants—typically have red-brown papular, vesicular lesions on head, neck, palms, and soles

4. Older child and adolescent—typically have red papular lesions on webs of fingers and folds of wrists, elbows, axillae, waist, buttocks, groin, umbilicus, abdomen, knees, ankles
 5. Pustules indicate secondary bacterial infection
 6. Regional adenopathy may be present
- Diagnostic Tests/Findings
 1. Skin scrapings of burrow or papule material and microscopic examination for body parts of mite, ova, or feces
 2. Culture of pustule will confirm agent of secondary infection
 - Management/Treatment
 1. Obtain a detailed history of onset, duration, severity, and progression of symptoms
 2. Describe and monitor lesions in terms of morphology/structure, size shape, number, color, location, and distribution
 3. Bathe and dry skin, then treat with topical medication
 - a. Infants and young children—permethrin 5% (drug of choice)
 - b. Older children and adolescents—permethrin 5%; lindane, crotamiton 10%; sulfur in petrolatum
 - c. Since 2004, Lindane has had an FDA-mandated boxed warning about not using this product on children weighing < 110 pounds
 4. Use topical steroids to reduce inflammation, immune response, and pruritus, e.g., hydrocortisone, triamcinolone
 5. Use oral antihistamines to reduce pruritus, e.g., hydroxyzine, diphenhydramine
 6. Treat secondary bacterial infection
 - a. Use topical antibiotics, e.g., mupirocin
 - b. Use oral antibiotics if extensive, recurrent, or unresponsive, e.g., dicloxacillin, cefadroxil, cephalexin
 7. Educate regarding medication dosage, signs of irritation, sensitivity
 8. Treat household and other close contacts
 9. Wash clothes, bed linens, towels, and hats with hot water and dry in hot dryer
 10. Store nonwashable items in plastic bags for one week; do not use
 11. Educate regarding characteristics of condition, treatment regime, and prognosis. Residual pruritus and skin irritation can persist for weeks after successful treatment
 12. Educate regarding medication dosage and signs of irritation and sensitivity

13. Educate regarding communicability and prevention
 - a. Avoid sharing personal items of clothes, linens, towels, and wash these items frequently
 - b. Maintain good daily personal hygiene and dry well after bathing
 - c. Wash hands before and after applying topical treatment
 - d. Avoid touching or scratching affected areas
14. Refer for dermatologist evaluation if condition does not improve

Pediculosis Infestation (Lice)

- Definition: Highly contagious parasitic louse infestation affecting hairy body surfaces
- Etiology/Incidence
 1. Caused by several species of lice
 - a. *Pediculus capitis*—affects scalp
 - b. *Pediculus humanus*—affects less hairy body surfaces
 - c. *Phthirus pubis*—affects pubic and axilla areas, eyelashes, eyebrows
 2. Worldwide distribution in all population groups regardless of hygiene practices
 3. More common in school age and adolescents due to sharing of personal items
 4. More common in Caucasians, less common in African Americans
 5. Spreads through direct contact with infected person or indirect contact with clothing, bed linens, and other personal items
 6. Lice do not fly or jump
 7. Incubation of 6 to 10 days from laying of eggs to hatching; hatched lice mature in 2 to 3 weeks
 8. Communicability present until all lice, neophytes, and ova are destroyed on body surface and environment
- Signs and Symptoms
 1. Pruritus; however, this may not be present until 4 to 6 weeks after initial infestation
 2. Tenacious white “flakes” on hair (nits); however, these egg casings may be empty (already hatched)
 3. Erythematous blotches and bumps (rare)
- Differential Diagnosis
 1. Bites from other insects
 2. Bacterial infection
 3. Dandruff
 4. Impetigo
 5. Scabies

- Physical Findings
 1. The only definitive way to diagnose an active infestation is to identify a live louse
 2. Small white nits (eggs) on hair strands— $\frac{1}{4}$ inch from skin surface; difficult to remove; the nits must be close to the scalp for a blood meal
 - a. Head lice most common on back of head, behind ears
 - b. Body lice most common in seams of clothing
 3. Macular, papular lesions with mild erythema and excoriation
 4. Pustules secondary to scratching (secondary bacterial infection)
 5. Regional adenopathy may be present
- Diagnostic Tests/Findings
 1. Clinical examination of hair shaft for ova is usually sufficient to confirm diagnosis
 2. Microscopic examination of ova may confirm questionable diagnosis
- Management/Treatment
 1. Obtain detailed history of onset, duration, severity and progression of symptoms
 2. Describe and monitor lesions in terms of morphology/structure, size, shape, number, color, location, and distribution
 3. Treat infestation with topical antiparasitics to destroy louse and ova, e.g., permethrin, pyrethrins, malathion, ivermectin
 4. Resistance to antiparasitics is increasing; permethrin only has 70% efficacy
 5. Since 2004, Lindane has had an FDA-mandated boxed warning about not using this product on children weighing < 110 pounds
 6. Two treatments of medication of choice recommended
 7. Educate regarding medication dosage, signs of irritation, sensitivity
 8. Remove ova/nits after topical treatment
 - a. Head lice—manually with fine tooth comb
 - b. Vinegar and water preparation may help soften cement
 - c. Eyelashes—may coat with petroleum jelly for several days
 9. Use topical antibiotics to treat secondary bacterial infection, e.g., mupirocin
 10. Only treat other infested family members; you do not need to automatically treat every household contact
 11. Remove infestation from surrounding environment
 - a. Wash clothes, bed linens, towels, and hats with hot water and dry in hot dryer

- b. Wash personal items such as combs and brushes with pediculocide
 - c. Lice can only live 24 hours away from a human host, so you do not need to vacuum drapes, rugs, floors, and furniture
12. Educate regarding characteristics of condition and prognosis. Residual pruritis and skin irritation can persist for weeks after successful treatment
13. Educate regarding communicability and prevention
 - a. Avoid sharing personal items of towels, hats, hair brushes, combs; wash these items frequently
 - b. Maintain personal hygiene, wash hands before/after treatments
 - c. Avoid touching or scratching affected areas
 - d. Avoid promoting a “no nit” return to school policy
14. Refer for dermatologist evaluation if condition does not resolve

■ MISCELLANEOUS CONDITIONS OF HYPERSENSITIVITY

Drug Eruptions

- Definition: Acute condition of the skin involving an allergic hypersensitivity reaction to a drug characterized predominately by a morbilliform generalized rash
- Etiology/Incidence
 1. Caused by release of histamine in reaction to immune system's response to drug allergen
 2. Most common drugs
 - a. Sulfates
 - b. Penicillins
 - c. Barbiturates
 - d. Dilantin
 3. Onset usually occurs within first week of exposure; may be delayed for more than 2 weeks and/or after drug has been discontinued
 4. Recurrences are frequent with re-exposures—response varies depending on antigen exposure
- Signs and Symptoms
 1. Intense generalized and localized pruritis
 2. Generalized erythematous lesions beginning on trunk and progressing to extremities
- Differential Diagnosis
 1. Syphilis
 2. Contact dermatitis
 3. Erythema multiforme

4. Rubeola
 5. Urticaria
 6. Gianotti-Crosti
 7. Erythema nodosum
- Physical Findings
 1. Generalized and localized pruritus
 2. Generalized morbilliform erythematous rash occurring first on trunk and progressing to extremities; initially macular, becoming papular and confluent
 3. Wheals are less typical and less frequent
 - Diagnostic Tests/Findings: None
 - Management/Treatment
 1. Obtain detailed history of onset, duration, severity and progression of symptoms, and possible precipitating factors
 2. Describe and monitor lesions in terms of morphology/structure, size, shape, number, color, location, distribution
 3. Discontinue contact with drug/medication allergen if known sensitivity
 4. Oral steroids to reduce inflammation and immune response in severe and extensive cases, e.g., prednisone
 5. Oral antihistamines for nocturnal pruritus, e.g., hydroxyzine, diphenhydramine; non-sedating oral antihistamines for daytime pruritus, e.g., OTC Loratadine, Citirizine
 6. Educate regarding medication dosage, signs of irritation, sensitivity
 7. Educate regarding characteristics of condition, cause, treatment prognosis, and recurrence
 8. Refer to dermatologist for evaluation if condition does not improve in 2 days or if becomes more severe at any time

Erythema Multiforme Minor

- Definition: Acute condition of the skin involving hypersensitivity reaction characterized by multimorphology skin and mucous membrane eruptions; lasts approximately 2 to 3 weeks with spontaneous resolution
- Etiology/Incidence
 1. Hypersensitivity caused by exposure to variety of substances
 - a. Infectious organisms—most common are enteroviruses, *Mycoplasma pneumoniae*, and herpes simplex, especially in recurrent conditions
 - b. Drugs—most common are barbiturates, sulfa, and penicillin drugs
 - c. Other substances—food reactions

2. More common in adults, however, approximately 20% of cases are in children and adolescents
 3. Recurrent episodes occur in approximately $\frac{1}{3}$ of cases
- Signs and Symptoms
 1. Itching may be present
 2. Pain, especially in mouth
 3. Redness and swelling may present with blisters and/or ulcers on hands, elbows, knees, ankles, feet, eyes, lips, mouth
 4. Develop in crops over period of 1 to 2 weeks with each crop lasting 1 week
 - Differential Diagnosis
 1. Allergic vasculitis
 2. Kawasaki disease
 3. Urticaria
 4. Varicella or other viral infections
 - Physical Findings
 1. Pruritus and pain may be present at site of lesions, especially those in oral cavity
 2. Erythema and edema with lesions progressing from macules to papules, vesicles, bulla, and petechiae
 3. Lesions occur on bilateral exposed areas predominately—includes hands, elbows, knees, ankles, feet, eyes, lips, oral mucous membranes, tongue, oral cavity, and less commonly on chest and trunk
 4. Lesions develop in crops over period of 1 to 2 weeks with each crop lasting 1 week
 5. Targetoid or “bull’s-eye” lesions may be present which have three distinct characteristics—a necrotic or vesicular center, a pale middle macular ring and an outer erythematous peripheral ring
 6. Lasts from 2 to 3 weeks with spontaneous resolution
 - Diagnostic Tests/Findings
 1. Chest radiograph to rule out *Mycoplasma pneumoniae*
 2. Tzanck test to rule out herpes simplex
 - Management/Treatment
 1. Obtain detailed history of onset, duration, severity and progression of symptoms, and possible precipitating factors
 2. Describe and monitor lesions in terms of morphology/structure, size, shape, number, color, location, distribution
 3. Cool compresses for pain and pruritus
 4. Antihistamines for nocturnal pruritus, e.g., hydroxyzine, diphenhydramine; non-sedating

- antihistamines for daytime pruritis, e.g., OTC Loratadine and Citirizine
- 5. Oral analgesics for generalized pain, e.g., acetaminophen
- 6. Topical anesthetics and mouthwashes for oral lesions, e.g., lidocaine
- 7. Maintain hydration with cool fluids, e.g., water, nonacidic juices
- 8. Determine underlying trigger and remove or treat as indicated
- 9. Educate regarding medication dosages, signs of irritation, sensitivity
- 10. Educate regarding characteristics of condition, cause, prognosis, and recurrence
- 11. Refer for dermatologist evaluation if:
 - a. Condition does not improve
 - b. Systemic symptoms of fever and malaise develop

Erythema Multiforme Major (Stevens-Johnson Syndrome)

- Definition: Skin condition involving hypersensitivity reaction characterized by multimorphology mucous membrane and skin eruptions with associated systemic involvement; also known as Stevens-Johnson Syndrome
- Etiology/Incidence
 1. Hypersensitivity caused by exposure to a variety of substances
 - a. Infectious organisms—most common are enteroviruses, *Mycoplasma pneumoniae*, and herpes simplex, especially in recurrent conditions
 - b. Drugs—most common are barbiturates, sulfa, and penicillin
 - c. Other substances—food reactions
 2. More common in adults, however, approximately 20% of cases are in children and adolescents
 3. Recurrent episodes occur in approximately $\frac{1}{3}$ of the cases
 4. Can be life threatening; approximately 5% mortality of diagnosed cases
- Signs and Symptoms
 1. Fever, fatigue, sore throat, headache, nausea, vomiting, diarrhea, muscle pain, and/or joint pain
 2. Skin rash develops in 2 to 3 days after generalized symptoms
 - a. Areas of erythema and edema
 - b. Variety of skin reactions on hands, elbows, knees, ankles, feet, eyes, lips, mouth, chest, and/or trunk
- 3. Pruritis may be present
- 4. Pain, especially in mouth
- Differential Diagnosis
 1. Gingivostomatitis
 2. Pemphigus
 3. Toxic epidermal necrolysis
 4. Urticaria
 5. Varicella
 6. Staphylococcal scalded syndrome
- Physical Findings
 1. Sudden onset of prodromal state—high temperature, malaise, weakness
 2. Multimorphology rash develops in progressive pattern
 - a. Macular erythematous with edematous areas
 - b. Progress to papules, vesicles, erosions, and petechiae
 3. Pruritus and pain, especially lesions in oral cavity
 4. Lesions occur on bilateral exposed areas predominately—includes hands, elbows, knees, ankles, feet, eyes, lips, oral mucous membranes, tongue; less common on chest and trunk
 5. Targetoid or herald lesions may be present which have three distinct characteristics—necrotic or vesicular center, pale middle macular ring, and an outer erythematous peripheral ring
 6. Lesions develop in crops over period of 1 to 2 weeks with each crop lasting 1 week
 7. Condition may progress to more severe stage involving the respiratory, renal, and gastrointestinal systems
- Diagnostic Tests/Findings
 1. Chest radiograph to rule out *Mycoplasma pneumoniae*
 2. Tzanck to rule out herpes simplex
 3. Skin biopsy to confirm diagnosis
- Management/Treatment
 1. Obtain detailed history of onset, duration, severity and progression of symptoms, and possible precipitating factors
 2. Describe and monitor lesions in terms of morphology/structure, size, shape, number, color, location, distribution
 3. Immediate dermatologist referral for evaluation and hospitalization due to potential life-threatening situation
 4. Educate regarding characteristics of condition, cause, and prognosis

Urticaria (Hives)

- Definition: Acute or chronic condition of the skin involving an allergic hypersensitivity reaction characterized by transient pale or skin-colored skin lesions
- Etiology/Incidence
 1. Caused by release of histamine as reaction to immune system's response to an allergen
 - a. Foods
 - b. Temperature changes of heat and cold
 - c. Viral infections
 - d. Vibrations and scratching
 - e. Emotional elation or stress
 - f. Insect bites
 - g. Materials and fabrics
 2. Symptoms may last for minutes or for up to 24 hours after initial exposure to antigen
 3. Recurrences are frequent with re-exposure to allergen; response is varied and may become chronic
- Signs and Symptoms
 1. Intense generalized and localized pruritis
 2. Mild erythema and edema of irregular shaped wheals—may involve eyelids, lips, hands, feet, mouth, and genitalia
- Differential Diagnosis
 1. Atopic dermatitis
 2. Contact dermatitis
 3. Erythema multiforme
- Physical Findings
 1. Intense generalized and localized pruritus at site of lesions
 2. Mild erythema and swelling with irregular shaped wheals on any skin surface
 - a. May have edema of eyelids, lips, hands, feet, and genitalia
 - b. May have edema and erythema of mucous membranes
 3. Individual lesions range in size from under 1 cm to over 15 cm
 4. Distribution pattern is generalized and scattered
 5. Become more pronounced with heat
 6. Will blanch with pressure
 7. Excoriation due to scratching secondary to severe pruritus
- Diagnostic Tests/Findings: No tests confirm condition
- Management/Treatment
 1. Obtain detailed history of onset, duration, severity and progression of symptoms, and possible precipitating factors
 2. Describe and monitor lesions in terms of morphology/structure, size, shape, number, color, location, distribution
 3. Discontinue contact with allergen if known sensitivity
 4. Cool compresses of Burow's solution for comfort
 5. Topical steroids to reduce immune response and pruritus, e.g., hydrocortisone, triamcinolone
 6. Oral steroids for severe and extensive reactions, e.g., prednisone
 7. Sedating oral antihistamines for nocturnal pruritus, e.g., hydroxyzine, diphenhydramine; OTC nonsedating antihistamines for daytime pruritis, e.g., cetirizine or loratadine
 8. Educate regarding characteristics of condition
 9. Educate regarding medication dosages, signs of irritation, sensitivity
 10. Teach measures for prevention, e.g., avoid known allergens and overheating
 11. Refer to dermatologist for evaluation
 - a. If acute episode does not improve
 - b. Consideration of skin testing for hypersensitivities after acute episode resolves

■ QUESTIONS

Select the best answer

1. J.D. is a postterm infant with lesions of varying morphology including wheals, vesicles, and pustules on her trunk. You suspect J.D. has:
 - a. Cutis marmorata
 - b. Erythema toxicum neonatorum
 - c. Milia
 - d. Contact dermatitis
2. In order to confirm your diagnosis of J.D., you order a Wright's stained smear. If your diagnosis is correct, what are the expected results of the smear?
 - a. Presence of eosinophils
 - b. Presence of neutrophils
 - c. Presence of keratinous material
 - d. Presence of staphylococcus bacteria
3. In addition to monitoring the skin for any changes, what is the best management for J.D.?
 - a. Topical antibiotics on lesions
 - b. Topical steroids on lesions

- c. A moisturizer on lesions
 - d. No treatment necessary since J.D.'s condition will resolve spontaneously in 5 to 7 days
4. You examine C.C., a newborn, and observe numerous white papular lesions on the cheeks, forehead, and nose. You suspect either milia or neonatal acne. Which physical finding helps to confirm a diagnosis of milia?
 - a. Papular lesions are intermixed with pale yellow macules
 - b. Papular lesions have an erythematous circular ring at the base
 - c. Papular lesions are surrounded by lacy-blue area with erythematous mottling
 - d. Papular lesions, yellow in color, are observed on the hard palate
 5. Newborn K.T. is three-weeks premature and you observe a macular erythematous lacy appearance to her skin when you undress her. K.T. has which condition?
 - a. Cutis marmorata
 - b. Erythema toxicum neonatorum
 - c. Salmon patch
 - d. Nevus flammeus
 6. In addition to monitoring the skin for any changes, what is the best management for K.T.?
 - a. Keep K.T. warm
 - b. Decrease the environmental temperature
 - c. Use a moisturizer on affected skin areas
 - d. Do nothing, as condition will resolve spontaneously in 5 to 7 days without intervention
 7. Newborn W.R. has a vascular lesion that will not fade as she gets older. What is your diagnosis?
 - a. Salmon patch
 - b. Capillary hemangioma
 - c. Café au lait
 - d. Port-wine stain (nevus flammeus)
 8. W.R.'s parents are concerned about her appearance and the psychological effect on their daughter as she becomes aware of her condition. In educating the parents, you tell them about several options. Which of the following is not an appropriate management or treatment consideration for W.R.?
 - a. Application of topical steroids to the affected area to prevent pruritus
 - b. Camouflage affected areas with cosmetics
 - c. Pulsed laser treatment of affected area
 - d. Counseling for psychological concerns
 9. Which condition is thought to be more apparent in darker-skinned individuals or during the summer months?
 - a. Tinea corporis
 - b. Psoriasis
 - c. Pityriasis alba
 - d. Pityriasis rosea
 10. J.R., an eight-year-old boy, has scaly, hyperpigmented lesions in a "fir tree" distribution, predominately on his trunk. One lesion on the buttocks is larger than all the other lesions and measures 4 cm in diameter. What is your likely diagnosis?
 - a. Psoriasis
 - b. Eczema
 - c. Pityriasis alba
 - d. Pityriasis rosea
 11. What symptom is commonly experienced in J.R.'s condition?
 - a. Pruritus
 - b. Pain at site of lesions
 - c. Nausea
 - d. Headache
 12. What management would you not recommend for J.R. with his condition?
 - a. Cool bath or cool compresses to lesions
 - b. Topical steroids to lesions
 - c. Oral antibiotics
 - d. Monitored and controlled daily sunlight exposure
 13. You have diagnosed D.L. with acute atopic dermatitis. Which of the following is not correct regarding the incidence of this condition?
 - a. D.L. is most likely an infant
 - b. D.L. has a greater chance of developing asthma later in childhood than the average individual
 - c. D.L. has a greater chance of developing malignant melanoma in adulthood than the average individual
 - d. D.L. has a condition associated with familial predisposition
 14. Which of the following management measures or treatments would you not recommend for D.L.?
 - a. Topical steroids to affected areas
 - b. Wet compresses to affected skin areas
 - c. Maintain a dry, warm environment
 - d. Eliminate all substances that dry the skin

15. In addition to having atopic dermatitis, you have diagnosed D.L. with a secondary bacterial infection at the site of several lesions. What is the best management for the infection?
 - a. Topical antibiotics to affected areas
 - b. Oral antibiotics
 - c. Hot compresses to affected areas
 - d. Monitored and controlled daily sun exposure until lesions resolve
16. You see B.D. for the first time at age six weeks. B.D. has a bright red, raised, rubbery lesion of irregular shape and 2 cm in diameter on the occiput. What condition do you suspect B.D. has?
 - a. Malignant melanoma
 - b. Port-wine stain
 - c. Capillary hemangioma
 - d. Burn
17. Which of the following is not characteristic of the lesion B.D. has?
 - a. It was not present at birth, however, B.D.'s mother noticed site was blanched
 - b. It will continue to grow for the first 9 to 12 months of B.D.'s life
 - c. It will begin to gradually resolve when B.D. is between 12 to 15 months
 - d. It is expected to completely resolve by the time B.D. is ten years old
18. You notice 10 macular tan lesions of varying sizes on D.D. and refer him for a medical evaluation to rule out neurofibromatosis or Albright's syndrome. What kind of lesion does D.D. have?
 - a. Malignant melanoma
 - b. Café au lait spots
 - c. Mongolian spots
 - d. Vitiligo
19. What is characteristic of the lesion that D.D. has?
 - a. More common in Caucasians than dark-skinned individuals
 - b. More common in males than females
 - c. Lesions can be present at birth, however, more lesions may develop at any age
 - d. Lesions usually fade spontaneously and completely resolve in adult life
20. You suspect that A.F., age nine years, has either pityriasis alba or vitiligo. Which of the following would not confirm the diagnosis of pityriasis alba?
 - a. A.F.'s skin would be normally pigmented except for areas of depigmentation
 - b. A.F.'s skin would have one or more scaly areas of hypopigmentation
 - c. A.F. complains of mild itching in areas of hypopigmentation
 - d. A.F.'s lesions became more pronounced when she was exposed to sunlight
21. A.F. was diagnosed with pityriasis alba. Which of the following is proper management of A.F.'s condition?
 - a. Bland moisturizers to reduce overdrying
 - b. Topical steroids to the affected areas
 - c. Expose affected areas to short periods of sunlight each day
 - d. Burow's wet compresses to affected areas
22. Patient education is a major part of the PNP's role. What would you teach A.F. and her parent regarding the progress and prognosis of pityriasis alba?
 - a. A.F. will continue to develop lesions for the rest of her life
 - b. A.F.'s condition should fade appreciably in three to four months
 - c. A.F.'s condition is permanent and affected areas will not repigment
 - d. A.F.'s condition will resolve completely, however, the affected areas can become slightly reddened when exposed to sunlight
23. Malignant melanoma is a form of much dreaded skin cancer. Which of the following is not characteristic of this condition?
 - a. Occurs in all ethnic groups but more commonly in light-skinned individuals
 - b. Severe sunburn or excessive exposure to sunlight before the age of ten years predisposes developing melanoma later in childhood or in adult life
 - c. Spreads through the lymphatic system and invades other distant skin surfaces and organs
 - d. Spreads primarily by invading skin surfaces that surround the major lesion
24. Which of the following does not characterize the lesion of malignant melanoma?
 - a. Irregular asymmetrical nodule with blurred borders
 - b. Raised with distinct symmetrical borders
 - c. Uneven coloring in which blue, black, brown, tan, and red may all be present in the same lesion
 - d. Bleeding, ulceration in later stages
25. Patient education regarding prevention of malignant melanoma is essential. Which of the

- following is not considered best prevention education?
- Avoid sunlight, especially during the hours of 9:00 a.m. and 1:00 p.m.
 - Avoid sun tanning lamps
 - Use cover-up clothing, hats, and sunglasses
 - Use sun blocks that protect against ultra-violet exposure with 30 SPF
26. You suspect M.N. as having chronic psoriasis. Which of the following is characteristic of her lesions if she has psoriasis vulgaris?
- Scaly erythematous patches and plaques 3 to 10 mm in diameter
 - Round or oval in shape
 - Large scaly silver-white plaque 5 to 10 cm in diameter
 - Located mainly on her trunk
27. M.N.'s condition of psoriasis is common in approximately 33% of children. Which of the following is not correct regarding the etiology or incidence of this condition?
- Occurs more commonly in dark-skinned ethnic individuals
 - Associated with constant rubbing or trauma to exposed affected areas such as elbows
 - Associated with overproduction of epithelial cells
 - Associated with epithelial cells that migrate to the skin surface much more quickly than normal
28. What would you not advise regarding the management or treatment of M.N.'s condition?
- Excise lesions
 - Apply topical steroids
 - Apply mineral oil and moisturizers
 - Expose to monitored short periods of sunlight
29. You have diagnosed Jale as having contact dermatitis. Which symptom is most characteristic of his condition?
- Headache
 - Difficulty breathing
 - Pruritus at site of affected areas
 - Pain at site of affected areas
30. Which of the following is not characteristic of Jale's condition?
- He has hypersensitivity to a substance within his environment when direct contact is made
 - He may experience a delayed reaction of several days with re-exposure to an allergen
 - His dermatitis may be caused by direct contact with topical medications, soaps, cosmetics, fabrics, and plants
 - Typical response is redness and edema at the site of contact which may progress to papules and vesicles
31. What would you not recommend as management and treatment of Jale's condition?
- Skin testing during the acute episode to determine if Jale has an allergy
 - Cool compresses of Burow's solution to affected areas
 - Topical steroids to affected areas for five days
 - Oral antihistamines
32. You diagnose Kelli, age seven months, with diaper dermatitis. Which of the following should not be included in the differential diagnosis?
- Atopic dermatitis
 - Child abuse
 - Contact dermatitis
 - Pityriasis alba
33. What management measure would you not prescribe to treat Kelli's condition?
- Oral antihistamines
 - Lubricants such as petroleum jelly to mildly affected areas
 - Low potency topical steroids to severely affected areas with erythema and papules
 - Topical antibiotics to severely affected areas with ulcerations
34. What would not be an appropriate recommendation to prevent Kelli from having subsequent episodes of diaper dermatitis?
- Expose diaper area to air several times each day
 - Increase oral fluids using orange juice to dilute urine
 - Make diaper changes immediately after soiling
 - Use a double rinse of vinegar and water for home-laundered diapers
35. Seborrhea dermatitis is common in both infants and adolescents. Which of the following is not correct of this condition?
- Can cause irritating pigment changes to include hyperpigmentation and hypopigmentation
 - Is associated with an overproduction of sebum in areas abundant with sebaceous glands

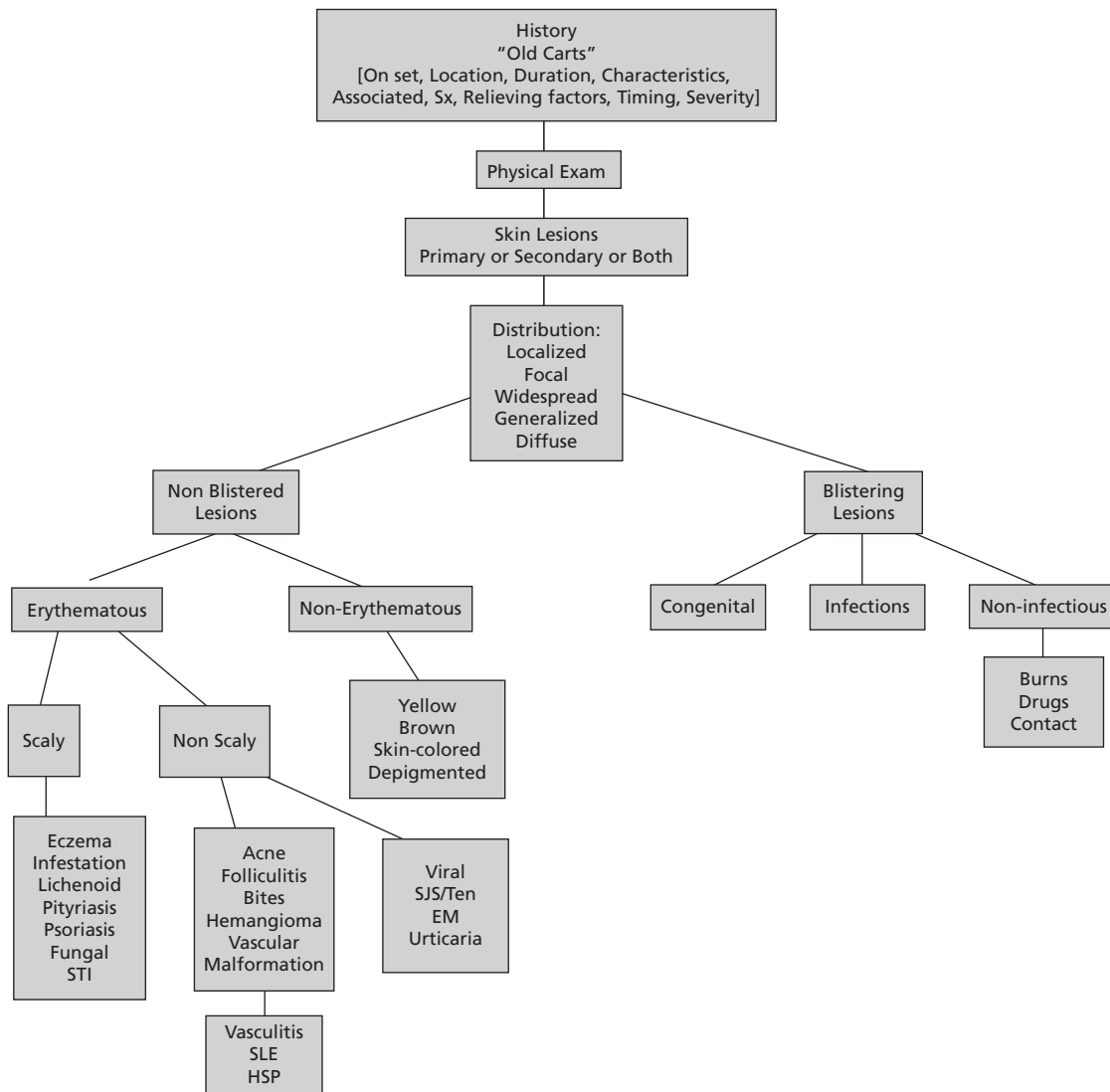
- c. The condition in infants is known as “cradle cap” in which lesions have erythematous base with yellow crusted areas and greasy scales
- d. The condition in adolescents is known as acne with comedones, papular and pustular lesions
36. What is the best treatment of seborrhea in the infant?
- Mineral oil to loosen crusts prior to washing affected areas with a nonperfumed baby shampoo
 - Topical antibiotics
 - Oral antibiotics in severe cases
 - Oral steroids for severe cases
37. You are evaluating F.P., age three years, who acutely sustained a burn when she pulled a pan of boiling water onto herself within the past hour. Since burns are classified according to the depth of injury to the skin layers and the amount of area involved, how would you rate the burn if 5% of her body surface is burned involving the epidermis and upper part of the dermis?
- She has a minor first and second degree burn
 - She has a major second degree burn
 - She has a major full thickness burn
 - She has major first and second degree burns
38. F.P.’s burn should appear:
- Dry, with mild edema and erythema
 - Dry whitish areas that blanch with pressure
 - Dry whitish to brownish areas with edema
 - Moist with edema, erythema, and a few vesicles
39. What is the best treatment for F.P.’s burn?
- Warm compresses to affected areas and mild analgesic for discomfort
 - Topical emollients to affected areas
 - Topical steroids to affected areas
 - Refer for urgent treatment in an ED
40. Jerry has been diagnosed as having folliculitis, an inflammatory condition involving the pilosebaceous follicle. What is the most common cause of this condition?
- Microsporum canis* tinea
 - Poxvirus
 - Staphylococcus aureus*
 - Streptococcus* group A
41. Jerry has a condition that most commonly occurs on which body surface?
- Neck and scalp
 - Upper arms
 - Chest and abdomen
 - Legs
42. You order a culture and the results confirm that Jerry’s condition is caused by the most common organism for this condition. What treatment do you prescribe?
- Oral penicillin
 - Dicloxacillin
 - Tinactin
 - Tretinoin
43. Sandra, age twelve years, has several vesicles and honey-colored crusted lesions on her face above the right nares. She has a history of having had a scratch in the same area several days ago. What condition do you suspect?
- Acne
 - Impetigo
 - Herpes simplex
 - Eczema
44. Judy, age fifteen years, has been diagnosed as having acne. Which of the following is not true of this condition?
- Poor hygiene is the primary cause of acne
 - Associated with increased androgenic hormonal activity
 - Females can have a “cyclic” component to their acne
 - Severe acne having a later onset in puberty is more common among males
45. Judy has a history of remission and exacerbation of acne that has followed the pattern of menses for two years. However, the condition over the last six months has worsened to a moderate degree of severity and has been chronic and persistent. You prescribe antibiotic therapy. Which of the following antibiotics would you not consider?
- Topical clindamycin
 - Oral erythromycin
 - Oral minocycline
 - Oral tetracycline
46. K.C., age thirteen years, has several firm, small (2 mm), white or skin-colored umbilicated papules on her neck. The lesions have been present for three months and have increased in number. What is your diagnosis?
- Acne
 - Molluscum contagiosum*
 - Warts
 - Cellulitis

47. What is the cause of K.C.'s condition?
- Microsporium canis tinea
 - Poxvirus
 - Staphylococcus aureus
 - Streptococcus group A
48. Which treatment would you not recommend for K.C.'s condition?
- Curettage lesions
 - Oral antibiotics
 - Observation
 - Topical Imiquimod
49. Paul has four superficial lesions on his anterior lower abdomen of one week duration. The lesions are 4 cm in diameter, scaly, irregular shaped plaques with skin colored centers and erythematous borders. The affected areas are slightly pruritic. What condition do you suspect Paul has?
- Psoriasis
 - Eczema
 - Tinea corporis
 - Pityriasis rosea
50. You performed two tests to confirm your diagnosis of Paul's condition. The KOH scraping was positive for the presence of hyphae. The Wood's lamp did not fluoresce the lesions. You are sure that Paul's condition was not caused by which organism?
- Epidermophyton floccosum
 - Microsporium canis
 - Trichophyton tonsurans
 - Trichophyton rubrum
51. You see Paul after eight weeks of treatment with a topical antifungal preparation. The original lesions have almost resolved, however, the condition has worsened with the development of several other larger lesions on the abdomen and groin area. Which of the following would you not consider?
- Oral antifungal medication, griseofulvin
 - Topical antibiotic preparation
 - Continue with the topical antifungal applications
 - Educate again regarding not sharing personal items
52. Dale, age 7 years, is complaining of pain and burning on his right leg where you observe two small red puncture marks surrounded by a blanched area with an erythematous border. He had been playing with his dog all morning outside in a grassy wooded area near his home and was wearing shorts. You suspect he has been bitten by which insect?
- Mosquito
 - Bee
 - Recluse spider
 - Black widow spider
53. Which of the following is not true of insect stings from bees, wasps, and fire ants?
- Greater reaction of hypersensitivity occurs most often with the initial exposure than with subsequent exposures
 - For mild reactions, cool compresses to the site of injury is the usual management
 - Occurs more often during the spring and summer months
 - Most stings occur in self-defense when the nonaggressive insect feels threatened or irritated
54. You diagnose W.A. with scabies. Which of the following is not characteristic of this condition?
- He has several erythematous papular, pustular, and crusted lesions on his face
 - He has several excoriated scratched areas around the umbilicus and waist area
 - He has several linear curved lines approximately 4 mm in length with a papule at the proximal end linear line
 - He complains of severe pruritus which is worse at night
55. Which of the following is not recommended as a management and treatment strategy for W.A.?
- Put nonwashable items in a plastic bag and store for one week
 - Prescribe topical antifungal applications
 - Prescribe topical antiparasitics
 - Prescribe topical steroids and/or oral antihistamines for pruritus
56. Pediculosis is a highly communicable, common condition in children. Which of the following is not correct of pediculosis humanus?
- Caused by an insect that does not fly or jump
 - Gravid females lay ova in seams of clothing
 - Likes hairy areas of the body better than the non-hairy body surfaces
 - Same medication used for scabies may be used to effectively eradicate this condition

57. Hypersensitivity may occur to a variety of substances causing a variety of reactions. It is important to determine if the body's hypersensitivity reaction will cause erythema multiforme condition. Which of the following is not typical of the erythema multiforme reaction?
- Target "bull's-eye" lesion with a necrotic center surrounded by a pale macular middle area and then by an erythematous peripheral ring
 - Itching at site of affected skin areas
 - Pain at site of affected areas, especially in the oral cavity
 - Lesions which all have the same morphology on the trunk
58. You see D.Y. in your clinic and suspect she has a form of erythema multiforme. Erythema multiforme minor must be differentiated from erythema multiforme major. Which of the following is the most important confirming evidence for making a diagnosis of erythema multiforme major?
- Presence of deeper lesions within the dermis
 - Presence of lesions on the exposed areas of the body
 - Presence of pustules indicating a secondary infectious process
 - Occurrence of prodromal systemic symptoms of fever, malaise, sore throat, headache, nausea, and/or vomiting
59. You suspect D.Y. has erythema multiforme major. What treatment or management is most indicated?
- Prescribe topical antibiotics due to secondary infection
 - Prescribe topical steroids to lesions for pruritus
 - Refer for medical evaluation
 - No treatment is indicated as condition will resolve spontaneously in one week
60. Urticaria is a hypersensitivity allergic reaction to a variety of substances and agents. You suspect W.P. has urticaria due to the typical morphology of lesions on her trunk and arms which are:
- Erythematous papules
 - Vesicles
 - Pustules
 - Wheals
61. During W.P.'s acute episode of urticaria which of the following is not considered an appropriate management or treatment measure?
- Oral antibiotics to prevent secondary infection
 - Oral antihistamines for pruritus
 - Topical steroids to affected areas to reduce the immune response
 - Cool compresses to affected areas

▣ ANSWERS

- | | |
|--------------|--------------|
| 1. b | 32. d |
| 2. a | 33. a |
| 3. d | 34. b |
| 4. d | 35. d |
| 5. a | 36. a |
| 6. a | 37. d |
| 7. d | 38. d |
| 8. a | 39. d |
| 9. c | 40. c |
| 10. d | 41. a |
| 11. a | 42. b |
| 12. c | 43. b |
| 13. c | 44. a |
| 14. c | 45. b |
| 15. b | 46. b |
| 16. c | 47. b |
| 17. b | 48. b |
| 18. b | 49. c |
| 19. c | 50. b |
| 20. a | 51. b |
| 21. a | 52. d |
| 22. b | 53. a |
| 23. d | 54. a |
| 24. b | 55. b |
| 25. a | 56. c |
| 26. c | 57. d |
| 27. a | 58. d |
| 28. a | 59. c |
| 29. c | 60. d |
| 30. b | 61. a |
| 31. a | |



■ REFERENCE MATERIALS

DERMATOLOGY TERMINOLOGY

PRIMARY	SECONDARY
Macule	Scale
Non palpable	Crust ["scabbed"]
Papule	can be hemorrhagic
Palpable	can be moist
< 1 cm in diameter	Excoriation [scratched, rubbed]
Pustule	Oozing
Pus-filled papule	Erosion
Open comedone	Ulceration
["blackhead"]	
Closed comedone	
["whitehead"]	
Nodule	
Raised, palpable	
Deep; in dermis	
Tumor	
A large nodule	
Wheal	
Edematous	
Slightly raised	
Plaque	
Raised, flat topped	
> 1 cm in diameter	
Vesicle	
Fluid-filled papule	
Cyst	
Raised	
Fluid-filled sac	
Bulla	
A cyst > 1 cm in size	

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Gastrointestinal Disorders

Lindsay Wilson

■ GASTROESOPHAGEAL REFLUX IN INFANCY (GER)

- Definition: Reflux of gastric contents into esophagus, due to immaturity of lower esophageal sphincter; normal physiologic process that occurs throughout the day in healthy infants, children and adults; GER occurs during periods of transient relaxation of the lower esophageal sphincter
- Etiology/Incidence
 1. Etiology multifactorial
 - a. Occurs in 50% of infants in first 3 months of life, 67% of 4-month-old infants and 5% of 10- to 12-month-old infants.
 - b. Resolves as child becomes more upright and starts solids
 - c. Often caused by overfeeding or incomplete burping
 - d. Childhood diagnoses with higher incidence of GERD include: neurological impairment and delay, esophageal atresia, hiatal hernia, bronchopulmonary dysplasia, asthma, and cystic fibrosis
- Signs and Symptoms
 1. Physiologic GER
 - a. Effortless, painless spitting of varying amounts, often within 40 minutes of eating
 - b. No choking or color changes
 - c. Normal growth—growth chart is key
 - d. Feeding history may indicate excessive intake; burp heard during vomiting may indicate incomplete burping
- 2. Gastroesophageal Reflux Disease (GERD)
 - a. Reflux may cause other physical complications, such as:
 - (1) Failure to thrive (FTT)—caused by long-term, forceful regurgitation
 - (2) Esophagitis—causing irritability, anemia, and guaiac positive stools or hematemesis; dysphagia
 - (3) Aspiration—pneumonia, wheezing, apnea
 - (4) Sandifer syndrome—abnormal posturing of head and neck
 - b. May be “silent GER”—no overt vomiting, but complications may be presenting symptom
 - c. 60% show improvement by 16 months; 30% may remain symptomatic up to 4 years
- Differential Diagnosis
 1. Pyloric stenosis
 2. (Partial) anatomical obstruction
 3. Formula intolerance (cow milk protein or soy allergy)
 4. Eosinophilic esophagitis
 5. Gastroenteritis
 6. Infections—urinary tract infection (UTI); otitis media (OM); pneumonia
 7. Increased intracranial pressure (ICP)/neurological disorder
- Physical Findings
 1. May have wheezing or respiratory symptoms with aspiration

2. Abdominal examination normal—no masses, olive, or peristaltic waves
 3. Neurological examination normal—no signs of increased ICP
- Diagnostic Tests/Findings
 1. Diagnosis often made by observation and history; testing only to determine if reflux is causing problems since vomiting indicates reflux
 2. UGI to ligament of Treitz—only evaluates anatomy, not sensitive or specific for GER; use to rule out anomalies of the GI tract such as malrotation
 3. pH probe—indicates amount of reflux occurring; does not confirm diagnosis; can determine association of reflux with recurring symptoms
 4. Upper endoscopy—assesses presence and degree of esophagitis, can diagnose other disorders such as eosinophilic infiltration associated with allergy (eosinophilic esophagitis), infectious esophagitis, etc.
 5. Scintigraphy—“Milk scan;” evaluates for slow gastric emptying and aspiration; lack of standardized techniques and age-specific norms limit usefulness of test
 6. Guaiac stool/emesis—positive for occult blood if abnormal
 7. Empiric medical therapy—trial of medication to relieve specific symptoms. If symptom is relieved with medication then GER can be determined to be the cause
 - Management/Treatment
 1. Conservative therapy
 - a. Positioning—(most important) postprandial, prone position for 1 to 2 hours if infant can be observed; infant seats/swings worsen by increasing intra-abdominal pressure; caution with diapering/playing postfeeding
 - b. Breastfeeding or predominately whey formula
 - c. A 1 to 2 week trial of hypoallergenic formula may be warranted if vomiting or other symptoms severe enough to consider drug treatment
 - d. Thickening agents such as rice cereal have not been proven to decrease reflux but may decrease vomiting
 - e. Avoid over-feeding—age in months plus 3 equals number of ounces every 3 to 4 hours for most infants to age 5 months
 - f. Small feedings with frequent burping
 - g. Reassure parents with growth charts
 - h. Decrease anxiety in mother-infant interaction
 - i. Monitor for problems—aspiration/esophagitis
 - j. Since the risk of SIDS is greater than the risk of aspiration from birth to 12 months, nonprone position (preferably supine) during sleep to prevent SIDS is the recommendation of the AAP except in very severe cases of GER
 - 2. Medications: (if conservative therapy has failed)
 - a. H₂ blockers (1st line), proton pump inhibitors (2nd line), and antacids (short term) if irritable from esophagitis (aluminum-containing antacids increase plasma aluminum levels in infants; there are no studies on safety of magnesium or calcium carbonate preparations, antacids should not be used long term)
 - b. Cisapride has been removed from market for most patients
 - c. Metoclopramide may be cautiously considered for severe reflux under the supervision of a physician or gastroenterologist—“black box” warning from FDA; can cause tardive dyskinesia, a serious irreversible movement disorder
 - d. Erythromycin 2 mg/kg up to 4 times prior to meals can promote gastric emptying (motilin agonist), should only be used infants over 4 weeks of age because of increased incidence of pyloric stenosis when given in first few weeks of life
 - 3. Surgery—Nissen fundoplication only as last resort in an infant with GERD

■ PYLORIC STENOSIS

- Definition: Obstruction due to thickening of circular muscle of the pylorus
- Etiology/Incidence
 1. Unknown cause, genetic disposition, environmental factors
 2. Young infants exposed to erythromycin in the first few weeks of life are at increased risk for developing pyloric stenosis
 3. Occurs in 3:1000 infants; male > female; more likely in first-born males
 4. Familial predisposition—25% chance if mother had pyloric stenosis, 15% chance if other family member, 22% if identical twin
 5. More common in Caucasians than African-Americans or Asians

6. Symptoms occur later in breastfed infants; greater muscle thickness required to obstruct smaller-sized breastmilk curd
 7. Delayed timing in premature infants
- Signs and Symptoms
 1. Not present at birth; may occur in first week; average age of presentation from 3 to 6 weeks through 3 to 4 months of age
 2. Vigorous, nonbilious vomiting after eating; with time becomes projectile with brownish color
 3. “Hungry” after emesis; progressing to lethargy and irritability
 4. Weight loss or poor weight gain
 5. Constipation
 6. Dehydration, metabolic alkalosis, malnutrition, and gastritis can occur with prolonged symptoms
 - Differential Diagnosis
 1. Overfeeding
 2. Gastroesophageal reflux
 3. Milk protein allergy
 4. Gastroenteritis
 5. Malrotation/volvulus if bilious emesis
 - Physical Findings
 1. Visible peristaltic waves progressing from left to right across abdomen—darken room, shine bright light on abdomen of naked, supine baby; feed bottle of sugar water; peristaltic waves visible
 2. Palpable pyloric “olive” after vomiting—palpate epigastrium in RUQ deep under liver edge; need very relaxed abdomen; hard, smooth, mobile, nontender mass may be palpable
 3. Dehydration as obstruction increases
 - Diagnostic Tests/Findings
 1. Abdominal ultrasound to determine size of pylorus—preferred test

2. Upper GI—avoid due to risk of barium aspiration; shows “string sign;” elongated pyloric channel and delayed gastric emptying
3. Electrolytes to determine dehydration status

- Management/Treatment

1. Surgical treatment after correction of fluid and electrolyte deficits
2. Postoperative monitoring for hypoglycemia
3. Excellent prognosis

■ ACUTE INFECTIOUS GASTROENTERITIS

- Definition: Illness of rapid onset, includes diarrhea with possible nausea, vomiting, fever, or abdominal pain
- Etiology/Incidence
 1. 70% to 80% caused by viral agents; 25% by rotavirus
 2. < age 3 years 1.3 to 2.3 episodes/year; higher if in childcare facility
 3. Predisposing factors (see **Table 8-1**)
 - a. Child-care facility
 - b. Poor sanitation—improper hand washing, food preparation, or water quality
 - c. Recent travel, especially to endemic areas
 - d. Ill contacts—animals or humans
 - e. Immunocompromised children at risk
 - f. Recent antibiotic use
- Signs and Symptoms
 1. Rotavirus—nonbloody diarrhea, preceded or accompanied by vomiting and fever; symptoms last 3 to 8 days; dehydration may develop in severe cases
 2. Adenovirus—URI tract infection most common; children less than 4 susceptible to enteric infection; symptoms similar to rotavirus, but last longer
 3. Norwalk—nausea, fever, abdominal cramps, headache, malaise, myalgia; vomiting more frequent than diarrhea

■ **Table 8-1** Most Common Causative Agents in U.S.

Agent	Source/Transmission	Risk Factors	Incubation	Duration
VIRUS				
<i>Rotavirus</i>	Infected persons; contaminated toys; hard surfaces	< 3 years; childcare; low SES*; winter	2–4 days	3–8 days
<i>Adenovirus</i>	Infected person; contaminated toys; hard surfaces	< 4 years; childcare; not seasonal	3–10 days	5–12 days
<i>Norwalk</i>	Infected persons; contaminated food or water	older children & adults	12 hrs–4 days	12–48 hrs

continues

■ Table 8–1 Most Common Causative Agents in U.S. (continued)

Agent	Source/Transmission	Risk Factors	Incubation	Duration
BACTERIA				
<i>Shigella</i>	Infected persons; contaminated food, water, objects; houseflies	1–4 years; childcare; crowding; poor sanitation; summer/fall months, need very few organisms for transmission	1–7 days	48–72 hours, even without treatment
<i>Salmonella</i> (nontyphoidal)	Contaminated foods (poultry, red meat, eggs, milk, fruits); contaminated water; infected persons or animals, pet snakes, turtles	< 5 years; especially first year	6–72 hrs	2–7 days
<i>Salmonella typhi</i> (typhoid fever)	Infected persons; rare in U.S., may be acquired during international travel	All ages	7–14 days	2–21 days
<i>Campylobacter jejuni</i>	Contaminated poultry; unpasteurized milk; pets	All ages; summer months	1–7 days	5–7 days
PARASITIC				
<i>Giardia lamblia</i>	Infected persons, animals; contaminated food or water	Childcare, institutions; campers	1–4 weeks	May be long-term
<i>Cryptosporidium</i>	Infected persons, animals; contaminated water supplies	Parasite is chlorine resistant	2–14 days	1–20 days
<i>Entamoeba histolytica</i>	Mebic cysts; contaminated food and water	Tropical areas; crowding; poor sanitation	1–4 weeks	May be long-term
TOXIN				
<i>S. aureus</i>	Ingestion of contaminated food products: ham, poultry, salads (egg and potato), cream filled pastries	Inadequate cooking, refrigeration	$\frac{1}{2}$ –6 hrs	1–2 days
<i>E. coli</i> (O157:H7)	Infected persons or carriers; contaminated foods (undercooked ground beef, apple cider, raw vegetables, salami, yogurt, water)	Childcare; poor sanitation; inadequate cooking; poor handwashing; travel to developing countries	3–4 days, up to 8 days	Varies
<i>Clostridium difficile</i>	Infected persons or hospital environment	Hospitals, childcare, normal intestinal flora altered—antibiotics; repeated enemas, prolonged NG tube; intestinal surgery, inflammatory bowel disease; c.dif disease rare < 12 months	Unknown	May improve with discontinuation of antibiotic if cause

*SES—socioeconomic status

Millonig, V.L., & Mobley, C. (Eds.). (2004). Pediatric nurse practitioner certification review guide, (4th ed.). Sudbury, MA: Health Leadership Associates, Inc.

4. *Shigella*—high fever, headache, abdominal pain and tenderness; large, watery stools in which blood and mucus may be seen; can lead to dehydration
 5. *Salmonella*—fever, abdominal pain and cramps; watery, mucoid, or bloody stools
 6. *Campylobacter jejuni*—fever, malaise, abdominal pain, can mimic appendicitis or intussusception, bloody stools
 7. *Giardia lamblia*—flatulence, abdominal pain, failure to thrive, anorexia, range of stools, e.g., asymptomatic carrier to foul, steatorrhea, consider in persistent diarrhea (> 7 days)
 8. *Cryptosporidium parvum*—frequent watery stools most common symptom with abdominal pain, anorexia and weight loss, fever, vomiting common; consider in persistent diarrhea (> 7 days)
 9. *Entamoeba histolytica*—asymptomatic, mild symptoms, e.g., abdominal distention, constipation, occasionally loose stools; or severe abdominal pain with increasingly severe bloody and mucoid diarrhea, weight loss, fever in approximately $\frac{1}{3}$ of patients
 10. *Staphylococcus aureus*—abrupt onset of nausea, vomiting, abdominal pain, watery stools
 11. *Escherichia coli* (EHEC 0157:H7)—fever < $\frac{1}{3}$ of cases, severe abdominal pain, cramping, watery diarrhea, stools usually progress to grossly bloody or occult; hemolytic uremic syndrome (HUS) can occur 1 week or more after diarrhea
 12. *Clostridium difficile*—abdominal pain and cramps; pseudomembranous colitis, stools bloody with leukocytes, mucus, pus; symptom free carrier state common < 1 year
- Differential Diagnosis
 1. Urinary tract infection (UTI)
 2. Other infections—otitis media, streptococcal pharyngitis
 3. Inflammatory bowel disease
 4. Malabsorption (lactose intolerance, celiac disease, cystic fibrosis)
 5. Milk protein allergy
 6. Chronic/functional diarrhea
 7. If only vomiting:
 - a. Trauma
 - b. Congestive heart failure
 - c. Toxic ingestion
 - d. Metabolic disorder
 - e. Increased intracranial pressure
 - Physical Findings
 1. Assess hydration—see **Table 8-2**
 2. Recent weight (wt) very helpful, but often not available. Preillness wt minus wt today/preillness wt $\times 100 =$ fluid deficit as % body wt loss

■ **Table 8-2** Assessment of Dehydration

Variable	Mild, 3%–5%	Moderate, 6%–9%	Severe $\geq 10\%$
Blood pressure	Normal	Normal	Normal to reduced
Quality of pulses	Normal	Normal or slightly decreased	Moderately decreased
Heart rate	Normal	Increased	Increased*
Skin turgor	Normal	Decreased	Decreased
Fontanel	Normal	Sunken	Sunken
Mucous membranes	Slightly dry	Dry	Dry
Eyes	Normal	Sunken orbits	Deeply sunken orbits
Extremities	Warm, normal capillary refill	Delayed capillary refill	Cool, mottled
Mental status	Normal	Normal to listless	Normal to lethargic or comatose
Urine output	Slightly decreased	< 1 mL/kg/hr	< 1 mL/kg/hr
Thirst	Slightly increased	Moderately increased	Very thirsty or too lethargic to indicate

Note: From American Academy of Pediatrics, Provisional Committee on Quality Improvement, Subcommittee on Acute Gastroenteritis. (1996). Practice parameters: The management of acute gastroenteritis in young children. *Pediatrics*, 97(3), 427.

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The percentages of body weight reduction that correspond to different degrees of dehydration will vary among authors. The critical factor in assessment is the determination of the patient's hemodynamic and perfusion status. If a clinician is unsure of the category into which the patient falls, it is recommended that therapy for the more severe category be used.

*Bradycardia may appear in severe cases.

- Diagnostic Tests/Findings
 1. Testing not necessary unless:
 - a. Blood or mucus in stools—test for specific organism
 - b. No improvement in symptoms > 5 to 6 days
 - c. Signs of severe dehydration—BUN, specific gravity, electrolytes
 2. Test for specific organism
 - a. Virus—EIA, rarely necessary
 - b. Bacterial—stool culture; specific testing for *E. coli* 0157:H7 if bloody stools
 - c. Giardia—Giardia antigen (more specific); stools for ova and parasites
 - d. Cryptosporidium—stools for ova and parasites, specify cryptosporidium
 - e. *Clostridium difficile*—*Clostridium difficile* antigen, toxins A and B
 3. Persistent vomiting as only sign; diarrhea lasting longer than 10 days or with failure to thrive (FTT) need more extensive testing
- Management/Treatment
 1. Self-limiting in most healthy children
 2. Increased risk associated with dehydration with fever; prematurity, infancy, and adolescent mothers
 3. Assess degree of dehydration and correct deficit following the guidelines suggested by the AAP Subcommittee on Acute Gastroenteritis
 - a. Oral rehydration therapy (ORT) can safely and effectively treat mild to moderate dehydration in otherwise healthy child
 - b. Maintenance solutions have 45 to 50 mmol/L of sodium, suitable for rehydration; continue for a maximum of 24 hours
 - c. Determine replacement volume; give over 4 hour period
 - (1) 50 cc/kg for mild dehydration
 - (2) 80 to 100 cc/kg for moderate to severe
 - (3) Plus replace ongoing losses:
 - (a) 5 to 10 cc/kg for each diarrheal stool
 - (b) 2 cc/kg for each episode of emesis
 - d. Small frequent feedings are key—1 teaspoon every 1 to 2 minutes initially; if tolerated, amount may be advanced; do not allow child to drink large amounts quickly; process is labor intensive; parent or staff needs to be available to administer
 - e. Home remedies such as juice or sports beverages are nonphysiologic and should be avoided in the treatment of young children with dehydration
 - f. Once rehydrated or in children with diarrhea but no dehydration, feeding with age-appropriate diet should be encouraged; rice, wheat, potatoes, bread, cereals, lean meats, yogurt, fruits, and vegetables; early refeeding promotes healing of the GI mucosa and decreases stool output; old practice of “bowel rest” may worsen condition
 - g. In a change from earlier recommendations, once dehydration is corrected, full strength formula or milk can be given; monitor for the 20% of patients who develop transient (4 to 8 weeks) lactose intolerance
 - h. Breastmilk may be continued
 - i. Antidiarrheal medications are not appropriate and may be dangerous
 - j. UA and CBC with smear, BUN and creatinine to monitor *E. coli* (0157:H7) infection for changes indicative of HUS; may develop microangiopathic hematologic changes and/or nephropathy; close follow-up essential because potential long-term sequelae so severe
 4. Antimicrobials only in select cases
 - a. Shigella—disease generally self-limiting within 48 to 72 hours; antibiotics will shorten course of diarrhea and decrease shedding of organisms; children in childcare, large group settings, immunosuppressed, or who have severe disease should be treated; sensitivities must be done due to increased resistance; TMP/SMX (first line) or azithromycin (in TMP/SMX resistant areas) for 5 days; no return to daycare until 2 repeat cultures at least 24 hours apart, 48 hours after antibiotics completed are negative; household contacts must be cultured also
 - b. Salmonella—only for infants < 3 months or patients with suspected sepsis or immunosuppressed since antibiotics can prolong excretion; amoxicillin, TMP/SMX, cefotaxime, or ceftriaxone
 - c. *Campylobacter jejuni*—only helpful if caught in early stages; erythromycin or azithromycin 5 to 7 days; doxycycline > age 8
 - d. *Giardia lamblia*—metronidazole, tinidazole, or nitazoxanide drugs of choice; metronidazole 15 mg/kg/day in 3 divided doses for 5 days; tinidazole 50 mg/kg (up to 2 g) as a single dose; nitazoxanide 100 mg twice daily in children 1 to 3 years, 200 mg twice a day in children 3 to 11 years, and 500 mg twice a day for children over 12 years for 3 days; urazolidone 6 mg/kg/day in 4 divided doses for 7 to 10 days is less effective, but available in liquid form for children; contraindicated with G6PD deficiency and neonates; albendazole (available as liquid) for children > age 2,

- 400 mg every day for 5 days; all *Giardia* treatments are potential teratogens in pregnant teens; paromomycin (less effective) useful in pregnancy, 25 to 35 mg/kg/day in 3 doses for 7 days
- e. *Entamoeba histolytica*—metronidazole 35 to 50 mg/kg/day in 3 divided doses for 7 to 10 days followed by iodoquinol or paromomycin
 - f. *E. coli* (O157:H7)—antibiotics may not prevent hemolytic uremic syndrome (HUS); may increase risk but studies are inconclusive, AAP recommends no antimicrobial treatment
 - g. *Clostridium difficile*—Stop the causative antibiotic once diagnosed and if no improvement once antibiotic stopped then start metronidazole 30 mg/kg/day (maximum 2 g/day) in four divided doses for 7 to 10 days; vancomycin 40 mg/kg per day in 4 divided doses, maximum 500 mg for 10 days can be used for patients who do respond to metronidazole
 - h. *Cryptosporidium parvum*—self limiting diarrhea, average 10 days; nitazoxanide 100 mg twice daily in children 1 to 3 years, 200 mg twice a day in children 3 to 11 years, and 500 mg twice a day for children over 12 years for 3 days
5. Severe dehydration/HUS requires physician referral
 6. Prevention
 - a. Teach children importance of frequent, thorough hand washing
 - b. Encourage mothers to breastfeed
 - c. Childcare centers need strict policies for hand washing and food preparation; surfaces and fomites need frequent cleaning with chlorine-based disinfectant
 - d. Careful food preparation and storage
 - e. *E. coli* (O157:H7), shigella, amebiasis (*entamoeba histolytica*), campylobacter, cryptosporidiosis, giardiasis, and salmonella require public health involvement particularly for childcare attendees; may vary from state to state
 - f. Teach parents signs/symptoms of dehydration and early at home measures

■ PINWORMS (ENTEROBIASIS VERMICULARIS)

- Definitions: Nematode parasite with infestation of intestines and rectum
- Etiology/Incidence
 1. Human pinworm is ubiquitous; adult worm lives in rectum, comes out at night to lay

eggs on perianal skin and dies causing pruritus; scratching and finger to mouth contact transfers eggs to intestine; these develop into mature worms and repeat cycle (approximately 2 weeks)

2. Found in children of all socioeconomic classes
 3. Eggs float easily in air and can be swallowed by others
- Signs and Symptoms
 1. Nocturnal anal itching
 2. Vaginal itching (pinworm crawls into vagina)
 3. Insomnia (itching)
 4. Worm-like “threads”—seen in toilet or on underwear
 - Differential Diagnosis
 1. Vulvovaginitis secondary to local irritation
 2. Poor hygiene
 - Physical Findings
 1. Excoriation of perianal and perineal area
 2. Thread-like worms will be seen on visualization of anus (early morning using flashlight)
 - Diagnostic Test/Findings: Adhesive cellophane tape “paddle” with kits available for parental use; or can be made with clear Scotch tape and glass slide; prior to arising and bathing, paddle is pressed against anus and then examined for eggs
 - Management/Treatment
 1. Medication (over age 2 and non pregnant)
 - a. Pyrantel pamoate 11 mg/kg one dose (maximum dose 1 g); repeat in 2 weeks
 - b. Mebendazole 100 mg single dose (same dose for all ages and weights); repeat in 2 weeks
 2. Reassure parents ubiquitous nature of organism; reinfection likely
 3. Test other family members and treat at same time if infected
 4. Prevention
 - a. Keep nails clean and short
 - b. Bathing will remove eggs from skin and decrease pruritus
 - c. Excellent hand washing

■ INFLAMMATORY BOWEL DISEASE (IBD)

- Definition: Chronic intestinal inflammation with two specific entities of ulcerative colitis (UC) and Crohn’s disease (CD); may have extraintestinal symptoms and acute or insidious onset
 1. Location of inflammation in GI tract—CD occurs in any part of GI tract, terminal ileum typical, inflammation extends through entire

- thickness of intestinal wall, and strictures and fistulae may develop; UC only affects the lining of the colon
2. Pattern of inflammation—CD skip pattern; discrete areas of inflammation interspersed with normal mucosa; UC mucosal and submucosal inflammation, diffuse and continuous
- Etiology/Incidence (for both types of IBD)
 1. Etiology—genetic disposition, environmental factors, and alteration in intestinal flora
 2. Genetic link—30% of children with IBD have a positive family history
 3. Occurs more often in Caucasians than African-Americans and Asians; highest in descendants of Ashkenazic Jews
 4. Age of onset 25% of cases diagnosed before 20 years of age; IBD can begin in infancy, but most commonly diagnosed in children between 10 to 20 years of age
 - Signs and Symptoms
 1. Symptoms may be acute or unrecognized for years, dependent on location of lesions; more variability with CD since any part of GI tract can be involved
 2. Diarrhea
 - a. CD—loose with blood if colon involved, or can have pain but no diarrhea
 - b. UC—mild to profuse bloody diarrhea
 3. Weight loss/delayed pubertal maturation—often due to inadequate food intake because eating causes cramps, bloating, diarrhea
 - a. Growth failure may be only presenting problem, especially in CD
 - b. Weight loss and delayed puberty more common with CD
 4. Abdominal pain
 - a. CD—located in right lower quadrant sometimes as fullness or mass; food related
 - b. UC—left lower abdomen
 5. Severe cramps, low-grade fevers, anorexia
 - Differential Diagnosis (based on area of bowel involved)
 1. Ulcerative Colitis
 - a. Enteric infection (particularly those involving bloody diarrhea)
 - b. Irritable bowel syndrome (IBS)
 2. CD (particularly if manifested as extraintestinal symptoms)
 - a. Rheumatoid arthritis
 - b. Acute appendicitis
 - c. Lupus erythematosus
 - d. Lactose intolerance
 - e. Celiac disease
 3. Crohn's Disease
 - f. Infection—tuberculosis, *C. difficile*, yersinia, campylobacter
 - g. IBS
 - Physical Findings (common to both UC and CD unless noted)
 1. Weight deceleration or poor growth
 2. Diffuse abdominal pain or no tenderness
 3. Extraintestinal symptoms
 - a. Fever of unknown origin (FUO)
 - b. Short stature
 - c. Uveitis/iritis
 - d. Aphthous stomatitis
 - e. Arthritis/arthralgias
 - f. Inflammatory lesions of skin
 - g. Liver disease
 - h. Perianal fissures/tags/abscesses with CD
 - Diagnostic Tests/Findings
 1. Blood studies—CBC with differential shows microcytic anemia, increased WBC; sedimentation rate (ESR) elevated; chemistry panel shows low serum total protein and albumin; findings suggest IBD and indicate the need for further studies for both CD and UC
 2. Stool studies—infectious agents that cause bloody diarrhea for both CD and UC
 3. Endoscopy and colonoscopy with biopsy—for diagnosis and to differentiate Crohn's vs. UC
 4. Upper GI with small bowel—shows IBD changes and extent of disease in small intestine in areas not accessible by endoscopy
 - Management/Treatment
 1. Refer to pediatric gastroenterologist
 2. Nutritional therapy to ensure adequate growth and pubertal development; total parenteral nutrition (TPN) or elemental formula may be necessary and can improve clinical symptoms; in milder cases, high protein, high carbohydrate, normal fat diet providing 75 to 90 kcal/kg/day; avoid overly restrictive diets
 3. Anti-inflammatory agents
 - a. Induction of remission—corticosteroids, biologic agents
 - b. Maintenance—mesalamine, immunosuppressants (6-mercaptopurine, azathioprine); biologics-infliximab
 4. Long-term patients may require surgery, often ileocectomy in CD and colectomy/ostomy; curative for UC
 5. At higher risk for colorectal cancer
 6. Need emotional support to deal with chronic illness with reassurance that emotional factors are not primary cause

■ CHRONIC DIARRHEA

- Definition: Gradual or acute increase in the number and volume of stools that are liquid/watery
 1. Noninfectious causes
 - a. Antibiotic therapy—caused by alteration or eradication of usual intestinal flora, diarrhea usually watery, not associated with systemic symptoms, probiotics may help in its resolution; *C. difficile* infection occurs in 0.2 to 10% of patients treated with antibiotics
 2. Extraintestinal infections—infections of urinary tract and upper respiratory tract (otitis media) are sometimes associated with diarrhea
 3. Allergy to whole cow's milk or other foods
 4. Functional or nonspecific diarrhea—daily painless, recurrent passage of 3 or more large, unformed stools for 4 or more weeks; most common cause of diarrhea in young children; assess for overfeeding, excessive fruit juice or sorbitol consumption, excessive carbohydrate ingestion with low fat intake; older children may have diarrhea associated irritable bowel syndrome
 5. Malabsorption (see below)
 6. Secretory diarrhea—very watery, voluminous stools that do not resolve when child is fasting; due to rare disorders, including malignancies (neuroblastoma and GI neuroendocrine tumors), autoimmune enteropathy, microvillus inclusion disease

■ MALABSORPTION

- Definition: Impaired intestinal absorption of nutrients and electrolytes
- Etiology/Incidence: many causes—classified according to stage of digestion affected
 1. Intraluminal phase—exocrine pancreatic deficiency; cystic fibrosis most common cause of pancreatic deficiency in children (see Lower Respiratory Disorders, Chapter 6)
 2. Intestinal phase—abnormalities of mucosal surface area; absorption (celiac disease); inflammation, infections can damage mucosa
 - a. Lactose malabsorption—most common cause of malabsorption in children, results in gas, pain, and diarrhea, but growth normal; can be temporary following gastrointestinal infection; lactose intolerance less common < age 4, but occurs in 80% Asian, African-American adults; primary lactase deficiency very rare

- b. Infectious—bacterial, viral, parasites, e.g., *Giardia lamblia*
 - c. Celiac disease—immune-mediated systemic disease due to intestinal intolerance to gluten; rate in population 1:1000; more common in children with Irish, Swedish heritage, but increasingly diagnosed in all ethnic groups; more common in children with type 1 diabetes, Down syndrome
 - d. Crohn's disease
 - e. Food allergy
3. Decreased conjugated bile acids
 - a. Biliary atresia
 - b. Hepatitis
 - c. Short bowel syndrome

- Signs and Symptoms

1. Failure to thrive
2. Adequate or increased intake per dietary history
3. Severe, chronic diarrhea
4. Bulky, foul, pale, steatorrhea stools
5. Abdominal distention

- Differential Diagnosis

1. Renal disease
2. Poor dietary intake
3. Failure to thrive

- Physical Findings: Disease specific findings

1. Lactose intolerance—persistent diarrhea after infectious diarrhea with normal growth
2. Cystic fibrosis—recurrent pulmonary infection, “salty” taste to skin; nasal polyps; rectal prolapse, failure to thrive
3. Celiac disease—vomiting, abdominal pain, irritability, anorexia, pallor; protuberant abdomen, failure to thrive often noted around 6 months of age with the introduction of solids

- Diagnostic Tests/Findings

1. Stool—(most important) inspection, culture, microscopic examination
 - a. Hemoccult test—intestinal mucosa damage
 - b. Ova and parasite; *Giardia* antigen to test for *Giardia* and other parasites
 - c. pH reducing substances—to rule out carbohydrate malabsorption
 - d. Sudan stain for fat (microscopic examination of stool for fat)
 - e. 3-day quantitative fecal fat (quantitates amount of fat losses in stool)
2. Urinalysis/culture
3. CBC, comprehensive metabolic panel, ESR
4. Celiac screen—serum IgA (have to screen for IgA deficiency to ensure accuracy of test);

tissue transglutaminase (most sensitive and specific test)

5. Sweat test > 60 mEq/L chloride—cystic fibrosis
 6. Hydrogen breath test—increased with lactose intolerance
- Management/Treatment:
 1. Refer to gastroenterologist
 2. Lactose intolerance—avoid lactose containing foods
 3. Celiac disease—gluten free diet; no wheat, oats, rye, barley
 4. Cystic fibrosis—pancreatic enzyme replacement

■ ACUTE ABDOMINAL PAIN

Intussusception

- Definition: Acute episode of prolapse of one portion of intestine into the lumen of the adjoining part, usually ileocolic
- Etiology/Incidence
 1. Unknown cause—85% idiopathic; may be caused by polyps, adenovirus, or rotovirus with Peyer patch hypertrophy, Henoch Schönlein purpura, celiac disease, CF; Meckel diverticulum of small intestine; lymphoma primary cause in children > 6 years; intermittent intussusception is a rare cause of abdominal pain
 2. Greater incidence in males than females
 3. 60% occur before first birthday; 80% by 2 years
- Signs and Symptoms
 1. Healthy infant/child presents with sudden cycle of inconsolable screaming, flexing of leg, colicky abdominal pain
 2. 90% have nonbilious vomiting after pain
 3. Periods of quietness or sleepiness between episodes; lethargy if intussusception not reduced
 4. Eventually shock-like state develops
 5. Within 12 hours of onset, “currant jelly” (blood with mucus) stool is passed; late presentation
- Differential Diagnosis
 1. Gastroenteritis
 2. Incarcerated hernia
 3. Volvulus/obstruction
- Physical Findings
 1. Abdomen soft between episodes; may palpate sausage shaped mass RUQ or upper-mid abdomen
 2. Distention and tenderness increased as obstruction increases
- 3. Guaiac positive or grossly bloody stool
- 4. If not reduced, develops perforation and peritonitis leading to fever and shock
- Diagnostic Tests/Findings
 1. Radiography only to clarify diagnosis—no gas RLQ, air fluid levels consistent with obstruction
 2. Barium enema—diagnostic and results in reduction
 3. CBC and electrolytes—dehydration and anemia
 4. Ultrasound—tubular mass longitudinal image, doughnut on transverse view
- Management/Treatment
 1. Reduction via barium/air enema; air becoming more common
 2. Emergency surgery
 3. Can recur; fatal if untreated

Appendicitis

- Definition: Acute inflammation of the appendix
- Etiology/Incidence
 1. Cause—obstruction of lumen by fecaliths or parasites
 2. Incidence increases with age, peaks between 15 and 30 years of age
 3. Incidence in males greater than females
 4. Seasonal distribution—autumn and spring
 5. Increased incidence following gastrointestinal infection
 6. Most common cause of pediatric abdominal surgery
- Signs and Symptoms
 1. Young child may not appear ill or have severe pain, particularly in early phase
 2. Abdominal pain—earliest symptom
 - a. Vague, possibly midline, constant pain for several hours
 - b. Pain eventually localized in RLQ; in some, pain may begin in RLQ
 - c. Can wake at night over time with increasing severity of pain
 - d. Pain on ambulation
 - e. Pain precedes vomiting
 3. Anorexia, nausea, and vomiting
 4. Variable changes in bowel patterns—constipation or diarrhea may be noted
 5. Afebrile to very low-grade fever in early phase
- Differential Diagnosis
 1. Gastroenteritis—especially if fever (early stage) and/or diarrhea

2. Mittelschmerz
 3. Ovary cyst/ovary torsion/ruptured follicle
 4. Lower lobe pneumonia
 5. Pelvic inflammatory disease (PID)
 6. Constipation
 7. UTI/pyelonephritis
 8. Ruptured ectopic pregnancy
 9. Inflammatory bowel disease
 10. Intussusception
 11. Perforated peptic ulcer
- Physical Findings: Depends upon the stage of appendicitis
 1. Observe child—may be motionless, with legs flexed
 2. Tenderness localized to RLQ; intense at McBurney's point, halfway between umbilicus and anterior superior iliac crest
 3. Rebound tenderness
 4. Rovsing's sign—pain RLQ with left-side pressure; highly indicative of appendicitis in children
 5. Local, right-sided tenderness or mass on rectal examination
 6. Won't jump/difficulty ambulating
 7. Obturator sign—rotating thigh may produce pain in RLQ
 8. Complete examination to rule out other causes of abdominal pain, especially throat, chest, testicles
 9. Perforation and peritonitis within 24 to 48 hours
 - a. Rigidity
 - b. Higher fever
 - c. Pain improves
 - d. Generalized tenderness
 - e. Increased vomiting
 - f. 40% incidence in young children
 - Diagnostic Tests/Findings
 1. Diagnosis based on history and physical; tests often not helpful in diagnosing appendicitis, but can rule out other sources of pain
 2. CBC with differential—may have a mild leukocytosis with left shift
 3. CT—most sensitive and specific; useful, especially in early stages of diagnosis; better to identify other diagnoses; US versus CT often depends upon institution; some will do US because it is quicker, and if equivocal proceed to CT scan
 4. Ultrasound—if ovarian condition part of differential or findings unclear
 5. Radiography of abdomen to rule out constipation
 6. UA to rule out UTI—UA can be abnormal in appendicitis

- Management/Treatment: Immediate surgical referral

■ FUNCTIONAL GASTROINTESTINAL DISORDERS

Functional GI disorders include functional abdominal pain, functional dyspepsia, abdominal migraine, and irritable bowel syndrome. A thorough history and physical examination will often be sufficient to lead to the diagnosis of most common disorders.

Once any underlying organic disease is eliminated it is the primary care provider's role to reassure the child and family and to encourage participation in normal activities including school attendance. Workup is similar for each functional disorder.

Functional Abdominal Pain (also called chronic abdominal pain, recurrent abdominal pain)

- Definition: Episodic or continuous abdominal pain that occurs at least weekly for at least 2 months where there is no evidence of an inflammatory, anatomic, metabolic, or oncological process to explain the child's symptoms
- Etiology/Incidence
 1. Unclear mechanism of pain; multifactorial, altered brain-gut interaction
 2. Multifaceted problem that includes predisposition aggravated by early life events or personality
 3. Most common cause of chronic pain in school-aged and young adolescent; incidence unknown, accounts for 2 to 4% of pediatric office visits; 13 to 17% of all adolescents have had chronic abdominal pain
 4. Most common between age 8 to 15 years, uncommon under 4 years of age
 5. Greater incidence in girls than boys; average age for females 9 to 10 years, males 10 to 11 years
 6. Family history of GI complaints and somatization disorders, e.g., migraines, peptic ulcers
 7. Differential diagnosis
 - a. Organic—approximately 10%
 - (1) Peptic ulcer disease
 - (2) Helicobacter pylori
 - (3) GER
 - (4) Eosinophilic gastroenteritis
 - (5) Pancreatitis—more common if positive family history; corticosteroid use
 - (6) Cholecystitis—rare < 9 years of age; increased with obesity; positive family history of gall stones; birth control pills

- (7) Inflammatory bowel disease
 - (8) Celiac disease
 - (9) Constipation
 - (10) Malabsorption/lactose intolerance
 - (11) Parasites (especially Giardia)
 - (12) Gynecological disorders
 - (13) Urinary tract infection
 - (14) Sexual abuse
- Signs and Symptoms
 1. Certain personality traits (maladaptive coping skills, anxiety, internalization of feelings) and family characteristics (protective parents) more frequent; often adversely affects school performance as child frequently absent due to pain (red flag) and child's quality of life
 2. Nature of pain
 - a. Onset of crampy or dull ache; no radiation of pain
 - b. Pain usually periumbilical
 - c. Nothing relieves pain
 - d. Interferes with activities, but no night waking
 - e. Unrelated to meals
- Physical Findings: Indicate nonorganic cause
 1. Normal weight
 2. Afebrile
 3. Abdomen may have diffuse tenderness, but no guarding
 4. Normal findings on complete examination
- Diagnostic Tests/Findings: Diagnosis of exclusion
 1. Excellent history and physical examination key to diagnosis; special attention to growth parameters
 2. Exacerbating factors (foods, stress, time of day) and relieving (acid suppression, diet/food avoidance) factors
 3. Family history—peptic ulcer disease, IBD
 4. Guaiac stool—negative; rule out inflammatory bowel disease
 5. Blood tests—CBC with differential; ESR to rule out infection/inflammation
 6. Urinalysis/culture—rule out UTI
 7. Ova and parasites; Giardia antigen
 8. Additional/selected studies may be warranted depending on symptoms
 - a. Pelvic examination of adolescent female
 - b. Endoscopy for esophagitis, peptic ulcer—dysphagia, chest pain, weight loss present
 - c. Upper gastrointestinal (UGI) with small bowel—if recurrent vomiting, stool blood to rule out obstructive lesions, Crohn's disease
 - d. Hydrogen breath test—increased with lactose intolerance
 - e. Pregnancy test

- f. Endoscopy or serology—rule out *Helicobacter pylori*
- g. Abdominal ultrasound—rule out cholecystitis
- h. Amylase/lipase—increased with pancreatitis

- Management/Treatment
 1. Emphasize to child and family—pain is real, even though no organic cause can be found; workup should reassure them child is healthy
 2. Reinforce normal behavior; go to school, don't allow secondary gain; may go to school nurse briefly if symptoms severe; if too sick to attend school, follow-up with provider required
 3. Decrease hectic lifestyle and hurried meals
 4. Limited evidence-based knowledge on the use of medications and functional abdominal pain
 5. Try to identify source of stress in patient and/or family; stress reduction techniques may help, such as biofeedback and relaxation techniques; may need referral to behavior psychologist
 6. Keep pain diary to identify situations associated with symptoms
 7. Treat identified organic disease

Functional Dyspepsia

- Definition: Persistent or recurrent pain in the upper abdomen above the umbilicus. No evidence, including upper endoscopy, of organic disease. No evidence that dyspepsia is relieved by defecation. Acid suppression can be useful in relief of symptoms.

Abdominal Migraine

- Definition: Three or more paroxysmal episodes of acute, midline, abdominal pain lasting for 2 hours to several days with intervening symptom-free episodes. No evidence of organic disease, including CNS. And at least 2 of the following: headache during episodes, photophobia during episodes, family history of migraines, headache confined to one side only and aura. Pizotifen (serotonergic agent) has been found to improve symptom.

Irritable Bowel Syndrome

- Definition: Chronic disorder with a range of symptoms, including abdominal pain, altered bowel habits (diarrhea or constipation), bloating, fecal urgency, and a feeling of incomplete evacuation
- Treatment
 1. Diarrhea predominant—consider fructose and lactose elimination from diet for 2 to 3 weeks;

antidiarrheal agents and antispasmodic agents may be helpful

2. Constipation predominant—increased dietary soluble fiber (5 + age in years = daily dose in grams); stool softener such as polyethylene glycol 3350

■ CONSTIPATION

- Definition: Alteration in frequency, passage, size, or consistency of stool
- Etiology/Incidence
 1. Functional—less than 5% of pediatric constipation has an organic cause; constipation most commonly due to voluntary withholding of stool following the passage of a painful bowel movement; painful stools can be due to many factors, including change in diet, toilet training, stressful events, intercurrent illness, delayed defecation due to play or lack of available toilets; stoolholding leads to increased stool mass, stools become hard as fluids are reabsorbed into the colon; a cycle of stoolholding and painful stools occurs, resulting in constipation, which if untreated leads to impaction and encopresis
 2. Encopresis (involuntary fecal soiling)—chronic withholding leads to impaction and soiling; eventually loses urge to stool and results in megacolon; psychological problem not primary cause
 3. Anatomical abnormalities—anal or rectal abnormalities; problems often seen in immediate newborn period
 4. Intrinsic motor disorder—Hirschsprung's disease (most common), congenital absence of ganglion in segments of colon; 1:5000 births; 4:1 male to female ratio; increased with positive family history or Down syndrome
 5. Metabolic, e.g., hypothyroidism
 6. Neurologic, e.g., tumors, spinal injury, myelomeningocele
- Signs and Symptoms
 1. Onset
 - a. Functional—during infancy, particularly after change from breastmilk to formula or starting solids; 1 to 3 years of age after life change, e.g., new sibling, home, introduction of toilet training
 - b. Encopresis—4 to 7 years, male > female
 - c. Hirschsprung's disease—Less than 10% of infants with Hirschsprung's disease will pass meconium in first 24 hours; history of failure to have bowel movement without aid of laxative or enema; short-segment

Hirschsprung's may manifest itself beyond the newborn period

2. Stools
 - a. Functional—hard, dry stools, “pellets;” occasionally stool caliber very large/wide; may be dark or have strong odor
 - b. Encopresis—soiled underwear, may appear to be diarrhea; may occur daily
 - c. Hirschsprung's—small, ribbon-like stools; no leakage
3. Complaints
 - a. Functional—abdominal pain, blood streaked stools, straining or “dancing around” indicates withholding
 - b. Hirschsprung's—no stooling
- Differential Diagnosis
 1. Tumor, sacral teratoma
 2. Anatomical defect
 3. Metabolic and gastrointestinal disorders—celiac disease, cystic fibrosis
 4. Infantile botulism—(recent onset)
 5. Tethered spinal cord and other neuropathic disorders
 6. Drugs
 7. Heavy metal ingestion (lead)
 8. Cow's milk protein intolerance
- Physical Findings
 1. Functional—rectal examination may show fissure, ampulla full of stool, normal tone; may have no palpable abdominal mass; may have abdominal pain or cramping, but no distention; normal growth and development
 2. Encopresis—may have impacted stool and/or large, dilated rectal vault, normal tone; abdominal distention with sausage-shaped mass in left pelvis or midline
 3. Hirschsprung's—tight, empty rectum in presence of palpable abdominal stool mass, may be explosion of stool on withdrawal of examining finger; stool may be guaiac positive; abnormal bowel sounds; abdominal distention; failure to thrive
 4. Anal wink, neurological examination, muscle strength and tone should be normal
- Diagnostic Tests/Findings
 1. Radiograph of abdomen to examine for stool
 2. Unprepped barium enema—rule out Hirschsprung's disease
- Management/Treatment:
 1. Emphasize to parents the definition of constipation; straining with soft stool in infancy is normal
 2. Ensure proper preparation of formula

3. Infants > 6 months of age—prune juice; malt soup extract $\frac{1}{2}$ to 3 teaspoons twice per day for maximum of 3 days
4. Constipation causing abdominal pain or encopresis needs more aggressive treatment; withheld stool causes intestinal muscle stretching; multifaceted treatment involves emptying intestines, leading to a return of sensation, preventing recurrence of painful stools and bowel training
5. Plan for otherwise healthy child
 - a. If impacted—day 1 mineral enema to soften stool
 - b. No impaction or day 2—sodium phosphate enema one time per day for 2 to 3 days
 - c. May choose oral medications for disimpaction, polyethylene glycol 3350, 1 to 1.5 g/kg/day for 3 days
 - d. After intestines emptied, keep stool soft to prevent recurrence of withholding cycle—polyethylene glycol 3350, 1 g/kg/day
 - e. Prevent pain cycle—emphasize to child that medicine will prevent painful stools
 - f. Bowel retraining—child should sit on toilet for one minute per year of age twice per day; don't expect bowel movement every sitting
 - g. Goal—soft bowel movement every day or every other day without encopresis
6. Hirschsprung's—GI/surgery referral

■ HEPATITIS

- Definition: Inflammation of the liver
- Etiology/Incidence
 1. Hepatitis A virus (HAV)
 - a. Most common form of viral hepatitis in children
 - b. Highest rates of symptomatic infection 5 to 14 years; less than 10% of infected children under 6 years are symptomatic
 - c. Transmission—fecal/oral, raw shellfish, contaminated water; in U.S., HAV shed from asymptomatic, infected children to adults
 - d. Incubation—15 to 50 days, average 25 to 30 days; can infect others up to 2 weeks before onset of illness and 1 week after
 - e. High risk populations—Native-Americans, Alaskan natives, homosexuals, IV drug users; some Hasidic, Hispanic communities; travelers to developing countries; children in daycare
 2. Hepatitis B virus (HBV)
 - a. Most common form of hepatitis in world

- b. Transmission—blood or body fluids; virus can survive more than one week on inanimate objects
 - c. Incubation—45 to 160 days, average 120 days
 - d. 5% to 10% of infected people become chronic carriers; inverse relationship between age of infection and carrier state; majority of perinatally infected infants become carriers
 - e. High risk populations—sexually active; institutionalized; IV drug users; patients with clotting disorders; household contacts of HBV carriers; hemodialysis patients; infants of Alaskan natives or Pacific Islanders; travelers to China, Southeast Asia
3. Hepatitis C virus (HCV)
 - a. Very low rates in children under 12 years of age
 - b. Transmission—blood and blood products, occasionally blood transfusion; maternal-fetal transmission 3 to 5%
 - c. Incubation—6 to 7 weeks, range of 2 weeks to 6 months
 4. Hepatitis D (HDV)
 - a. Coinfection or super infection with HBV
 - b. Transmission—blood or blood products, IV drug use, sexual contact
 - c. Incubation—2 to 8 weeks, if simultaneous with HBV, average is 120 days
 5. Hepatitis E (HEV)
 - a. Endemic to Asia, Africa, Mexico; U.S. travelers to those areas at risk
 - b. Transmission—fecal/oral route, especially contaminated water
 - c. Incubation—40 days, range 15 to 60 days
 - d. More common in adults than children
- Signs and Symptoms
 1. HAV
 - a. Infants and young children—asymptomatic or nonspecific symptoms, e.g., nausea, vomiting, diarrhea; no jaundice; misdiagnosed as gastroenteritis
 - b. Adults—fever, malaise, anorexia, nausea, abdominal pain, jaundice; later pruritus
 - c. Self-limiting disease—several weeks to occasionally 6 months; no chronic or carrier state
 2. HBV and HDV
 - a. Children often asymptomatic—mild to severe disease; macular rash and arthritis (early sign); anorexia, nausea, malaise, arthralgia
 - b. Up to 90% of perinatally infected infants can develop carrier state

- c. Chronic carrier state can lead to chronic liver disease or cancer
 - 3. HCV
 - a. Children often asymptomatic; those with symptoms have mild disease; < 25% become icteric; gradual onset of headache, fever, nausea, fatigue, anorexia
 - b. 50% to 85% become chronic carriers; can lead to chronic liver disease or cancer
 - 4. HEV
 - a. Acute illness with arthralgia, abdominal pain, jaundice, malaise, anorexia, and fever
 - b. No carrier state
 - Differential Diagnosis
 1. Viral gastroenteritis
 2. Hemolytic-uremic syndrome
 3. Reye's syndrome
 4. Cytomegalovirus illness
 5. Toxin/medication exposure
 6. Fitz-Hugh Curtis syndrome with gonorrheal PID
 - Physical Findings
 1. Possible hepatosplenomegaly; RUQ tenderness
 2. If jaundiced, may develop dark urine and light stools, scleral icterus
 - Diagnostic Tests/Findings
 1. Nonspecific findings
 - a. Elevated liver enzymes—AST, ALT
 - b. Elevated serum bilirubin
 - c. Elevated erythrocyte sedimentation rate (HAV)
 2. Specific serologic antigen/antibody testing
 - a. Hepatitis A (HAV)
 - (1) Anti-HAV IgM—current or recent infection; usually disappears within 4 months
 - (2) Anti-HAV IgG—resolved infection and immune status; may last for years following infection; 40% to 45% general population carry anti-HAV IgG
 - b. Hepatitis B (HBV)
 - (1) HBsAg (surface antigen)—earliest marker of acute infection; persistence beyond 6 months indicates carrier status
 - (2) Anti-HBc IgM—current or recent acute infection; usually disappears within 6 months
 - (3) Anti-HBc IgG—chronic infection of at least 6 months duration
 - (4) Anti-HBs—immune status following resolved infection or immunization (Heptavax)
 - (5) Anti-HBe—recovery phase of infection
 - c. Hepatitis C (HCV)
 - (1) Anti-HCV (ELIZA)—may need serial testing due to false negatives up to 5 or more weeks following onset of symptoms
 - (2) Anti-HCV (RIBA)—positive after several weeks; confirms infection sooner than Anti-HCV
 - d. Hepatitis D (HDV)—anti-HDV from reference laboratory
 - e. Hepatitis E (HEV)—serologic and polymerase chain reaction assays are available experimentally at this time; diagnosis by exclusion of acute hepatitis A, B, C, D and other causes of viral hepatitis
- Management/Treatment
 1. Treatment is supportive, good nutrition, decreased activity, monitor hydration and chronic state
 2. HAV—immunoglobulin (IG) available for decreasing course of disease in early stages or prevention in exposed individuals; not recommended for HCV or HEV; defer measles or MMR immunization for 3 months following administration of immune globulin
 3. Treatment for HBV
 - a. Interferon α -2B treatment for chronic HBV; limited improvement (30% to 40% HBV; 10% to 20% HCV)
 - b. Expensive
 - c. Side effects
 - d. Lamivudine 3 mg/kg/day may be used if interferon fails
 - e. Adefovir 10 mg/day can be used in children > 12 years of age
 4. Treatment of HCV Interferon α -2B, three times per week, plus oral ribavirin Pegylated interferon weekly, plus oral ribavirin, for children > 3 years of age
 5. Report to state health department
 6. Prevention
 - a. HAV—two inactive HAV vaccines recommended for children > age 12 months (see Health Maintenance and Health Promotion, Chapter 3); hepatitis A vaccine should be given to previously unvaccinated individuals prior to travel to affected areas; HAV vaccine can also be given as postexposure prophylaxis; for children aged < 12 months, immunocompromised persons, persons with chronic liver

- disease, and persons who are allergic to the vaccine or a vaccine component HAV IG should be used
- b. HBV—vaccine available and highly recommended for all newborns and adolescents (See Health Maintenance and Health Promotion, Chapter 3)
- c. Cautious intake of food and water when traveling to endemic area
- d. Avoid unprotected sex and drug use

▣ HERNIA

- Definition: Abnormal protrusion of abdominal tissue/structures through umbilical ring in umbilical hernia or external inguinal ring in inguinal hernias
- Etiology/Incidence
 1. Umbilical—due to imperfect closure or weakness of umbilical ring; common in infancy, reported in up to 60% of African-American infants; many umbilical hernias resolve by age 2, but in the African-American population closure can occur as late as 11 years of age; generally, if not closed by 4 to 5 years of age, the patient should be referred to a surgeon
 2. Inguinal—failed closure of processus vaginalis
 - a. Congenital defect—can be noticeable at birth
 - b. Nine times more frequent in males
 - c. Greater risk with premature births
 - d. Hydrocele can increase risk; indicates opening present
- Signs and Symptoms
 1. Intermittent or constant bulge of abdominal wall or inguinal region that may worsen with crying or straining
 2. Uncomplicated hernias—asymptomatic
 3. Umbilical—incarceration or strangulation extremely rare
 4. Inguinal
 - a. Incarcerated—cranky, anorexia, nausea, vomiting, groin discomfort, constipation
 - b. Strangulated—area becomes tender, swollen, and progressively reddened in addition to above symptoms, possible fever
- Differential Diagnosis
 1. Hydrocele
 2. Lymphadenopathy
 3. Undescended testes
- Physical Findings
 1. Umbilical hernia—size of defect varies from 1 to 5 cm in diameter

2. Inguinal hernia
 - a. Maneuvers that increase intra-abdominal pressure (sitting up, crying, coughing) will increase visibility of hernia
 - b. May be bilateral; if unilateral, right side more common
 - c. “Silk” sign can be diagnostic; elicited by palpation of the spermatic cord over the pubic tubercle, the layers of the peritoneum rubbing together will have a “silky feel”
 - d. Transillumination of scrotal sac will highlight the presence of bowel

- Diagnostic Tests/Findings: None may be needed; ultrasound if unclear
- Management/Treatment:
 1. Monitor umbilical hernias; reassure parents
 2. Refer inguinal for surgical correction
 3. Emergency referral if incarcerated/strangulated

▣ QUESTIONS

Select the best answer

1. A ten-month-old child has been diagnosed with gastroenteritis. He attends a child-care facility. What is the most likely cause of his illness?
 - a. Clostridium difficile
 - b. Rotavirus
 - c. Salmonella
 - d. Cryptosporidium
2. In a healthy, eight-month-old with diarrhea but no dehydration, what would be the most appropriate advice to give parents?
 - a. Encourage $\frac{1}{2}$ strength formula for 12 hours
 - b. Give oral rehydration solution (ORT) for 12 hours
 - c. Give only fluids until stools return to normal
 - d. Give bananas and cereal as tolerated
3. When evaluating a child with abdominal pain, what symptom would lead to a likely organic etiology?
 - a. Night waking
 - b. Pallor
 - c. Suprapubic pain
 - d. Sweating

4. Vomiting in infancy has a long list of differential diagnoses. Which accompanying symptom would most likely point to pyloric stenosis?
 - a. Diarrhea
 - b. Appropriate growth
 - c. Acts hungry after vomiting
 - d. Sausage shaped mass in abdomen
5. Which of the following is the appropriate regimen for pinworm medication?
 - a. Daily times 7 days, repeat as needed
 - b. Three times a day for 10 days, repeat as needed
 - c. Twice daily for 3 days, repeat in 2 weeks
 - d. 1 dose/1 time, repeat in 2 weeks
6. Mrs. Doyle is upset. Two-month-old John's frequent vomiting has her convinced that "something is seriously wrong." Which of the following is most suggestive of GER (gastroesophageal reflux)?
 - a. He's gained 5 ounces this month
 - b. He has a slight wheeze today
 - c. He eats hungrily after vomiting
 - d. He drinks 7 to 8 ounces every 3 to 4 hours
7. You see Jack, a 20-month-old toddler with normal growth and development in your office for diarrhea. His mother tells you that he is passing up to three loose stools a day and that he drinks 20 ounces of apple juice a day. What is the most likely diagnosis?
 - a. Crohn's disease
 - b. Giardia lamblia
 - c. Celiac disease
 - d. Nonspecific "toddler" diarrhea
8. Baby Sally was in your office last week for her 6-month check up. Her weight was 7 kg. Today she presents with diarrhea and vomiting for four days. Today her weight is 6.5 kg. What is her percentage of dehydration?
 - a. 5%
 - b. 7%
 - c. 10%
 - d. < 1%
9. What clinical signs would you expect to see in Sally on your examination?
 - a. Normal capillary refill
 - b. Normal fontanel
 - c. Cool mottled skin
 - d. Dry mucous membranes
10. Sally's vomiting and diarrhea has stopped. If she needs oral replacement therapy (ORT) today, what would be the appropriate amount to recommend?
 - a. 325–350 cc over 4 hours
 - b. 600–700 cc over 4 hours
 - c. 600–700 cc over 12 hours
 - d. 325–350 cc over 8 hours
11. Pinworms can cause which of the following?
 - a. Constipation
 - b. Anal itching
 - c. Abdominal pain
 - d. Diarrhea
12. In evaluating Billy, a child with bloody diarrhea, which of the following would not be an appropriate first action?
 - a. Check growth chart
 - b. Stool culture
 - c. Upper GI
 - d. Hemoccult test stools
13. Billy's family eats at fast food restaurants 4 to 5 times each week. If you suspect the diarrhea is infectious in nature, what is a likely causative organism?
 - a. Adenovirus
 - b. E. coli
 - c. Giardia lamblia
 - d. S. aureus
14. Which of the following conditions would be most likely to occur in a four-year-old boy?
 - a. Pyloric stenosis
 - b. Recurrent abdominal pain
 - c. Intussusception
 - d. Giardia infection
15. Which of the following findings could be expected to occur in a baby with intussusception?
 - a. Inconsolable screaming
 - b. Olive shaped mass
 - c. Left to right peristaltic waves
 - d. Weight loss
16. Which of the following may occur with suspected appendicitis?
 - a. Pain not relieved with ambulation
 - b. Young children appear very ill in the early phase
 - c. Fever is 102 to 103°F
 - d. Leukopenia with left shift

17. In the U.S., parasitic gastroenteritis is most commonly caused by which organism?
 - a. *Enterobius vermicularis*
 - b. *Entamoeba histolytica*
 - c. *Cryptosporidium parvum*
 - d. *Giardia lamblia*
18. Which of the following serological findings indicates a carrier state for HBV?
 - a. HBs Ag negative for 6 months
 - b. IgM anti-HBc negative and HBsAg positive
 - c. Anti-HBc positive
 - d. Anti-HBs positive
19. Children in child-care facilities are at greater risk of being exposed to which of the following infections?
 - a. HAV
 - b. HBV
 - c. HCV
 - d. HDV
20. Infant immunization for hepatitis B often raises many parental questions about the disease. Which of the following is not true about hepatitis B virus?
 - a. It can survive for more than 1 week on fomites
 - b. It is the most common form of hepatitis in the world
 - c. Contaminated water and shellfish are the major source
 - d. Perinatally infected infants are likely to become carriers
21. Two-day-old baby Jamie is in the hospital nursery and still has not passed meconium. This is a red flag for what condition?
 - a. Intussusception
 - b. Hemolytic uremic syndrome
 - c. Pyloric stenosis
 - d. Hirschsprung's disease
22. Consistent with the condition in question 21, Jamie's findings on rectal examination would be which of the following?
 - a. Tight anal canal with no stool in vault
 - b. Impacted stool with fissure
 - c. Large, dilated rectum
 - d. Soft stool, normal tone
23. What treatment would be appropriate for Jamie's condition?
 - a. Emulsified mineral oil, $\frac{1}{2}$ tablespoon per day
 - b. Referral to gastroenterologist/surgeon
 - c. Malt soup extract, 2 teaspoons for 3 days
 - d. Rectal dilatation with thermometer
24. When evaluating a child with suspected IBD, which of the following diagnostic tests would not be helpful?
 - a. Amylase and lipase
 - b. ESR
 - c. Serum total protein and albumin
 - d. CBC with differential
25. Your patient has inflammatory bowel disease. Which finding is most consistent with ulcerative colitis?
 - a. Occult blood
 - b. Perirectal abscess
 - c. Aphthous ulcers
 - d. Left sided abdominal pain
26. Antimicrobials will improve the condition of a four-year-old child with diarrhea caused by which of the following organisms?
 - a. *Salmonella*
 - b. Rotavirus
 - c. *Shigella*
 - d. *E. coli* (O157:H7)
27. Katie has functional abdominal pain. When counseling her family on management of painful episodes, you would recommend which of the following?
 - a. Take ibuprofen 200 mg for pain
 - b. Stay home from school during episode
 - c. Decrease milk products
 - d. Go to school during episode
28. Which of the following would not be consistent with a diagnosis of functional constipation in an infant?
 - a. Vomiting
 - b. Anal fissure
 - c. Straining
 - d. Starting solids
29. A child has developed her second perirectal abscess in six months. She should be evaluated for which condition?
 - a. *Giardia lamblia*
 - b. Crohn's disease
 - c. Ulcerative colitis
 - d. Enterobiasis
30. Which of the following symptoms are most common in the early phase of appendicitis in children?
 - a. Abdominal pain after eating
 - b. Fever and diarrhea
 - c. Severe localized RLQ pain with pallor and sweating
 - d. Anorexia, vague, diffuse pain

31. Steatorrhea is not consistent with which of the following?
- C. difficile*
 - Giardia lamblia*
 - Celiac disease
 - Cystic fibrosis
32. Jamil has had diarrhea for three days. His mother calls concerned. Which of the following would not be helpful advice?
- Monitor stool for blood or mucus
 - Encourage solid food
 - Avoid milk products
 - Monitor for urination at least every 6 hours
33. Of the following advice, which would be most helpful for the parents of a baby with gastroesophageal reflux?
- Most babies continue to vomit until they are walking; at around one year of age
 - Laying prone after eating will decrease the amount of vomiting
 - Increase the interval between feedings to a minimum of four hours
 - Medications are generally necessary to prevent further problems
34. Which of the following foods would be appropriate for a child with celiac disease?
- Oatmeal for breakfast
 - Boiled rice with butter
 - Commercially baked bread
 - Cream of wheat
35. A parent requests that her 6-month-old child receive immunoglobulin (IG) as protection against hepatitis A prior to international travel. Which of the following does this parent need to know?
- After IG administration, a 3-month interval is needed prior to the next measles vaccine
 - There is no impact on future immunizations
 - No immunizations can be given for 1 year
 - Since children do not have symptoms with hepatitis A, IG is not necessary

ANSWERS

- | | |
|--------------|--------------|
| 1. b | 19. a |
| 2. d | 20. c |
| 3. a | 21. d |
| 4. c | 22. a |
| 5. d | 23. b |
| 6. d | 24. a |
| 7. d | 25. d |
| 8. b | 26. c |
| 9. d | 27. d |
| 10. b | 28. a |
| 11. b | 29. b |
| 12. c | 30. d |
| 13. b | 31. a |
| 14. d | 32. c |
| 15. a | 33. b |
| 16. a | 34. b |
| 17. d | 35. a |
| 18. b | |

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Infectious Diseases

Jody K. Roblyer

■ SEPTICEMIA (SEPSIS)

- Definition: A generalized systemic response to infection; usually severe and associated with presence of pathogenic microorganisms or associated toxins (usually bacterial) in bloodstream; neonates are most susceptible due to immature immune system
- Etiology/Incidence
 1. Etiologic agents vary by age, immunologic status, and mechanism of transmission
 - a. Newborn (< 3 days)—placental transfer of pathogens from infected maternal blood stream or from vaginal mucosa during birth
 - (1) Group B streptococcus (GBS)—high mortality
 - (2) Escherichia coli—most common gram negative organism
 - (3) Listeria monocytogenes
 - b. Neonates < 28 days—cross-contamination in nurseries or other crowded conditions; poor hand washing and housekeeping
 - (1) Staphylococcus aureus—most common
 - (2) Klebsiella
 - (3) Enterococci
 - (4) Pseudomonas
 - c. Older infants in the community can be at increased risk for sepsis due to inadequate immunization status—Haemophilus influenzae type B
 - d. Hospitalized immunosuppressed children at risk for nosocomial sepsis—cancer, postoperative, and patients with HIV/AIDS
 2. Leading cause of morbidity and mortality among hospitalized patients due to associated hemodynamic changes affecting tissue perfusion and oxygenation; if not treated, may lead to septic shock
 3. Increased risk of septicemia
 - a. High risk, premature infants
 - b. Males > females
 - c. Invasive procedures—IV, intubation
 - d. Bottle feeding—breastmilk may be protective
 - e. Immunocompromised children and adolescents
 - f. Age-related risk groups
 - (1) Neonates < 28 days with fever without source (FWS) > 38°C
 - (2) Infants 28 to 90 days with FWS > 38°C
 - (3) Infants > 90 days with FWS > 39°C who have received HIB and PCV-7
- Signs and Symptoms
 1. Neonates—symptoms may be vague and nonspecific, e.g., poor feeding, color changes (pallor, mottling), changes in muscle tone, apnea, cyanosis, low temperature, or fever
 2. Other immunosuppressed children—fever may be only sign; reports by caregiver that child “isn’t him or herself”

3. Older children (rarer) with normal immune function—fever, irritability, anorexia, general toxic appearance
- Differential Diagnosis
 1. Viral sepsis
 2. Fungal infection
 3. Rocky mountain spotted fever
 4. Toxic shock syndrome
 5. Noninfectious causes—hemorrhagic, anaphylactic, or neurogenic shock
 6. UTI
 7. Pneumonia
 8. Early bacterial meningitis
 - Physical Findings
 1. Neonates—hyper or hypothermia; bradycardia or tachycardia; hepatosplenomegaly, jaundice
 2. Older children—irritability, stiff neck
 3. Later phase—lethargy, delayed capillary refill, hypotension, and subsequent septic shock
 4. 20% of infants and young children have fever without an apparent source after a complete history and physical examination
 - Diagnostic Tests/Findings
 1. CBC with differential—WBC $> 15,000$ with increased neutrophils
 2. Chest radiograph to rule out pneumonia
 3. Positive blood cultures identifying source of infection
 4. Urinalysis, urine culture, and sensitivity to rule out UTI, especially in infants with FWS
 5. Cerebral spinal fluid (CSF)—pleocytosis, decreased glucose, increased protein, positive culture
 6. Coagulation studies—prolonged prothrombin time; decreased fibrinogen levels
 7. Low risk infants age 28 to 90 days will present with the following—previously healthy full term infant, uncomplicated nursery stay, clinically nontoxic looking, no focal bacterial infection on exam (except otitis media), urine free of leukocyte esterase and nitrites or < 10 WBCs/hpf, WBC 5 to 1500 and < 1500 bands or band/neutrophil ratio < 0.2 , no blood and < 5 WBCs/hpf when diarrhea present, CSF < 8 WBCs/mm³ and negative gram stain, chest x-ray clear without infiltrates
 - Management/Treatment
 1. Medical referral for hospitalization
 2. Close observation, monitoring, and supportive care
 3. IV fluids to maintain hemodynamics
4. Broad spectrum parenteral antibiotics for gram positive and negative organisms pending culture and sensitivities
 5. Vasoactive medication if septic shock ensues
 6. Decision making regarding age and relative risk
 - a. Neonates < 28 days with FWS $> 38^{\circ}\text{C}$, complete sepsis workup with CBC with differential, blood cultures, U/A and urine culture, CSF studies, and chest x-ray; requires hospitalization with IV antibiotics, pending culture results
 - b. Infants 28 to 90 days with FWS $> 38^{\circ}\text{C}$ can be managed as outpatient without antibiotics if all low-risk criteria are met; outpatient treatment for UTI if able to take oral meds; if not UTI and low-risk criteria not met, then admit for LP and IV antibiotics pending culture results
 - c. Infants > 90 days with FWS $> 39^{\circ}\text{C}$ who have received Hib and PCV-7, require at a minimum a U/A and UC; for all females, uncircumcised males < 24 months, circumcised males < 6 months, and any infant with a previous UTI
 7. Close follow-up, often seen outpatient within 24 hours after discharge from hospital

■ DIPHTHERIA

- Definition: Highly contagious acute infection of the upper respiratory tract and/or trachea that is relatively rare in the U.S. but may still occur in under-immunized or unimmunized children
- Etiology/Incidence
 1. Caused by *Corynebacterium diphtheriae*
 2. Transmission through direct contact with infected person, carrier, or contaminated food/objects; spread by respiratory droplets or by contact with discharges from mucous membranes or cutaneous lesions
 3. Rare in the U.S. due to DTaP/dT immunization; approximately 5 cases reported annually
 4. Increased risk among unimmunized children living in crowded conditions
 5. Fall/winter incidence most common
 6. Incubation period is 2 to 7 days
- Signs and Symptoms
 1. Sore throat
 2. Low-grade fever
 3. Nasal discharge
 4. Bloody nose
 5. Hoarseness or cough
 6. Difficulty breathing (severe cases)

- Differential Diagnosis
 1. Acute streptococcal pharyngitis
 2. Nasal foreign body
 3. Mononucleosis
 4. Viral croup
 5. Epiglottitis
 6. Post-tonsillectomy faucial membrane
 7. Thrush
 8. Acute toxoplasmosis
 9. Acute cytomegalovirus
 10. Tularemia
 11. Leukemia
- Physical Findings
 1. Greyish-white pseudomembrane found at location of infection—nasopharynx, pharynx, or trachea
 2. Other findings vary by location of membrane
 3. Toxin-related severe complications include—severe neck swelling (bull neck), myocarditis, Guillain-Barré type neuritis, and paralysis
- Diagnostic Tests/Findings
 1. Positive culture of *C. diphtheriae* from nose or throat or any lesion; notify lab of suspected diphtheria case
 2. CBC—normal or slight leukocytosis and thrombocytopenia
- Management/Treatment
 1. Hospitalization
 2. Evaluation of sensitivity to horse serum; if negative, single dose of equine antitoxin
 3. Antibiotic treatment—erythromycin or penicillin G; antimicrobial treatment is not a substitute for antitoxin; erythromycin (EES) parenterally or orally for 14 days at 40 to 50 mg/kg/day divided by 6 hours; or penicillin G benzathine for 14 days IM, 600,000 units for children < 30 kg and 1.2 million units for children and adults > 30 kg
 4. Respiratory isolation with droplet precaution
 5. Report to state health department
 6. Identification of contacts for follow-up care, immunization, and treatment according to current American Academy of Pediatrics *Red Book* recommendations; includes surveillance for 7 days, culture for *C. diphtheria*, and antibiotic prophylaxis for 7 days with the same dosing as above treatment
 7. Elimination of organism should be documented 24 hours after completion of treatment by 2 consecutive negative cultures 24 hours apart
 8. Prevention through universal immunization; disease may not confer immunity

■ PERTUSSIS (WHOOPIING COUGH)

- Definition: Highly contagious bacterial infection involving the respiratory tract; characterized by prolonged coughing episodes ending in an inspiratory “whoop”
- Etiology/Incidence
 1. Caused by *Bordetella pertussis*
 2. Infection occurs following person-to-person contact via aerosolized droplets from respiratory tract
 3. Pertussis is most common and most severe in infants less than 6 months of age, especially for premature infants and those not immunized, and these children have the highest mortality
 4. Incubation period is 5 to 21 days, usually 7 to 10 days
 5. Infectivity is highest in catarrhal stage and the first 2 weeks after onset of cough
 6. In 2003, the number of middle-school-age cases 10 to 14 years of age, which is traditionally the group for the second highest incidence, exceeded the number of infant cases
 7. No life-long immunity is conferred by disease or immunity
- Signs and Symptoms
 1. Catarrhal stage—mild URI symptoms with cough for approximately two weeks; low-grade fever
 2. Paroxysmal stage—severe coughing episodes with inspiratory “whoops” that may persist for weeks; vomiting, sucking, or crying precipitates coughing episodes; poor feeding and poor weight gain, especially in infants
 3. Convalescent stage—symptoms, mostly coughing, gradually subside over several weeks to months
 4. Infants < 6 months of age may present with atypical early signs—short catarrhal stage, gagging, gasping, apnea spells possibly with cyanosis, absence of whoop, unexpected death, and then followed by a prolonged convalescence
- Differential Diagnosis
 1. Pneumonia (bacterial, viral, chlamydial, mycoplasma)
 2. Acute bronchitis
 3. Croup
 4. Upper respiratory infection
 5. Foreign body aspiration
 6. Cystic fibrosis

- Physical Findings
 1. Catarrhal stage—mild URI symptoms
 2. Paroxysmal stage—severe coughing episodes with inspiratory “whoops,” cyanosis
- Diagnostic Tests/Findings
 1. Chest radiograph—may reveal thickened bronchi and evidence of atelectasis and bronchopneumonia
 2. WBC count reveals marked leukocytosis—usually presents during paroxysmal period and persists for 3 to 4 weeks, more often in infants and young children
 3. Nasopharyngeal culture is still the “gold standard;” requires special transport media which may need to be obtained by the local health department; usually grows in 3 to 4 days, but culture is continued for 10 days; positive in initial phase of illness but not in paroxysmal stage
 4. A negative culture does not exclude diagnosis of pertussis
- Management/Treatment
 1. Medical referral is necessary
 2. Antibiotic therapy—erythromycin (40 to 50 mg/kg/day divided four times a day for 14 days; maximum 2 g/day) or azithromycin; azithromycin is now recommended as drug of choice for treatment and prophylaxis of infants < 1 month of age; dose is 10 mg/kg/day as a single dose for 5 days; any infant receiving a macrolide must be monitored for infantile hypertrophic pyloric stenosis for 1 month after completing the course; the risk of developing severe pertussis and life-threatening complications far outweighs the potential risk of pyloric stenosis that has been associated with erythromycin; trimethoprim-sulfamethoxazole is the alternative for patients unable to tolerate macrolides or who may have a macrolides-resistant strain; dosing at trimethoprim 8 mg/kg/day, twice a day for 14 days
 3. Hospitalized children should remain in isolation until they have received five days of erythromycin
 4. Children receiving oral erythromycin at home should not attend childcare or school until they have received five days of therapy
 5. Supportive treatment for children who cannot tolerate oral intake due to paroxysmal coughing episodes—intravenous hydration, oxygen supplementation, ventilatory support
 6. Report cases of pertussis to state health department
 7. Complication for infants—pneumonia, seizures, encephalopathy, death
- 8. Complications for adolescents and adults—syncope, sleep disturbances, incontinence, rib fractures, pneumonia
- 9. Prevention
 - a. Appropriate pertussis immunization according to schedule
 - b. Children less than 7 years of age, with close contact with infected individual, who are unimmunized or have received fewer than four doses of pertussis vaccine, should complete the series with the minimal intervals
 - c. If the third dose of vaccine was received six months or more prior to the exposure, should be given a fourth dose at time of exposure
 - d. Children who have not received a vaccine within past three years or those ≥ 6 years of age should receive a booster dose of pertussis vaccine at time of exposure; this can be given as Tdap
 - e. Chemoprophylaxis—erythromycin (40 to 50 mg/kg/day divided four times a day for 14 days; maximum 2 g/day) is recommended for all household contacts and other close contacts, regardless of vaccination status

▣ INFLUENZA

- Definition: Highly contagious, viral illness characterized by sudden onset of fever, chills, malaise, headache, myalgia, and dry cough
- Etiology/Incidence
 1. Epidemic influenza caused by types A and B; recent subtypes have included H1N1, H1N2, and H3N2 viruses
 2. Transmission occurs by direct person-to-person contact, via airborne droplet, or by articles contaminated with nasopharyngeal secretions
 3. During outbreak, school-aged children are most frequently infected and infect household contacts
 4. Period of highest infectivity 24 hours prior to onset of symptoms and while symptoms are most severe; viral shedding peaks first 3 days of illness with direct correlation to height of fever
 5. Incubation period 1 to 4 days, mean of 2 days
 6. Influenza season mid-October through mid-February
- Signs and Symptoms
 1. Fever
 2. Chills

3. Malaise
 4. Headache, myalgia
 5. Dry cough
 6. Anorexia
 7. Rhinorrhea
 8. Sore throat
 9. Less frequently—conjunctivitis, abdominal pain, nausea, vomiting, diarrhea
- Differential Diagnosis
 1. Upper respiratory infections
 2. Pneumonia
 - Physical Findings
 1. Listlessness
 2. Nonproductive cough
 3. Rhinorrhea
 4. Rigors
 5. Fever
 6. Conjunctivitis
 7. Pharyngitis
 - Diagnostic Tests/Findings
 1. Nasopharyngeal cultures obtained within first 72 hours of illness may reveal influenza; or if using a rapid antigen test for influenza, it can designate the virus as type A or B
 2. Diagnosis is usually made based on clinical signs and available prevalence data
 - Management/Treatment
 1. Management of influenza is primarily supportive
 - a. Bed rest
 - b. Acetaminophen or ibuprofen for fever (avoid aspirin-containing products due to risk of developing Reye's syndrome)
 - c. Adequate hydration
 2. Rimantadine diminishes severity of influenza A but is not effective in treatment of influenza B; oseltamivir, a neuraminidase inhibitor (NA), is approved for treatment of uncomplicated influenza A and B within 2 days of symptom onset; NA inhibitors are the only available antiviral drugs active against the type B influenza viruses
 - a. Not approved for use in infants < 12 months of age
 - b. Dosages—rimantadine 5 mg/kg/day for 5 days (not to exceed 150 mg/day for children 1 year to 9 years of age; for children ≥ 10 years of age and < 40 kg give 5 mg/kg/day, or 200 mg/day if ≥ 40 kg; oseltamivir ≤ 15 kg–30 mg orally once daily for 5 days; 16 to 23 kg–45 mg orally for 5 days; 24 to 40 kg–60 mg orally for 5 days; children > 40 kg and adults 75 mg orally for 5 days)
3. Complications of influenza—secondary bacterial infection; sepsis-like picture in infants; febrile seizures; encephalopathy; myocarditis; sudden death, even in previously healthy children
 4. AAP and CDC recommendations at time of publication are for annual vaccinations in all children 6 to 59 months of age, for household contacts and out-of-home caregivers of children 0 to 59 months of age, and for children and adolescents in high-risk groups
 5. High-risk children include—asthma, cystic fibrosis, other chronic pulmonary disease, significant cardiac disease, immunosuppressive disorders and treatment, HIV, sickle cell anemia, other hemoglobinopathies, medical conditions requiring long-term salicylate medication such as JRA or Kawasaki disease, chronic renal or metabolic (diabetes) diseases, or any cognitive dysfunction, spinal cord injury, or neuromuscular diseases that compromise respiratory functioning
 6. There are 2 forms of influenza vaccine—inactivated trivalent influenza vaccine (TIVA) and live-attenuated influenza vaccine (LAIVA); LAIVA is only for healthy children with no known risk factors and approved for ages 5 to 49 years; also do not give to children with a history of anaphylactic reaction to egg protein or a history of Guillain-Barré syndrome; TIVA is given to all other infants and children and for close contacts of severely immunocompromised individuals
 7. Children younger than 9-years-old who are getting an influenza vaccine for the first time need to receive 2 doses of TIVA or LAIVA at least 1 month apart, preferably finishing the 2 dose regimen by December; this boosts antibody response; in subsequent years, children < 9 years old who had 1 or 2 doses of vaccine need only 1 dose per year; children > 9 years old require just 1 dose per year
 8. The CDC and state health departments will post recommendations for prevention and treatment guidelines each year and periodically as warranted

▣ RUBELLA (GERMAN MEASLES)

- Definition: An acute, contagious, viral disease characterized by a minor or absent prodrome, swelling of suboccipital, postauricular and cervical nodes, and followed by generalized rash
- Etiology/Incidence
 1. Caused by an RNA virus, a Rubivirus in the Togaviridae family

2. Postnatal transmission occurs via contact from nasopharyngeal secretions
 3. Incubation period ranges from 14 to 23 days
 4. Peak incidence is late winter and early spring
 5. Preventable by active immunization
- Signs and Symptoms
 1. History of inadequate immunization
 2. Rash starts on forehead and face and spreads over trunk and extremities during the 1st day; facial exanthem fades by 2nd day, disappears by 3rd day
 3. Associated signs and symptoms
 - a. Malaise, low-grade fever
 - b. Transient joint pain, polyarthralgia or polyarthritis rare in children, but more commonly seen in adolescents and adults, particularly in females
 - c. Bruising (rare)
 - Differential Diagnosis
 1. Rubeola
 2. Scarlet fever
 3. Erythema infectiosum
 4. Adenovirus
 5. Rocky mountain spotted fever
 6. Roseola
 7. Drug eruption
 - Physical Findings
 1. Generalized erythematous, maculopapular discrete rash—usually first indication of illness
 2. Listlessness
 3. Postauricular, suboccipital, and posterior cervical lymphadenopathy—usually precedes rash
 4. Petechiae on soft palate and uvula (Forchheimer's sign)
 5. Purpura/petechiae (rare)
 6. Meningeal signs (rare)
 - Diagnostic Tests/Findings
 1. Presence of rubella-specific IgM antibody indicates recent postnatal infection or congenital infection in newborn
 2. Refer to current American Academy of Pediatrics *Red Book* for further information on available assays for detecting rubella infection
 - Management/Treatment
 1. Management of uncomplicated infection is primarily supportive—includes fever and pain (in lymph nodes); control with acetaminophen or ibuprofen
 2. Determine contacts that may require immunization

3. Infected children should limit contact with susceptible persons, including women of childbearing age; out of school for 5 days after onset of rash
4. Educate adolescent females regarding teratogenic nature of rubella in pregnancy, resulting in congenital rubella syndrome—multiple congenital anomalies affecting eyes, heart, auditory with hearing loss, and neurologic systems
5. Educate caretakers regarding complications of arthritis, and rarely thrombocytopenia and encephalitis

▣ RUBEOLA (RED MEASLES)

- Definition: An acute, highly contagious viral disease characterized by prodrome of upper respiratory manifestations followed by generalized maculopapular eruptions
- Etiology/Incidence
 1. Caused by an RNA virus, a morbillivirus, in Paramyxoviridae family
 2. Transmitted by direct contact with infected secretions or via airborne droplets through sneezing or coughing
 3. Incubation period is 7 to 18 days
 4. Infected individuals are contagious 3 to 5 days before appearance of rash, to 4 days after appearance of rash
 5. Increased incidence during late winter and spring
 6. Preventable by active immunization
- Signs and Symptoms
 1. History of inadequate immunization
 2. Acute onset of fever, coryza, cough, conjunctivitis, malaise, anorexia
 3. Confluent, erythematous, maculopapular rash 3 to 4 days after initial symptoms; progresses in caudal direction, beginning behind the ears and at the hairline
- Differential Diagnosis
 1. Roseola or rubella
 2. Viral infections (e.g., echovirus, coxsackievirus, adenovirus)
 3. Infectious mononucleosis
 4. Scarlet fever
 5. Rickettsial diseases
 6. Serum sickness
 7. Morbilliform drug eruption
 8. Secondary syphilis

- Physical Findings
 1. Confluent, erythematous maculopapular rash; after 3 to 4 days, rash assumes a brownish appearance
 2. Profuse coryza
 3. Conjunctivitis, photophobia, periorbital edema in prodrome
 4. Pulmonary findings (crackles, rhonchi), a hacking, bark-like cough
 5. Koplik spots (red eruptions with white centers on buccal mucosa) prior to appearance of rash
 6. Generalized lymphadenopathy
- Diagnostic Tests/Findings: Presence of measles-specific IgM antibody suggests recent infection
- Management/Treatment
 1. Medical referral necessary
 2. No specific antiviral therapy available; WHO recommends vitamin A to any child who may have a vit A deficiency, especially in third world countries
 3. Management of uncomplicated measles is primarily supportive—bed rest, adequate hydration; acetaminophen or ibuprofen for fever; antitussive therapy
 4. Otitis media is most common complication of measles infection—treated with same antibiotics as in standard otitis media
 5. Educate caretakers regarding complications, including otitis media, diarrhea, encephalitis, croup, and pneumonia

■ ROSEOLA (EXANTHEM SUBITUM)

- Definition: An acute contagious disease characterized by high fever and appearance of a rash with simultaneous decrease in fever
- Etiology/Incidence
 1. Caused by human herpesvirus 6 (HHV-6)
 2. Mode of transmission not known
 3. Incubation period is 5 to 15 days
 4. Period of infectivity is thought to be during the febrile episode, prior to appearance of the rash
 5. Most commonly occurs in children 6 to 24 months of age
 6. Most cases occur in spring and summer
 7. One attack confers life-long immunity
- Signs and Symptoms
 1. Abrupt onset of high fever (102 to 105°F) lasting 3 to 5 days
 2. Appearance of a rash follows resolution of fever
 3. Associated symptoms include irritability and swelling of eyelids
 4. Febrile seizures

- Differential Diagnosis
 1. Rubeola
 2. Scarlet fever
 3. Rubella
 4. Erythema infectiosum
 5. Other viral exanthems
 6. Meningococemia
 7. UTI
- Physical Findings
 1. Generalized erythematous, maculopapular rash; starts on trunk and spreads to arms and neck with less involvement of face and legs; generally a well-appearing infant
 2. Irritability
 3. May have mildly inflamed edematous conjunctiva
- Diagnostic Tests/Findings: Progressive leukopenia during febrile period
- Management/Treatment
 1. Acetaminophen or ibuprofen for fever; tepid baths; fluids
 2. Medical referral if meningeal signs appear or if fever persists
 3. Education
 - a. Potential for febrile seizures
 - b. Reassurance that appearance of rash is sign of recovery

■ FIFTH DISEASE (ERYTHEMA INFECTIOSUM–EI)

- Definition: A contagious, usually afebrile, exanthematous disease; commonly known as “fifth disease”, it was the fifth childhood exanthem described after measles, rubella, scarlet fever, and roseola
- Etiology/Incidence
 1. Human parvovirus B19
 2. Typically seen in 5- to 14-year-old children
 3. Outbreaks occur most often during late winter and spring months
 4. Incubation period between 4 and 14 days, up to 21 days
 5. Mode of transmission includes respiratory secretions and blood
 6. Most infectious prior to rash
 7. About 60% of adults are immune
- Signs and Symptoms
 1. May have prodromal symptoms of a mild URI for 2 to 3 days, preceding the rash by 7 to 10 days; low-grade fever, headache, chills,

malaise, myalgia, pharyngitis, conjunctivitis, arthralgias, arthritis

2. Rash
 - a. Begins as bilateral erythema on cheeks with circumoral pallor (“slapped cheek” appearance)
 - b. Spreads to upper arms, legs, trunk, buttocks, hands, and feet
 - c. Palms and soles are spared
 - d. Lacy-reticular exanthem, slightly raised, appears as facial erythema, begins to diminish
 - e. May reappear when skin is exposed to sunlight, temperature extremes, or friction
 - f. Rash lasts from 2 to 39 days, average 11 days
 3. Less common symptoms—mild URI with no rash, rubelliform rash, papulopurpuric glove-and-socks-syndrome (painful and pruritic papules, petechiae, purpura of hands and feet)
 4. Can cause aplastic crises in young children, patients with hemolytic diseases, or if immunocompromised
 5. Arthralgias and arthritis are more common in adolescents and adults
- Differential Diagnosis
 1. Drug reactions
 2. Rubella, atypical measles
 3. Enteroviral diseases
 4. Systemic lupus erythematosus
 5. Juvenile rheumatoid arthritis
 - Physical Findings
 1. Early—bilateral erythema on cheeks (“slapped cheek” appearance)
 2. Late—erythematous, lacy-reticular rash appears as facial erythema begins to diminish and is seen on upper arms and legs, trunk, hands and feet; palms and soles are spared
 - Diagnostic Tests/Findings
 1. Parvovirus B19 IgM antibody confirms current infection, or infection within past several months
 2. Parvovirus B19 IgG antibody indicates previous infection and immunity
 - Management/Treatment
 1. None indicated
 2. Reassure parent of benign nature of disease
 3. Avoid sunlight as exposure may exacerbate the condition
 4. Period of high infectivity in persons with EI is prior to onset of symptoms; unlikely to be infectious after rash develops; conversely patients with aplastic crises are highly

contagious prior to the onset of symptoms and through week of onset or longer

5. Can result in spontaneous abortion or stillbirth; complications to a developing fetus can occur, including hydrops fetalis and possible fetal death, with the greatest risk in the first half of the pregnancy, but Parvo B 19 has not been proven to cause congenital anomalies; the risk of fetal death is 2 to 6%; pregnant women exposed to the Parvo virus should discuss the implications with their OB/GYN provider
6. Prevention—hand hygiene and appropriate disposal of facial tissues

▣ VARICELLA-ZOSTER VIRUS (VZV) (CHICKENPOX)

- Definition: An acute contagious disease caused by a herpes virus; characterized by a short or absent prodrome and usually a sequential rash consisting of papules, vesicles, pustules, and crusts
- Etiology/Incidence
 1. VZV is a herpes virus
 2. Transmission occurs by direct contact with varicella lesions or by airborne droplet infection
 3. Susceptible individuals can contract chickenpox from patients with varicella zoster (shingles)
 4. Incubation period between 10 and 21 days, most commonly 14 to 16 days
 5. Infected individual contagious for 24 to 48 hours prior to outbreak of lesions, until all lesions have crusted over
 6. Most cases occur in children younger than 10 years old
 7. VZV occurs commonly in late winter and early spring
 8. Primary infection generally confers life-long immunity
 9. There is a latent phase in which the virus resides in the dorsal root ganglia and may be reactivated at a later time as eruptions of “shingles”
 10. If children received only 1 dose of the 2 series Varicella vaccines, there can be a second episode of varicella
- Signs and Symptoms
 1. Early lesions appear as faint erythematous macules that progress to papules, followed by appearance of vesicles primarily on trunk, scalp, face; lesions eventually crust over
 2. Lesions continue to erupt for 3 to 4 days and may be present in various stages

3. Associated symptoms of a prodrome may include fever, headache, pruritus, malaise, general aches and pains, anorexia, joint pain
- Differential Diagnosis
 1. Herpes zoster
 2. Bullous impetigo
 3. Insect bites
 4. Urticaria
 5. Disseminated herpes simplex virus
 6. Scabies
 7. Smallpox
 - Physical Findings
 1. Crops of skin lesions that may appear as maculopapular (early), vesicular, pustular with eventual crusts; many maculopapular lesions may progress to vesicular stage and resolve without crusting
 2. Rash usually present on scalp, face, trunk and extremities; most lesions on face and trunk with anywhere from a few to several hundred lesions; all stages of lesions are often seen simultaneously; vesicles become umbilicated and progress to pustules and crusts within 8 to 12 hours; vesicles are watery yellow, pustules have white pus, crusts are brownish-red and fall off in 1 to 3 weeks leaving pink, often punched-out permanent scars
 3. Hepatomegaly (rare)
 4. Meningeal signs (rare)
 5. Pulmonary findings—crackles, wheezes (rare)
 - Diagnostic Tests/Findings: None routinely performed
 - Management/Treatment
 1. VZV is a self-limited disease lasting 7 to 10 days
 2. Supportive treatment
 - a. Control of pruritus with oatmeal baths, diphenhydramine, calamine lotion
 - b. Acetaminophen for fever (avoid aspirin-containing products due to risk of developing Reye's syndrome)
 3. Oral acyclovir is beneficial in reducing duration of new lesion formation and total number of lesions (20 mg/kg/dose, four times a day; maximum 800 mg, four times a day); should be started within 24 hours of onset for maximum benefit
 4. Oral acyclovir not usually recommended in healthy children with uncomplicated varicella
 5. Varicella-zoster immune globulin (VZIG) should be given to immune suppressed contacts to provide passive protection
 - 6. Medical referral necessary for immune suppressed children who are at risk for severe disease, such as pneumonia, encephalitis, glomerulonephritis, and hepatitis
 - 7. Education
 - a. Avoid contact with elderly, pregnant women, neonates, and immunocompromised children
 - b. Children may return to school when all lesions are crusted, or in immunized children without crusts when the lesions appear to be resolving
 - c. Live-attenuated varicella vaccine is available in U.S., and 2 doses are now recommended for all healthy children at age 12 months and 4 to 6 years of age; adolescents past their 13th birthday who lack a reliable history of varicella should be given 2 doses of varicella vaccine spaced 4 weeks apart
 - d. Signs and symptoms of complicated varicella infection—meningeal signs, respiratory distress, dehydration, ocular involvement, secondary bacterial infection, thrombocytopenia, pneumonia
 - e. Signs and symptoms of Reye's syndrome—persistent vomiting, lethargy, agitation, disorientation, combativeness, coma

▣ MUMPS

- Definition: A contagious, systemic, viral disease characterized by swelling of the salivary glands
- Etiology/Incidence
 1. Caused by an RNA virus, Rubulavirus in the Paramyxoviridae family
 2. Spread by direct contact via respiratory airborne droplet and fomites contaminated with infected saliva
 3. Incubation period between 12 to 25 days after exposure, usually 16–18 days
 4. Infected individual is contagious for as many as 7 days prior to, and as long as 9 days after onset of symptoms
 5. Infection occurs throughout childhood; rarely during adulthood
 6. More common in late winter and spring
- Signs and Symptoms
 1. History of inadequate immunization
 2. Swelling of salivary glands (specifically parotid gland), pain with swallowing
 3. Malaise, fever
 4. Orchitis, which is scrotal swelling and pain, rare cases of sterility

- Differential Diagnosis
 1. Submandibular or preauricular lymphadenitis
 2. Salivary duct obstruction
 3. Infectious mononucleosis
 4. Epididymitis
- Physical Findings
 1. Swelling of salivary glands (specifically parotid gland)
 2. Lethargy
 3. Scrotal swelling and pain
- Diagnostic Tests/Findings: Serum for complement fixation (CF)—positive test for complement-fixing antibody against mumps virus suggests recent infection
- Management/Treatment
 1. Acetaminophen for pain and fever
 2. Warm compresses for salivary gland swelling
 3. Soft or liquid diet
 4. Education
 - a. Complications include pancreatitis, oophoritis, meningitis, orchitis
 - b. May return to childcare or school when all symptoms have resolved or 9 days after onset of symptoms
 5. Report cases to state health department

■ CAT SCRATCH DISEASE (CSD)

- Definition: An infection characterized by regional lymphadenopathy in an otherwise healthy person, following contact with an infected cat, as a result of a cat bite or scratch, or contact with cat saliva on broken skin or the conjunctivae of the eye
- Etiology/Incidence
 1. Most cases are caused by *Bartonella henselae*
 2. Cats are common reservoir for human disease; no human-to-human transmission
 3. More common in children, peak incidence 5 to 14 years of age
 4. Late fall, winter, early spring; July and August in warmer climates
 5. Often multiple cases in the same family
- Signs and Symptoms: Mild systemic symptoms
 1. History of cat exposure, usually a kitten < 6 months old
 2. Usually do not appear ill
 3. Swollen lymph nodes
 4. May have low-grade fever, general malaise, headache, nausea, chills, general aching
 5. Anorexia
 6. May have rash
- Differential Diagnosis
 1. Bacterial lymphadenitis
 2. Lymphoma
 3. Tularemia
 4. Lymphogranuloma venereum
 5. Atypical mycobacteria
 6. Infectious Mononucleosis
 7. Toxoplasmosis
- Physical Findings
 1. Papule or pustule at site of cat scratch or bite 7 to 12 days after cat contact, followed in 1 to 4 weeks by enlargement of an associated regional lymph node
 2. Lesion may be present for several days to several months
 3. May be erythematous, hot, firm, and tender to touch
 4. Most common on the head, neck, or extremities
 5. Conjunctivitis—if portal of entry is the conjunctiva, a granuloma on the palpebral conjunctiva with associated tender preauricular and/or cervical lymphadenopathy is present
 6. Involved node, usually single, draining the site of inoculation
- Diagnostic Tests/Findings
 1. May have elevated ESR
 2. Immunofluorescence assay (IFA) detects antibody to CSD
 3. Warthin-Starry silver stain used to identify CSD in lymph node, skin, or conjunctival tissue
- Management/Treatment
 1. CSD is a self-limited disease lasting 2 to 4 months
 2. Supportive treatment, may be unresponsive to antibiotics
 - a. Analgesics for discomfort and fever, warm compresses
 - b. Limited activity per comfort level
 - c. Needle aspiration of painful, suppurative nodes questionable; may result in chronic draining sinus tract; I & D not recommended; rarely is surgical removal of nodes necessary; no treatment required for animal that transmitted CSD; no declawing is needed, nor is removal of cat from home necessary

■ ROCKY MOUNTAIN SPOTTED FEVER (RMSF)

- Definition: A systemic, febrile illness causing a generalized vasculitis with characteristic petechial or purpuric rash
- Etiology/Incidence
 1. Caused by *Rickettsia rickettsii*
 2. Transmitted to humans via tick bites
 3. Usually occurs in persons younger than 15 years of age
 4. Widespread in U.S.; most prevalent in southern states April to September
 5. Incubation period ranges from 2 to 14 days
- Signs and Symptoms
 1. Fever, myalgia, severe headache, anorexia, nausea, and vomiting precede appearance of rash; abdominal pain, diarrhea, sudden shaking rigor, photophobia, and prostration may also be present
 2. Erythematous, macular rash (usually appearing before the sixth day of illness), on wrists, ankles, spreading within hours to the trunk, and the palms and soles are often involved; macules become papular in 1 to 3 days
 3. In some cases rash fails to develop or develops late in the illness
 4. Disease can last 3 weeks with multisystem involvement and significant long-term sequelae (e.g., CNS, cardiac, pulmonary, GI, renal, DIC)
- Differential Diagnosis
 1. Rubeola
 2. Rubella
 3. Lyme disease
 4. Septicemia
 5. Meningococemia
 6. Ehrlichiosis and other rickettsial diseases
 7. Drug reaction
 8. Immune-complex-mediated vasculitis
 9. Acute appendicitis and acute cholecystitis
- Physical Findings
 1. Characteristic petechial, erythematous, maculopapular rash
 2. Neurologic deficits, altered consciousness, transient deafness
 3. Heart murmur
 4. Pulmonary findings (crackles)
 5. Decreased urine output
 6. Jaundice, hepatomegaly, splenomegaly
- Diagnostic Tests/Findings
 1. Culture is not attempted due to transmission risk for laboratory personnel
 2. Renal failure
 3. WBC typically is normal, leukopenia, anemia, thrombocytopenia, hyponatremia; elevated liver enzymes
 4. Increase in antibody titer as established by serologic testing; indirect hemagglutination (IHA) and microimmunofluorescence (micro-IF) are most sensitive
- Management/Treatment
 1. Medical referral
 2. Doxycycline is the drug of choice and is initiated based on clinical picture before lab results are available; it is given until patient is afebrile for 3 days and is showing signs of clinical improvement; the usual duration is 7 to 10 days; treatment by day 5 of illness provides the best chance for a good outcome
 3. Tetracyclines are not routinely given to children less than 8 years of age, but doxycycline is still the drug of choice to be used at any age; doxycycline has less tooth enamel staining than tetracycline and is effective against ehrlichiosis, which can mimic RMSF
 4. Chloramphenicol or a fluoroquinolones are alternative medicines, chloramphenicol has serious major side effects
 5. Untreated RMSF has a mortality rate as high as 20%; with adequate treatment, the mortality rate is lowered to 3%; early diagnosis and treatment has the best outcome
 6. Education includes preventive measures such as use of tick repellent and protective clothing in tick-infested areas; checking for ticks after spending time outdoors and removing them promptly
 7. Patients with multisystem organ involvement may require rehabilitative services
 8. Report cases to state health department

■ LYME DISEASE

- Definition:
 1. An immune-mediated, inflammatory response
 2. Affects multiple organ systems
 3. Transmitted primarily via the deer tick
- Etiology/Incidence
 1. Caused by a spirochete *Borrelia burgdorferi*; is most often transmitted via the deer tick; bacterial transmission usually occurs after the tick is embedded and feeding for 36 hours
 2. Most often seen in Northeast from Massachusetts to Maryland; the Midwest, primarily Wisconsin and Minnesota; and in California; primarily in heavily wooded areas with nearby tall grassy areas

3. Persons of all ages and both sexes are affected, but the incidence in the U.S. is highest among children ages 5 to 9 years and adults 45 to 54 years
 4. Most cases occur from April to October with > 50% of cases during June and July
 5. Incubation period is between 1 to 55 days with an average of 11 days
- Signs and Symptoms in 3 stages
 1. Early localized stage
 - a. Appearance of well-circumscribed, erythematous, annular rash with central clearing (erythema migrans) at site of recent tick bite; an expanding macule that is painless and not pruritic, usually flat except at the central bite mark, and may have partial central clearing
 - b. Accompanied by fever, malaise, headache, arthralgia, conjunctivitis, or mild neck stiffness, myalgia
 - c. May initially present as the above flu-like illness if erythema migrans does not occur, or not recognized
 2. Early disseminated stage
 - a. Multiple erythema migrans occurring several weeks after the known or unknown tick bite
 - b. Accompanied by systemic symptoms like arthralgia, myalgia, headache, and fatigue; carditis is rare in children
 3. Late disease stage (weeks to months later)
 - a. Migratory pain in joints, muscles, and bones
 - b. Transient, but severe, headaches and stiff neck
 - c. Poor memory, mood changes, somnolence
 - d. Muscle weakness and poor coordination
 - e. Chest pain, cardiac abnormalities
 - f. Dizziness/fainting
 - g. Facial palsies, peripheral neuropathies
 - h. Joint stiffness, recurrent arthritis that is pauciarticular affecting large joints, especially the knees
 - Differential Diagnosis
 1. Tinea corporis (ringworm)
 2. Herald patch of pityriasis rosea
 3. Insect bite
 4. Cellulitis
 5. Urticaria
 6. Acute rheumatic fever
 7. Influenza
 8. Aseptic meningitis
 9. Juvenile rheumatoid arthritis
 10. Henoch-Schönlein purpura
 - Physical Findings
 1. Well-circumscribed, erythematous, annular rash with central clearing
 2. Malar rash, diffuse erythema or urticaria
 3. Heart murmur
 4. Neurologic findings—seventh cranial nerve palsy
 - Diagnostic Tests/Findings
 1. Enzyme-linked immunosorbent assay (ELISA)—detects antibodies against *B. burgdorferi*
 2. Western blot—used to validate a positive or equivocal ELISA
 3. Serum immunoglobulins—IgM, IgG elevated, but not until after the first few weeks and if treatment is begun based on characteristic rash, seroconversion is blocked by antibiotic use
 4. Culture of erythema migrans lesion—expensive, time to isolation may take four weeks
 5. White blood cell count—normal or elevated
 6. Erythrocyte sedimentation rate (ESR)—elevated
 - Management/Treatment
 1. Medical referral is necessary for late disease or chronic symptoms
 2. Early disease
 - a. Children > 8 years of age—doxycycline, which is the drug of choice, or amoxicillin; doxycycline 100 mg twice a day for 14 to 21 days
 - b. Children ≤ 8 years of age—amoxicillin, 50 mg/kg/day twice a day for 14 to 21 days
 - c. For penicillin-allergic patients—cefuroxime or erythromycin and azithromycin may be used, although erythromycin and azithromycin are less effective
 3. Early disseminated disease
 - a. multiple erythema migrans is treated the same as for early localized disease, but for 21 days
 - b. isolated facial palsy is the same for 21 to 28 days
 - c. arthritis is the same for 28 days
 4. Late disease for persistent arthritis, carditis, neurologic disease—parenteral ceftriaxone or penicillin G, referral to infectious specialist or Lyme specialist
 5. Education/prevention
 - a. Ticks that carry Lyme disease are 4 to 5 mm in diameter; nymphs are smaller
 - b. Avoidance of tick-infested areas; prompt removal of ticks from the skin and use of tick repellent decreases the incidence of Lyme disease

- c. Use blunt-end tweezers to grasp tick as close to skin surface as possible and gently and steadily pull it straight off; wear rubber gloves; look on the head, neck, behind the ears, the axilla, belt line, and groin
- d. Disinfect skin where tick bite occurred
- e. Early intervention leads to improved prognosis
- f. Wear protective clothing in heavily wooded areas, light-colored clothing is better to see ticks, long-sleeved shirts tucked into pants, long pants tucked into socks, wear a hat, spray clothes with a permethrin; use DEET insect repellents to exposed areas of skin with cautious use on young children; check skin closely after outdoor activities, daily if live near woods where deer and mice live or if live in an endemic part of the country
- g. Keep pets tick free with daily inspection and prompt removal of any ticks
- h. Report cases of Lyme disease to state health department

■ INFECTIOUS MONONUCLEOSIS (IM)

- Definition: An acute disease characterized by fever, exudative pharyngitis, lymphadenopathy, hepatosplenomegaly, and atypical lymphocytosis
- Etiology/Incidence
 1. Most commonly caused by Epstein-Barr virus (EBV), a gammaherpesvirus of the lymphocryptovirus genus
 2. Contact with infected secretions or blood is required for transmission, virus is viable in saliva for several hours outside the body, fomite transmission is unknown
 3. Incubation period is 30 to 50 days
 4. Commonly diagnosed in adolescents and young adults
 5. No seasonal pattern
 6. Viral shedding occurs many months after acute infection and intermittently lifelong; asymptomatic carriage is common
 7. Range of illness is wide, from asymptomatic to fatal infections
- Signs and Symptoms
 1. Fever (101–104°F), malaise, fatigue, headache, rhinitis, cough
 2. Abdominal pain, anorexia, nausea, vomiting
 3. Severe sore throat, difficulty swallowing, possibly to the point of dehydration
 4. Rash, especially with administration of ampicillin derivatives
- 5. Tender, enlarged lymph nodes, including posterior cervical chain
- 6. Generally begins as URI signs and symptoms, with increasing sore throat and enlarged tonsils and lymph nodes, along with increasing fatigue
- Differential Diagnosis
 1. Streptococcal pharyngitis
 2. Hepatitis
 3. Influenza or viral illness
 4. Measles
 5. Blood dyscrasias, especially leukemia
 6. Cytomegalovirus
 7. HIV
- Physical Findings
 1. Exudative tonsillitis with patchy white or gray exudates
 2. Inflamed pharynx; possibly petechiae at junction of hard and soft palates
 3. May have hepatosplenomegaly
 4. Tender anterior and posterior cervical nodes
 5. May have jaundice
 6. May have erythematous, macular, papular rash
 7. Periorbital edema
- Diagnostic Tests/Findings
 1. Positive monospot after 7 to 10 days, or positive Epstein-Barr virus IgM titer with acute illness appears in the first 2 weeks of disease and may need 7 to 10 days to show a positive test; children younger than 4 years of age are more often negative
 2. Positive IgG reveals post acute or past infection
 3. WBC count reveals leukocytosis, 10,000 to 20,000 cells/mm³, with ≥ 60% lymphocytes and 20 to 40% atypical lymphocytes
 4. Liver function tests may be elevated with hepatomegaly and/or jaundice
 5. Rapid strep test and throat culture—identifies presence of β-hemolytic streptococcal infection, if present
- Management/Treatment
 1. IM is a self-limited disease lasting 2 to 3 weeks
 2. Supportive treatment
 - a. Bedrest and liquids during acute phase
 - b. Antipyretics for fever and analgesics for pharyngitis and lymphadenitis
 - c. Saline gargles for sore throat
 - d. Isolation is unnecessary and may return to school or work.

- e. Schedule return appointments weekly until completely recovered and no longer with splenomegaly
- f. Avoid strenuous exercise and contact sports until child is fully recovered, which can be 3 to 4 weeks or up to 2 months from illness onset, and the spleen is no longer palpable; splenic ultrasound may be needed to show resolution prior to return to sports, particularly contact sports such as football, hockey, soccer, lacrosse, and swimming
- g. Antibiotic therapy as needed for pharyngitis—avoid use of ampicillin derivatives such as amoxicillin and other penicillins; may result in a nonallergic morbilliform rash
- h. Short-course corticosteroids are not routinely recommended, but may be prescribed for patients with marked tonsillar hypertrophy and impending airway obstruction or dehydration, massive splenomegaly, myocarditis, or hemolytic anemia; oral prednisone is prescribed at 1 mg/kg/day for 5 to 7 days with a taper
- i. Acyclovir or other antivirals are not needed for immunocompetent patients

■ INFANT BOTULISM

- Definition: A neuroparalytic disorder affecting young infants (< 6 months) resulting from ingestion of *Clostridium botulinum* spores with release of toxins as organism colonizes gastrointestinal tract
- Etiology/Incidence
 1. Etiologic agent is *Clostridium botulinum*; disease is caused by toxins produced by this anaerobic bacillus
 2. Toxin inhibits acetylcholine release at myoneuronal junction resulting in impaired motor activity
 3. *C. botulinum* spores have been associated with honey, reported association with corn syrup not substantiated
 4. Rural, farm environments associated with increased incidence
 5. May occur in breastfed infants when first introducing nonhuman milk
 6. Incubation period about 3 to 30 days from time of ingestion
- Signs and Symptoms: Evolving symptomatology
 1. May be asymptomatic or rapidly progressive to apnea and sudden death
 2. Constipation (most common presenting symptom)

3. Poor or slow feeding
4. Weak cry
5. Truncal weakness
6. Loss of head and neck control, decreased movement
7. Loss of facial expression
8. Progressive, descending generalized weakness and hypotonia (floppiness)

- Differential Diagnosis
 1. Sepsis
 2. Benign congenital hypotonia
 3. Benign constipation
 4. Hypothyroidism
 5. Hirschsprung's disease
 6. Other neuromuscular disorders
- Physical Findings
 1. Generalized weakness
 2. Diminished deep tendon reflexes
 3. Cranial nerve deficits
 4. Swallowing difficulties
 5. Loss of muscle tone
 6. Diminished gag reflex
 7. Ocular palsies
- Diagnostic Tests/Findings
 1. Stool specimen for toxin assay is test of choice for infant botulism
 2. Stool culture positive for *C. botulinum*
 3. Blood culture—may or may not be positive for *C. botulinum*
- Management/Treatment
 1. Human-derived antitoxin (botulism immune globulin intravenous or known as BabyBIG) is treatment of choice
 2. Equine antitoxin not usually recommended for infant botulism
 3. Hospitalization; possibility of respiratory arrest
 4. Stool softener
 5. Prevention/education regarding honey as potential source of botulism; avoid feeding to infants < 12 months of age
 6. Report to state health department

■ POLIOMYELITIS

- Definition: An acute, contagious, potentially paralytic viral disease
- Etiology/Incidence
 1. Caused by enterovirus (EV)
 2. When susceptible person comes in contact with poliovirus, one of three responses occur:
 - a. Nonspecific febrile illness (most frequent)—fever, myalgia, sore throat, headache

- b. Aseptic meningitis (nonparalytic poliomyelitis)—headache, stiff neck, spinal rigidity, nausea
 - c. Paralytic poliomyelitis (least frequent)—flaccid paralysis, loss of reflexes, asymmetrical paralysis, proximal limb muscles and lower extremity muscles are more involved
3. Incubation period of asymptomatic or nonparalytic polio is 3 to 6 days; that of paralytic polio to onset of paralysis is 7 to 21 days
 4. Preventable by active immunization—IPV (inactivated poliovirus vaccine) is now used exclusively in the U.S. since 2000, when the OPV (oral poliovirus vaccine) was discontinued; OPV was known to cause VAPP (vaccine-associated paralytic poliomyelitis)
 5. Spread by fecal-oral routes and respiratory secretions; virus persists in throat for about 1 week and shed in feces for several weeks
 6. Most contagious shortly before and after onset of clinical signs of disease
 7. Occurs more often in infants and young children
 8. Occurs more commonly in conditions of poor hygiene
- Signs and Symptoms
 1. History of inadequate immunization or recent immunization
 2. Fever, weakness
 3. Anxiety
 4. Urinary incontinence
 5. Meningeal signs
 6. Respiratory compromise
 7. Speech disturbances
 8. Headache
 9. Nausea, anorexia
 - Differential Diagnosis
 1. Guillain-Barré syndrome
 2. Meningitis
 3. Encephalitis
 4. Peripheral neuritis or neuropathy
 - Physical Findings
 1. Meningeal signs
 2. Respiratory compromise
 3. Inability to speak without frequent pauses
 4. Muscle weakness
 - Diagnostic Tests/Findings: EV isolated from feces, throat, urine or CSF in cell culture; involve the local health department; isolate is sent to CDC
 - Management/Treatment
 1. Medical referral necessary
 2. Supportive management—bed rest, adequate hydration, pain control, acetaminophen or

ibuprofen for fever, respiratory support if paralysis ensues, physical therapy for deficits associated with muscle weakness and paralysis

3. Education

- a. Educate family regarding potential complications of possible paralysis including respiratory compromise and arrest, hypertension, renal calculi due to immobility

■ TETANUS (LOCKJAW)

- Definition: A neurologic disease characterized by severe muscle spasms that can be fatal
- Etiology/Incidence
 1. Caused by neurotoxin produced by anaerobic bacterium *Clostridium tetani* in contaminated wounds
 2. Occurs throughout the world; neonatal tetanus is common in countries where women are not immunized and the umbilical cord may be a source for entry
 3. Incubation period 2 days to 2 months with most occurring in 14 days; 5 to 14 days in neonates
 4. More common in warmer climates and warmer months
 5. Has dramatically decreased with advent of tetanus vaccine
 6. *Clostridium tetani* is ubiquitous in the environment, in soil and human and animal intestines
- Signs and Symptoms
 1. Incomplete tetanus immunization series
 2. History of deep puncture wound, laceration
 3. Insidious onset, gradual over 1 to 7 days
 4. Muscle spasms aggravated by stimuli—sound, light, movement
 5. Muscle rigidity
 6. Increased oral secretions
 7. Respiratory compromise
 8. Begins with pain at site of wound, followed by regional muscle spasm, by 48 hours there is difficulty opening the jaw (trismus), followed by generalized tetany
 9. In infants, first sign is irritability and inability to nurse or feed; followed by stiffness of jaw, neck, increasing dysphagia, generalized hyperreflexia with rigidity and spasms of the abdomen and back causing opisthotonus; characteristic facial expression of a grimace; there may be seizures; a high or low temperature is a bad prognostic sign
- Differential Diagnosis
 1. Muscle spasms
 2. Amyotrophic lateral sclerosis (Lou Gehrig's disease)

3. Hypocalcemic tetany
 4. Phenothiazine reaction
 5. Strychnine poisoning
 6. Poliomyelitis
 7. Bacterial meningitis
 8. Narcotic withdrawal
- Physical Findings
 1. Muscle spasms aggravated by stimuli
 2. Muscle rigidity
 3. Increased oral secretions
 4. Respiratory compromise
 5. Patients are fully conscious
 - Diagnostic Tests/Findings: Diagnosis made clinically
 - Management/Treatment
 1. Medical referral
 2. Wounds need to be cleaned and debrided properly
 3. Supportive management—treatment of muscle spasms, intravenous fluids, respiratory support
 4. Minimize external stimuli (e.g., loud noise, bright light) to prevent aggravating muscle spasms
 5. Human tetanus immune globulin (TIG) given to prevent circulating toxin from binding to central nervous system sites
 6. Infection with tetanus does not confer immunity; patient should be reimmunized in convalescent period to prevent future infection
 7. Education—educate family regarding potential complications from tetanus including respiratory compromise, inability to speak

■ MALARIA

- Definition: An infectious disease primarily acquired via mosquito bite, characterized by high fever, rigors, sweats, and headache
- Etiology/Incidence
 1. Caused by *Plasmodium* spp. and transmitted primarily via mosquito bite by the *Anopheles* species; although transmission can be congenital; via transfusions or contaminated needles
 2. Infection by *Plasmodium falciparum* is most serious and potentially fatal
 3. Endemic in tropical areas worldwide; most cases in U.S. reported annually (approximately 1000) are acquired during foreign travel
- Signs and Symptoms
 1. History of recent travel to endemic area
 2. Classic paroxysmal symptoms—high fever, rigors, diaphoresis, and headache

3. Fever and other symptoms eventually become synchronized, and depending on the infecting species of *Plasmodium*, fever will occur every other or every third day
4. Associated symptoms—nausea, vomiting, diarrhea, arthralgia, cough, abdominal and back pain, pallor, jaundice
5. Multisystem involvement can develop with *Plasmodium falciparum* infection; may be fatal
 - a. Neurologic—seizures, signs of increased intracranial pressure, confusion, stupor, coma, and death
 - b. Pulmonary—coarse breath sounds, pulmonary edema
 - c. Renal—decreased urine output, oliguria, hematuria
 - d. Cardiovascular—absent peripheral pulses, hypotension
 - e. Gastrointestinal—diarrhea
 - f. Vascular collapse and shock

- Differential Diagnosis
 1. Influenza
 2. Rocky Mountain spotted fever
 3. Septicemia
- Physical Findings
 1. Early findings—listlessness, rigors, muscle weakness, pallor, jaundice, hepatosplenomegaly
 2. Findings seen in *Plasmodium falciparum* infection—neurologic deficits, pneumonia, elevated liver enzymes, renal failure
- Diagnostic Tests/Findings: Diagnosis depends on identification of parasite on stained blood films
- Management/Treatment
 1. Medical referral
 2. Drug therapy based on the infecting *Plasmodium* species, possible drug resistance, and severity of the disease; refer to most current American Academy of Pediatrics *Red Book* for varying treatment regimens
 3. Prevention best achieved through prophylactic therapy prior to travel to endemic areas; doses for children are calculated according to weight and should never exceed adult doses; each drug has its own side effects, check the *Red Book* for listing
 - a. Use chloroquine for areas in which there is no chloroquine-resistant malaria; begin 1 week before arrival and continue for 4 weeks after departure from area
 - b. In areas of chloroquine-resistant malaria, 3 drugs may be used: atovaquone-

- proguanil, doxycycline, and mefloquine; one must refer to the *Red Book* for appropriate ages and doses, as well as side effects
4. Control measures include protection against mosquitoes—by use of mosquito nets containing insecticide, insect repellents containing DEET, and protective clothing; contact the CDC Malaria Hotline at (770) 488-7788 (or online information) for country-specific risks, drug resistant strains in that country, and recommendations for travelers
 5. Education should include natural history of the illness and specific follow-up or rehabilitation needed after infection with *Plasmodium falciparum*
 6. Report cases to state health department

■ QUESTIONS

Select the best answer

1. Septicemia in the newborn period is most likely caused by which organism?
 - a. *Listeria monocytogenes*
 - b. *Haemophilus influenzae*
 - c. *Neisseria meningitidis*
 - d. *Streptococcus pneumoniae*
2. Signs and symptoms of bacterial sepsis in children beyond the neonatal period include:
 - a. Cough, fever, abdominal pain
 - b. Vesicular rash, pruritus, fever
 - c. Irritability, fever, lethargy
 - d. Abdominal pain, diarrhea, vomiting
3. Which of the following vaccines provides protection against a common type of sepsis/meningitis?
 - a. Smallpox vaccination
 - b. Hepatitis B vaccine
 - c. *Haemophilus influenzae* vaccine
 - d. Inactivated polio vaccine
4. Although relatively rare in the U.S., diphtheria can occur among under-immunized children. Which of the following clusters of signs, symptoms, and physical findings would suggest diphtheria in a child presenting with upper respiratory complaints?
 - a. Low-grade fever, sore throat, nasal discharge, and grayish-white pseudo-membrane in his/her throat
 - b. Abrupt onset of high fever, severe sore throat, nasal discharge, and grayish-white pseudo-membrane in his/her throat
 - c. Low-grade fever, abrupt onset of severe sore throat with difficulty swallowing and drooling
 - d. Abrupt onset of high fever, severe sore throat, with difficulty swallowing and drooling
5. Infants younger than six months of age with pertussis frequently require hospitalization to manage:
 - a. Fever, cough, dehydration
 - b. Coughing paroxysms, apnea, cyanosis, feeding difficulties
 - c. Coughing paroxysms, dehydration, renal failure
 - d. Seizures, fever, pneumonia
6. One of the most appropriate agents used to treat influenza A is:
 - a. Acyclovir
 - b. Oseltamivir
 - c. Erythromycin
 - d. Tetracycline
7. Which of the following symptoms are characteristic of rubella?
 - a. Vesicular, crusted lesions and high fever
 - b. Postauricular lymphadenopathy and low grade fever
 - c. Intense pruritus, usually in finger webs, buttocks, thighs, and ankles
 - d. Rough textured maculopapular rash that blanches with pressure
8. Although uncommon, a potential sequela of rubella may include:
 - a. Pneumonia and chronic otitis media
 - b. Arthritis, thrombocytopenia and encephalitis
 - c. Oophoritis and infertility
 - d. Arthritis, carditis, and neurological involvement
9. Rubeola is:
 - a. Preventable by active immunization
 - b. Caused by human herpesvirus 6
 - c. Treated with intravenous acyclovir
 - d. Not associated with severe complications (e.g., encephalitis, pneumonia)
10. You are examining a child who has fever, coryza, cough, conjunctivitis, malaise, and anorexia. During the oral examination, you observe red eruptions with white centers on the buccal mucosa. What are these eruptions called?
 - a. Pastia's spots
 - b. Rubeola spots
 - c. Koplik's spots
 - d. Strawberry spots

11. Which of the following best describes the treatment for roseola?
 - a. Acetaminophen or ibuprofen for fever, parental reassurance
 - b. Warm compresses for salivary gland swelling
 - c. Oral acyclovir, 20 mg/kg/dose, four times a day
 - d. Bed rest, saline gargles for sore throat
12. Fifth disease is usually:
 - a. Seen in age 5- to 14-year-old children
 - b. Transmitted via the deer tick
 - c. Treated with oral erythromycin
 - d. Characterized by prolonged coughing episodes
13. Which of the following statements is not true regarding the transmission of chickenpox?
 - a. Susceptible individuals can contract chickenpox from patients with varicella zoster (shingles)
 - b. Children with chickenpox are infectious only during the period of time when skin lesions are present
 - c. Children with chickenpox are no longer infectious once crusting of skin lesions has occurred
 - d. Varicella-zoster immune globulin (VZIG) should be administered to susceptible immunocompromised individuals who are exposed to a patient with varicella zoster infection
14. A child with chickenpox and temperature of 102°F should receive which medication for fever?
 - a. Aspirin
 - b. Amoxicillin
 - c. Acetaminophen
 - d. Acyclovir
15. The most appropriate agent for use in treating varicella zoster infection in an immunocompromised host is:
 - a. Ganciclovir
 - b. Acyclovir
 - c. Ceftriaxone
 - d. Chloramphenicol
16. Varicella zoster infection is most commonly associated with which of the following skin lesions?
 - a. Vesicle
 - b. Comedone
 - c. Nodule
 - d. Macule
17. Which of the following is not a complication of mumps?
 - a. Meningitis
 - b. Pneumonia
 - c. Oophoritis
 - d. Pancreatitis
18. What recommendation would you make to a parent whose son has been diagnosed with mumps and wants to know when he can return to child-care?
 - a. He can return once he becomes afebrile and can tolerate eating
 - b. He can return 9 days after onset of symptoms
 - c. He can return when he is well enough to participate in activities
 - d. He can return after a minimum of 5 days of antibiotic therapy
19. Which of the following are symptoms of cat scratch disease?
 - a. Joint pain, conjunctivitis, mild neck stiffness
 - b. Irritability, fever, hypotension
 - c. Fever, malaise, lymphadenopathy
 - d. Severe coughing, vomiting, anorexia
20. The following describes a characteristic rash associated with which disease? Initially erythematous and macular, becoming maculopapular and petechial. The rash first appears on the wrists and ankles, spreading proximally to the trunk. The palms and soles are often involved.
 - a. Lyme disease
 - b. Roseola
 - c. Rubella (measles)
 - d. Rocky Mountain spotted fever
21. A 10-year-old child manifests symptoms of fever, sore throat, and swollen lymph nodes. Spleen tip is palpable. Throat culture and monospot test results are negative. The next logical diagnostic test would involve:
 - a. Repeat throat culture
 - b. Chest radiograph
 - c. Bone marrow examination
 - d. Epstein-Barr virus titer
22. Which of the following factors is not associated with increased risk for infantile botulism?
 - a. Rural environments
 - b. Use of honey
 - c. Use of corn syrup
 - d. Farm families

23. Which of the following interventions would not be appropriate for a 6-month-old infant with a suspected diagnosis of infantile botulism?
- Stool and blood cultures
 - Immediate administration of equine antitoxin
 - Stool softeners
 - Supportive care
24. Which of the following are associated with paralytic poliomyelitis?
- Lacy, erythematous, pruritic rash
 - Respiratory compromise, speech disturbances, urinary incontinence
 - Abdominal swelling, lymphadenopathy, and jaundice
 - Nonspecific abdominal pain, nausea and vomiting
25. Muscle spasms associated with tetanus are aggravated by which of the following?
- Fever
 - Tetanus immunoglobulin
 - External stimuli
 - NSAID
26. Classic symptoms associated with malaria include:
- Low-grade fever, upper respiratory congestion, cough
 - Annular rash, conjunctivitis, headache, arthralgia
 - High fever, chills, rigors, sweats, headache
 - High fever, jaundice, lethargy, vomiting
27. Lyme disease is most closely associated with which of the following skin lesions?
- Erythema migrans
 - Nodule
 - Scale
 - Pustule
28. Many infectious diseases present with rashes along with general complaints of fever, malaise, and headaches. Which of the following clusters of symptoms would make you consider Lyme disease as a likely diagnosis?
- Fever, malaise, headache, arthralgia and well-circumscribed, erythematous, annular rash with central clearing
 - Fever, malaise, headache, transient bone pain, and generalized erythematous, maculopapular rash that began on the face and spread to trunk and extremities
 - Fever, malaise, anorexia, and confluent, erythematous, brownish maculopapular rash
 - Fever, malaise, anorexia, and erythematous rash beginning on wrists and ankles then spreading to the trunk
29. Which of the following would be included in patient education regarding Lyme disease?
- Educate caretakers regarding complications, including hypertension and renal calculi due to immobility
 - Avoid use of aspirin-containing products for fever control due to association with increased risk for Reye's Syndrome
 - Protective clothing and tick repellent should be worn in heavily wooded areas
 - Educate caretakers regarding natural history of the illness, and specific follow-up needed after infection with *Rickettsia rickettsii*

▣ ANSWERS

- | | |
|-------|-------|
| 1. a | 16. a |
| 2. c | 17. b |
| 3. c | 18. b |
| 4. a | 19. c |
| 5. b | 20. d |
| 6. b | 21. d |
| 7. b | 22. c |
| 8. b | 23. b |
| 9. a | 24. b |
| 10. c | 25. c |
| 11. a | 26. c |
| 12. a | 27. a |
| 13. b | 28. a |
| 14. c | 29. c |
| 15. b | |

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Musculoskeletal Disorders

Rachel Lyons

■ TORTICOLLIS (WRY NECK)

- Definition: Abnormal position of head and neck, due to unilateral contracture of sternocleidomastoid muscle that may be congenital (most common) or acquired, or due to atlanto axial rotary subluxation which is a displacement of C1 on C2
- Etiology/Incidence
 1. Cause not well defined, but may be due to:
 - a. Compartment syndrome due to soft tissue compression of neck at time of delivery
 - b. Occlusion of venous outflow of sternocleidomastoid muscle
 - c. Uterine crowding
 - d. Neurogenic myopathy from trauma or ischemia
 - e. Sternocleidomastoid muscle tumor
 2. Neurogenic causes rare
 3. Higher incidence in children with breech presentation and forceps delivery, but occurs with vaginal births or C-sections
 4. 20% have developmental dysplasia of the hip (DDH)
 5. Familial tendency (rare)
 6. 0.4% live births; males more frequently affected
- Signs and Symptoms
 1. Child's head tilted toward side of contracture
 2. Chin rotated away from contracted side (origin of muscle on mastoid process)
- Differential Diagnosis
 1. CNS tumors
 2. Syringomyelia
 3. Arnold-Chiari malformation
 4. Ocular dysfunctions
 5. Paroxysmal torticollis of infancy
 6. Klippel-Feil syndrome
- Physical Findings
 1. Contracture of one of sternocleidomastoid muscles
 2. Fusiform, firm mass or “tumor”
 - a. In body of contracted muscle
 - b. Palpable after 4 weeks of age, then recedes
 3. Plagiocephaly or asymmetry of face/skull development present with progressive deformity
- Diagnostic Tests/Findings
 1. Cervical radiograph
 - a. To rule out congenital spine abnormalities, e.g., hemivertebrae
 - b. Normal with muscular torticollis
 2. Other imaging (MRI or CT scan) not indicated—abnormalities not detected unless neurogenic pathology (rare) exists
 3. Hip ultrasound or x-ray, depending on age of child, to rule out DDH
- Management/Treatment
 1. Conservative measures—initial treatment
 - a. Stretching exercises—guided by physical therapist

- b. Encourage infant stretching by placing toys, mobiles, items of interest in infant's line of vision on the affected side
- 2. Surgery recommended when:
 - a. Defect persists beyond 1 year of age
 - b. Stretching exercises have been unsuccessful
 - c. Infant between 1 and 4 years of age; best outcome for surgical release

■ DEVELOPMENTAL DYSPLASIA OF THE HIP (DDH)

- Definition: Abnormal development dislocation of the hip(s), or ligamentous laxity which is congenital, but may not be recognized until ambulation occurs
- Etiology/Incidence
 - 1. Multifactorial etiology in an otherwise normal child
 - a. Intrauterine mechanical factors
 - (1) Breech presentation
 - (2) Infants with oligohydramnios
 - b. Genetic effects in primary acetabular dysplasia
 - (1) Various degrees of joint laxity
 - (2) 20% have positive family history
 - c. Increased incidence in first-born Caucasian infants; tight uterine muscles limit movement
 - 2. Left hip involved more commonly than right
 - 3. Greater incidence in girls (girls more often in breech position); 80% cases
 - 4. Incidence 1 to 1.5:1000 live births
- Signs and Symptoms
 - 1. Newborn
 - a. Instability without significant fixed deformity
 - b. Detected during newborn examination
 - 2. Sometimes not detected at birth
 - a. Untreated dislocation becomes fixed, with less instability, more limitation of movement (abduction)
 - b. Limp noticeable at onset of walking
 - c. Afebrile, painless, may have ligamentous laxity
- Differential Diagnosis
 - 1. Leg length discrepancy
 - 2. Innocent hip “click”—not associated with movement of femoral head
 - 3. Arthrogryposis
 - 4. Cerebral palsy
 - 5. Fracture of femur
- Physical Findings
 - 1. Galeazzi's sign—knee height comparison with infant in supine position with flexed hips/knees
 - a. Asymmetry evident in DDH
 - b. Shortening of the femoral segment limits abduction and full extension
 - c. Not helpful finding for detecting bilateral dislocation
 - 2. Limited abduction of affected hip in older child
 - 3. Indicators of hip instability in newborn—test each hip individually
 - a. Barlow's sign
 - (1) Positive when movement of femoral head can be felt as it slips out onto the posterior lip of acetabulum
 - (2) Not diagnostic, but indicates need for surveillance
 - b. Ortolani's sign (positive findings)
 - (1) Newborn period—sometimes a “click” or “clunk” is heard as femoral head enters or exits acetabulum
 - (2) After newborn period—“click” is less apparent and decreased abduction of flexed legs (at hip) is more significant
 - 4. Three degrees of hip dysplasia
 - a. Subluxation
 - (1) First degree, least severe
 - (2) Femoral head rests in acetabulum
 - (3) Can be dislocated partially by examination
 - b. Dislocatable
 - (1) Second degree
 - (2) Hip can be dislocated fully with manipulation, but is reducible
 - c. Dislocated hips
 - (1) Third degree, most severe
 - (2) Fixed dislocation
- Diagnostic Tests/Findings
 - 1. Physical examination is most reliable
 - 2. Radiographs
 - a. Not commonly used before 6 months of age
 - b. After 6 months of age assess femoral head/acetabulum relationship
 - 3. Ultrasonography
 - a. Before 6 months of age due to lack of ossification of proximal femoral heads
 - b. Assesses hip stability and acetabular development
- Management/Treatment
 - 1. Identification during newborn period essential for good prognosis
 - 2. Goal is to restore contact between femoral head and acetabulum

3. Subluxation in newborn
 - a. High incidence of spontaneous improvement in perinatal period
 - b. Observe and reexamine 3 to 4 weeks after birth
4. Dislocated hips
 - a. Treat at time of diagnosis
 - b. Before 6 months of age
 - (1) If unstable, stabilize with an abduction-flexion device (e.g., Pavlik harness); triple diapers not effective
 - (2) If device is ineffective, surgery is indicated
 - c. If diagnosis made after 6 months of age
 - (1) Child usually too large and strong to tolerate brace (failure rate 50% after 6 months old)
 - (2) Surgical reduction indicated

■ TALIPES EQUINOVARUS CONGENITA (CLUBFOOT)

- Definition: Common foot deformity that involves the foot and entire lower leg; classified into three groups, (1) congenital, (2) teratologic, and (3) positional
- Etiology/Incidence
 1. Cause unknown, possible inheritance factors or neuromuscular cause
 2. Congenital form—about 75% of all cases
 - a. 50% cases are bilateral
 - b. 1:1000 live births; more common in males
 3. Teratologic form—associated with neuromuscular disorders, e.g., spina bifida, arthrogryposis, multiple congenital, or a syndrome complex
 4. Positional form—normal foot that has been held in deformed position in utero
- Signs and Symptoms
 1. Congenital form
 - a. Absence of other congenital abnormalities
 - b. Variable rigidity of the foot
 - c. Mild calf atrophy
 - d. Mild hypoplasia of the tibia, fibula, and bones of the foot
 2. Older child—calf and foot atrophy are more obvious than in infant, regardless of how well corrected
- Differential Diagnosis
 1. Internal femoral torsion
 2. Internal tibial torsion
 3. Metatarsus adductus
 4. Neuromuscular disorders

- Physical Findings
 1. Small foot with limited dorsiflexion; usually obvious at birth
 2. Combination of deformities
 - a. Results in 90 degree rotation of forefoot in all planes
 - b. Leg and foot resemble shape of club
 3. Deep crease on medial border of foot
 4. Calf muscles thin and atrophic (more obvious in older child)
- Diagnostic Tests/Findings: Radiographs
 1. Rule out other conditions
- Management/Treatment
 1. Refer to orthopedist
 2. Serial casting with manipulation begins at birth; usually 3 to 6 months
 3. If further corrections required, surgery to lengthen tendon Achilles indicated

■ METATARSUS ADDUCTUS/METATARSUS VARUS

- Definition: Congenital medial deviation of the forefoot on the hindfoot
- Etiology/Incidence
 1. Uncertain etiology; often associated with abnormal intrauterine positioning
 2. Most common congenital foot deformity
 3. 1:1000 live births
 4. 10% of children with metatarsus also have developmental dysplasia of the hip
- Signs and Symptoms
 1. Toeing-in; “pigeon-toed” gait in older child
 2. Usually not painful
- Differential Diagnosis
 1. Internal femoral torsion
 2. Internal tibial torsion
 3. Equinovarus
- Physical Findings
 1. Adductus—forefoot adducted only; full ROM
 2. Varus—forefoot adducted and inverted; limited ROM
 3. Ankle joint has normal dorsiflexion and plantar flexion
- Diagnostic Tests/Findings
 1. Physical examination—usually sufficient to establish treatment plan

2. Radiographs
 - a. Usually unnecessary
 - b. Used when:
 - (1) Underlying congenital anomalies are suspected
 - (2) Foot is unusually rigid
 - (3) Failure of spontaneous resolution with growth
- Management/Treatment
 1. Supple deformity (adductus)
 - a. Parents stretch forefoot in all planes of motion with each diaper change for 4 to 6 months
 - b. Observe and follow-up
 2. Rigid deformity (varus)
 - a. Serial casting or bracing in the first year of life
 - b. Then straight-laced/outflare shoes fitted for daytime until no chance of recurrence
 - c. Surgical intervention required for child older than 4 years if significant residual metatarsus adductus persists

■ TIBIAL TORSION (INTERNAL)

- Definition: Abnormal bowing (internal or external rotation) of the tibia
- Etiology/Incidence
 1. Combination of genetic factors and intrauterine position
 2. Usually not pathological
 3. Internal tibial torsion—12% at birth; usually resolves by 2 years
 4. External tibial torsion—develops after birth or by 2 years
- Signs and Symptoms
 1. Toeing-in appearance of child's legs when walking/running
 2. Rarely painful
 3. Tripping and falling may be noticed
- Differential Diagnosis
 1. Metatarsus adductus
 2. Femoral neck anteversion
 3. Femoral neck retroversion
 4. Neuromuscular disorders
 5. Equinovarus
- Physical Findings
 1. No obvious deformity
 2. Full ROM
 3. Internal rotation of affected leg, flat feet, increased lumbar lordosis
- Diagnostic Tests/Findings
 1. Observation
 2. Angle measurement
 - a. Between foot and thigh
 - b. With ankle and knee position at 90 degree angle
 - c. With child lying in prone position
- Management/Treatment
 1. Reassure parents that most children have spontaneous correction with growth and need no treatment
 2. Recommend supine sleeping position

■ GENU VARUM (BOWLEG)

- Definition: Lateral bowing of the tibia
- Etiology/Incidence
 1. Joint laxity may contribute to deformity
 2. Considered normal until 36 months
 3. May be related to intrauterine position
- Signs and Symptoms
 1. Parental concern common regarding appearance of legs
 2. Physiologic bowing of up to 20 degrees is normal in children until 18 to 24 months of age
 3. Bowing does not generally increase after 16 months and usually resolves by age 24 months
- Differential Diagnosis
 1. Hypophosphatemic rickets
 2. Blount disease (tibia vara)
 3. Injury to medial proximal epiphysis of tibia
 4. Osteogenesis imperfecta
 5. Achondroplasia and other skeletal dysplasias
 6. Extreme physiologic bowing
 7. Neoplasms
- Physical Findings
 1. With child standing
 - a. Clinically present—space between knees is greater than 5 cm or 2 in. with apposition of medial malleoli (relative, not absolute measurement)
 - b. Resolution usually occurs without treatment
 2. Full range of motion throughout lower extremities
- Diagnostic Tests/Findings: Radiographs used for extreme and/or unilateral bowing
- Management/Treatment
 1. Observation to verify resolution
 2. Avoid unnecessary treatment of mild to moderate bowing

3. Further evaluation with radiographs necessary if:
 - a. Genu varum is present after 2 years of age
 - b. Progressive after 1 year of age
 - c. Unilateral involvement
 - d. Appears to be severe
 - e. Occurs in a high risk group, e.g., obese; African-American children with early ambulation

■ GENU VALGUM (KNOCK-KNEE)

- Definition: Deformity in which the knees are abnormally close and space between the ankles is increased
- Etiology/Incidence
 1. A natural shifting occurs from varum to valgus between 30 to 60 months
 2. Normal alignment about 8 years of age
 3. Underlying bone disease can cause marked bilateral valgum
- Signs and Symptoms
 1. Parental concern common regarding appearance of legs
 2. Associated with pronation; more common in overweight children
- Differential Diagnosis
 1. Injury to lateral proximal tibial epiphysis causes unilateral valgum
 2. Hypophosphatemic rickets
 3. Pseudoachondroplasia
- Physical Findings
 1. Knees are together and distance between medial malleoli (ankles) is greater than 3 in. (7.5 cm) when standing (relative, not absolute measurement)
 2. Full range of motion
 3. No pain
 4. Child may walk and run awkwardly
 5. Normal knee alignment usually occurs before 8 years of age
- Diagnostic Tests/Findings: Radiographs used for extreme valgum; standing AP and lateral views
- Management
 1. Observation to verify resolution
 2. Further evaluation with radiographs necessary if:
 - a. Genu valgum is present after 7 years of age
 - b. Unilateral involvement

■ TRANSIENT (TOXIC) SYNOVITIS OF THE HIP

- Definition: Self-limiting inflammation of hip joint
- Etiology/Incidence
 1. Etiology uncertain; possible immune or viral process
 2. Most common cause of irritable hip
 3. Males affected twice as often as females
 4. Occurs most often in 3- to 12-year-olds, but also as young as 3 months
 5. Both hips equally affected
- Signs and Symptoms
 1. Painful limp, or hip (groin) pain with acute or insidious onset, usually unilateral and is often preceded by an upper respiratory infection
 2. Afebrile or low-grade temperature
- Differential Diagnosis
 1. Septic arthritis
 2. Osteomyelitis
 3. Legg-Calvé-Perthes disease
 4. Juvenile monoarthritis
 5. Rheumatoid arthritis
 6. Slipped capital femoral epiphysis
- Physical Findings
 1. Range of motion of hip causes spasm and pain, particularly with internal rotation
 2. No obvious signs on inspection or with palpation
- Diagnostic Tests/Findings
 1. Radiography normal or slightly widened joint space medially
 2. Ultrasonography is useful in determining effusion
 3. Normal or slightly elevated WBC
 4. Joint fluid aspiration—normal
- Management/Treatment
 1. Hospitalize child
 - a. If high fever or severe symptoms are present
 - b. To differentiate between transient synovitis and septic arthritis
 2. Analgesics (ibuprofen every 6 to 8 hours) \times 5 days
 3. Bedrest/nonweight bearing
 4. Benign, self-limiting illness

■ LEGG-CALVÉ-PERTHES DISEASE (LCPD)

- Definition: Aseptic or avascular necrosis of the femoral head
- Etiology/Incidence
 1. Unknown etiology; possibly due to vascular interruption, anthropometric abnormalities, transient synovitis, or nutritional deficits
 2. Generally, slightly shorter stature/delayed bone age compared to peers
 3. Most common in Caucasian boys, ages 4 to 9 years
 4. 15% of cases are bilateral
- Signs and Symptoms
 1. Insidious onset of limp with knee pain that is activity-related and resolves with rest
 2. Pain also in groin or lateral hip
 3. Pain less acute and severe than transient synovitis or septic arthritis
 4. Afebrile
- Differential Diagnosis
 1. Transient synovitis
 2. Septic arthritis
 3. Hematogenous osteomyelitis
 4. Various types of hemoglobinopathy
 5. Gaucher's disease
 6. Hypothyroidism
 7. Epiphyseal dysplasias
- Physical Findings
 1. Limited passive internal rotation and abduction of hip joint
 2. May be resisted by mild spasm or guarding
 3. Hip flexion contracture and leg muscle atrophy in long-standing cases
- Diagnostic Tests/Findings
 1. Radiograph studies
 - a. Show disease progression and sphericity of femoral head
 - b. Used initially for definitive diagnosis
 - c. Subsequently used to assess reparative process
 2. Other laboratory studies not indicated
- Management/Treatment
 1. Goal—to restore range of motion while maintaining femoral head within acetabulum
 2. Observation only if full ROM preserved
 - a. Children < 8 years of age
 - b. Involvement of < one half the femoral head
- 3. Aggressive treatment
 - a. Indicated when > one half femoral head involved and in children older than 8 years
 - b. Use of orthosis—rarely used today
 - c. Surgical treatment
 - (1) Femoral osteotomy
 - (2) Shelf arthroplasty
- 4. Patient/family education—inform family that LCPD lasts 1 to 3 years and is potentially serious if not treated properly

■ GROWING PAINS

- Definition: A controversial diagnosis of exclusion for (usually intermittent) lower extremity pain
- Etiology/Incidence
 1. Onset at 3 to 5 years of age, or more commonly, 8 to 12 years of age
 2. Related factors
 - a. Rapid growth
 - b. Puberty
 - c. Fibrositis
 - d. Weather
 - e. Psychological factors
- Signs and Symptoms: Pain/ache localized to lower extremities; usually intermittent and sometimes nocturnal
- Differential Diagnosis
 1. Trauma
 2. Infection
 3. Hematologic causes—sickle cell, hemophilia
 4. Slipped capital femoral epiphysis
 5. Osgood-Schlatter
 6. Osteochondritis dissecans
- Physical Findings (usually none)
 1. No history of traumatic insult
 2. No loss of ambulation or mobility
 3. No systemic changes
 4. No edema or erythema
 5. Full range of motion
- Diagnostic Tests/Findings
 1. All laboratory studies normal—CBC with differential, ESR, CRP
 2. Radiograph of affected area normal
- Management/Treatment
 1. Prescribe anti-inflammatory medication
 2. Massage and heating pad to area
 3. Rest during painful episodes, activity as tolerated

■ OSGOOD-SCHLATTER DISEASE

- Definition: Inflammation of tibial tubercle from repetitive stresses in athletes with immature skeletal development
- Etiology/Incidence
 1. Tiny stress fractures in apophysis likely etiology
 2. Associated with a rapid growth spurt
 3. Occurs 10 to 15 years of age when immature cartilage susceptible to repeated trauma
- Signs and Symptoms: Pain and tenderness localized to tibial tubercle in acute phase
- Differential Diagnosis
 1. Osteomyelitis
 2. Osteosarcoma
 3. Patellar tendonitis
- Physical Findings
 1. Point tenderness over tibial tubercle
 2. Prominence/enlargement of tibial tubercle compared with unaffected side after acute phase
 3. 50% have bilateral involvement
- Diagnostic Tests/Findings
 1. Diagnosis accurate by clinical examination
 2. CT and MRI scans rarely indicated
 3. Radiographs
 - a. Used to rule out presence of more serious bone pathology
 - b. Lateral view
 - (1) May demonstrate ossicle between patellar tendon and tibial tubercle
 - (2) For pain which persists after skeletal maturity
 - c. Rarely useful at follow-up
- Management/Treatment
 1. Self-limiting condition
 2. Conservative and largely symptomatic treatment
 3. Pain resolves with full ossification of tibial tubercle and closure of apophysis
 4. Activity limitations
 - a. Complete avoidance of sports activities not recommended
 - b. Limit activity to control pain at tibial tubercle
 - c. Stretching exercises before activity, icing helpful afterwards
 - d. Use knee immobilizer
 - (1) Pain relief—briefly to avoid muscle atrophy

- (2) In combination with thigh muscle strengthening
5. Corticosteroid injections—not recommended; may aggravate apophysitis
6. Surgery
 - a. Indicated if tubercle pain persists after skeletal maturity
 - b. Excision of ossicle may ameliorate symptoms

■ SCOLIOSIS (IDIOPATHIC)

- Definition: Lateral curvature of spine
- Etiology/Incidence
 1. Multifactorial etiology
 2. 70% cases idiopathic
 3. Most common occurrence immediately before or during adolescent growth spurt
 4. Female to male ratio of 8:1
 5. Mild curves occur equally between the sexes
 6. Positive family history in about 70% of cases
- Signs and Symptoms (usually asymptomatic)
 1. Infancy to school age—parents may notice alteration in back contour
 2. Adolescence—more likely detected on routine screening
 3. Rarely painful
- Differential Diagnosis
 1. Hip disease
 2. Transient synovitis
 3. Legg-Calvé-Perthes disease
 4. Slipped capital femoral epiphysis
 5. Leg length discrepancy
- Physical Findings: Inspection in standing position
 1. Asymmetry of shoulder height
 2. Uneven hip level
 3. Waistline uneven
 4. Thoracic spinal curve (usually right sided)
 5. Rib asymmetry
 6. Unequal arm length
 7. Asymmetry of scapulae
- Diagnostic Tests/Findings
 1. Adam's forward-bending test
 - a. Child bends forward 90 degrees or more, keeping knees straight, feet forward, dropping head with arms hanging downward, elbows extended
 - b. Observed from caudal aspect to detect abnormal prominence of thoracic ribs
 2. Radiographs evaluate degree of deformity

- Management/Treatment
 1. If pain occurs, further evaluation required
 2. Treatment mode depends on severity of curve and child's age
 - a. Curves of 25 degrees
 - (1) No further evaluation/treatment if child skeletally mature
 - (2) Follow-up for possible progression if child is still growing
 - (3) Bracing treatment
 - b. Curves of 40 to 50 degrees
 - (1) Likely to increase if curve > 50 degrees even after growth complete
 - (2) Surgery likely for thoracic curve > 50 degrees or lumbar curve > 40 degrees
 3. Clinical pulmonary restriction may occur with thoracic curves > 75 degrees

■ SPORTS INJURIES

- Definition: Musculoskeletal injuries occurring as a result of participation in athletic activities; most common include sprains/strains, fractures, or overuse injuries
 1. Head and neck injuries
 - a. Common in football and ice hockey
 - b. Generally not severe
 - c. Include brachial plexus injuries
 - d. Concussions
 2. Back injuries—low back pain caused by:
 - a. Muscle strain
 - b. Spondylolysis/spondylolysis defect—overuse from repetitive hyperextension of back as in gymnastics, most likely in lower lumbar area causes fracture
 - c. Spondylolisthesis—forward vertebral slippage after spondylolysis
 3. Upper extremity injuries
 - a. Anterior shoulder dislocation, shoulder separation
 - b. Overuse injuries
 - (1) Impingement syndrome—“pitcher’s shoulder,” “swimmer’s shoulder,” or “tennis shoulder”
 - (2) Lateral epicondylitis—“tennis elbow”
 4. Lower extremity injuries
 - a. Sprains (common in adolescents but not in children)
 - (1) Medial collateral ligament sprain
 - (2) Anterior cruciate sprain and tear
 - b. Overuse injuries
 - (1) Iliac apophysitis
 - (2) Femoral stress fracture
 - (3) Chondromalacia (chronic patellar pain “runner’s knee”)
- Etiology/Incidence
 1. Leading causes—trauma, improper training
 2. Contributing factors include fatigue and improper nutrition
 3. 20-million children participate in organized athletics
 4. One out of fourteen adolescents treated for athletic injury
 5. Highest frequency in adolescent boys; football, soccer, and wrestling cause most injuries
- Signs and Symptoms
 1. Fracture
 - a. Edema
 - b. Erythema
 - c. Ecchymosis
 - d. Pain
 - e. Obvious angulation
 - f. Bony point tenderness
 2. Sprain
 - a. Various degrees of pain
 - b. Swelling
 - c. Difficulty weight bearing
 - d. Detectable joint laxity
 - e. Decreased ROM
 3. Overuse—various degrees of pain with or without activity limitations
- Differential Diagnosis: Possible underlying disease process of metabolic, neoplastic, or infectious origins
- Physical Findings
 1. Fracture
 - a. Decreased ROM
 - b. Pain
 - c. Obvious deformity
 - d. Swelling
 - e. Evidence of injury seen on radiographs at site of injury
 - f. Localized tenderness
 2. Sprains—use subjective grading
 - a. Grade I
 - (1) Few fibers torn within ligament; does not compromise ligament’s strength
 - (2) Minimal pain and swelling
 - (3) Full ROM
 - (4) No increase in joint laxity
 - b. Grade II
 - (1) Tears portion of the ligament
 - (2) Clinically significant pain and swelling
 - (3) Impairment of ROM
 - (4) Detectable increase in joint laxity
 - c. Grade III
 - (1) Complete tear of ligament
 - (2) Marked laxity evident when ligament is stressed

- Diagnostic Tests/Findings
 1. Physical examination and accurate history to evaluate injury
 2. Radiographs
 - a. Injured area and contralateral area
 - b. View open growth plate
 - c. Rule out fracture or tumor
 3. MRI—utilized to evaluate torn ligaments or damage to cartilage
 4. Ultrasonography—utilized for visualization of effusion
- Management/Treatment
 1. Encourage sports physical examinations prior to participation in athletic events to identify conditions that may interfere/worsen with athletic participation
 2. Fractures—immobilization, pain management, ROM exercise
 3. Sprain/strain
 - a. Minimize hematoma and swelling with rest, ice, compression, elevation (RICE)
 - b. ROM exercise
 - c. Grade III sprains may require surgery
 4. Overuse injuries
 - a. Usually respond to conservative treatment, rest, ice, and gradual return to athletic activities
 - b. Nonsteroidal anti-inflammatory drugs prescribed to decrease inflammation and pain
 5. Refer to orthopedic practitioner
 - a. Fractures that involve the growth plates are open or displaced
 - b. If sprain/strain or overuse injuries not resolving with conservative treatment measures
 6. Child can return to athletic activity based on functional evaluation of actions required during the activity

■ SLIPPED CAPITAL FEMORAL EPIPHYSIS

- Definition: Spontaneous dislocation of femoral head (capital epiphysis) posteriorly and usually medially through the physis that typically occurs through adolescent growth spurt
- Etiology/Incidence
 1. Etiology unknown—thought to be precipitated by the interplay of hormones related to puberty
 2. Generally occurs without severe, sudden force or trauma; sometimes related to trauma
 3. Usually occurs during growth spurt (ages 10 to 17) and before menarche in girls

4. Annual incidence is 2 to 13 per 100,000
 5. More common in males and African-Americans
 6. 20% present with bilateral involvement
 7. Incidence greater among obese adolescents with sedentary lifestyles
- Signs and Symptoms
 1. Varies with acuity of the process
 2. Most children have limp (if greater than 3 weeks, considered chronic)
 3. Varying degrees of aching or pain (in groin, often referred to thigh/knee)
 4. Some have acute, severe pain, and inability to walk or move hip
 - Differential Diagnosis
 1. Knee complaint with no obvious cause
 2. Trauma
 3. Septic arthritis
 4. Transient (toxic) synovitis
 5. Juvenile arthritis
 6. Legg-Calvé-Perthes disease
 - Physical Findings
 1. Unable to properly flex hip as femur abducts/rotates externally
 2. May observe limb shortening, resulting from proximal displacement of metaphysis
 3. Loss of internal rotation with hip flexed to 90 degrees
 - Diagnostic Tests/Findings
 1. Accurate history combined with knowledge of etiological factors
 2. Radiographs
 - a. Confirms diagnosis
 - b. Shows degree of slipping between femoral head and neck
 3. Laboratory studies
 - a. Depend on findings from physical examination and history
 - b. Done to rule out associated causes of infection or inflammation
 - Management/Treatment
 1. Immediate referral to orthopedic practitioner
 2. Treatment goal is to prevent further slippage, promote closure of the physis, and avoid chondrolysis or osteonecrosis
 3. No ambulation is allowed on acute/unstable SCFE
 4. Surgery—in situ pin fixation to stabilize upper femur and cause growth plate to close
 5. Monitor other hip for same problem

■ JUVENILE IDIOPATHIC ARTHRITIS

- Definition: Chronic, autoimmune idiopathic arthritis characterized by presence of chronic synovial inflammation with associated swelling, pain, heat, and/or limited ROM
- Etiology/Incidence
 1. Cause unknown; possible etiological factors include infections, autoimmunity, genetic predisposition, or stress and trauma
 2. Most common autoimmune inflammatory disease of childhood
 3. Mean incidence 6-19:100,000; estimated 65,000 to 70,000 children in U.S. affected
 4. Mean age of onset 1 to 3 years; rarely before 6 months, and again between 8 to 10 years
 5. Females affected twice as often as males; pauciarticular and polyarticular disease occur more frequently in girls, while both sexes are affected with equal frequency in systemic-onset disease
- Signs and Symptoms
 1. Range of severity of disease
 - a. May be mild in one joint with no symptoms
 - b. May have severe disease in many joints with fever, rash, lymphadenopathy, and organomegaly
 2. Signs of joint inflammation
 - a. Swelling with heat
 - b. Redness
 - c. Pain
 - d. Limited ROM
 3. May also exhibit
 - a. Morning stiffness, limp, refusal to walk
 - b. Irritability, fatigue
 4. Hallmark of systemic disease is high spiking fever with rash
- Differential Diagnosis
 1. Hip disease
 2. Transient synovitis
 3. Legg-Calvé-Perthes disease
 4. Slipped capital femoral epiphysis
 5. Leukemia
 6. Other rheumatic diseases, e.g., Kawasaki, systemic lupus erythematosus (SLE), Lyme, spondyloarthropathy
- Physical Findings
 1. Diagnostic criteria
 - a. Age of onset < 16 years
 - b. Joint involvement
 - (1) Arthritis (swelling/effusion) in one or more joints, or
 - (2) Presence of 2 or more of these signs:
 - (a) Range of motion limitation
 - (b) Tenderness
 - (c) Pain with movement
 - (d) Increased heat
 - c. Duration of disease 6 weeks or longer
 - d. Further classified by onset type during first 6 months
 - e. Exclusion of other forms of juvenile arthritis
- 2. Polyarthritis (polyarticular disease)
 - a. 40% to 50% of all cases
 - b. 5 or more inflamed joints
 - c. May be acute or gradual onset
 - d. Symmetric pattern
 - e. Commonly affects large joints, e.g., knees, wrists, elbows, ankles
 - f. May not complain of pain
- 3. Oligoarthritis (pauciarticular disease)
 - a. 50% of cases
 - b. 4 or less joints affected (usually larger joints such as knees, ankles, and wrists)
 - c. Gradual onset
 - d. Can be painless
 - e. Hip monoarticular arthritis unusual
 - f. Usually no systemic signs, except for chronic uveitis
 - (1) Early onset (< 5 years of age)
 - (a) Asymmetric involvement
 - (b) Large joints commonly affected
 - (c) Hips and sacroiliac (SI) joint spared
 - (d) Systemic symptoms usually not present
 - (2) Late onset
 - (a) Asymmetric involvement
 - (b) Hips and SI joint involvement present
 - (c) Chronic involvement can result in atrophy of extensor muscles in the thigh, tight hamstring ligaments, and knee flexion contractures
- 4. Systemic
 - a. 10% to 20% of cases
 - b. Occurs in late childhood > 8 years of age
 - c. Systemic onset may precede arthritis appearance by weeks, months, or years
 - d. High, daily intermittent spiking fevers is hallmark symptom
 - (1) Temperature elevations occur once or twice/day
 - (2) To 39°C (102°F) or higher with quick return to baseline temperature or lower
 - e. Linear evanescent rash present
 - (1) Salmon-colored nonpruritic macular lesions

- (2) Commonly on trunk and proximal extremities
- (3) Most characteristic feature is transient nature
- f. Painful multiple joint involvement
- g. Associated findings
 - (1) Hepatosplenomegaly
 - (2) Lymphadenopathy
 - (3) Visceral disease, e.g., pericarditis, hepatitis
 - (4) Pulmonary involvement
 - (5) CNS
- h. 50% of cases have chronic, destructive arthritis
- 5. Associated problems
 - a. Periarticular soft tissue edema
 - b. Intra-articular effusion
 - c. Hypertrophy of synovial membrane
- 6. Synovitis—painful inflammation of synovial membrane, with fluctuating swelling
 - a. May develop insidiously, existing months to years without joint destruction
 - b. May cause joint damage in relatively short time
- Diagnostic Tests/Findings
 1. No specific laboratory studies, however, abnormalities may be found
 2. Human leukocyte antigen (HLA) B27
 3. CBC—characteristics of chronic anemia of inflammation
 - a. Moderately severe anemia; hemoglobin between 7 and 10 g/dL
 - b. Leukocytosis
 4. Erythrocyte sedimentation rate (ESR)
 - a. ESR is always elevated in children with systemic JRA
 - b. Usually elevated in those with polyarticular disease
 - c. Often within the reference range in those with pauciarticular disease
 - d. Elevated, ESR may be used to monitor success of medical treatment
 5. Alanine aminotransferase (ALT) test—obtain ALT levels to exclude the possibility of hepatitis (viral or autoimmune) prior to initiating treatment with NSAIDs, which can cause hepatotoxicity
 6. Urinalysis with microscopic examination—perform a urinalysis to exclude the possibility of infection (as a trigger of JRA or transient postinfectious arthritis) and nephritis (observed in individuals with SLE)
 7. Synovial fluid WBC count—moderately elevated 10,000/mm³ to 20,000/mm³
 8. Rheumatoid factor present in:
 - a. Approximately 15% to 20% of cases
 - b. Child with later onset or older child
 - c. Child with prominent symmetric polyarthritis with:
 - (1) Involvement of small joints
 - (2) Subcutaneous rheumatoid nodules
 - (3) Articular erosions
 - (4) Poor functional outcome
- 9. Antinuclear antibodies (ANA)—seropositivity for antibodies
 - a. Present in about 25% of cases
 - b. Presence correlated significantly with development of chronic uveitis
 - c. Less commonly found in older boys or in systemic disease
 - d. Valuable diagnostic measure for JRA
 - (1) Usually not positive in other childhood illnesses
 - (2) Positive in SLE, scleroderma, transient acute viral disease
- 10. Radiographs
 - a. Early changes
 - (1) Soft tissue swelling
 - (2) Juxta-articular osteoporosis
 - (3) Periosteal new bone apposition
 - b. Development of ossification centers may be age accelerated
 - c. Stunting of bone growth secondary to premature epiphyseal closure
 - d. Cervical spine disease characteristic feature
 - e. Joint disease may be better evaluated with MRI, CT, bone scans
- Management/Treatment
 1. Goal is to control clinical manifestations and prevent/minimize deformity
 2. Suppress inflammation and fever
 - a. Use NSAID (ibuprofen, naproxen) for most children
 - b. In severe, progressive disease resistant to therapy
 - (1) Methotrexate
 - (a) Most successful and safe drug
 - (b) 10 to 15 mg/m²/once a week
 - (c) No oncogenic potential or risk of sterility
 - (2) Hydroxychloroquine—useful adjunctive agent
 - (a) Retinopathy possible adverse reaction
 - (b) Low dose, e.g., 5 mg/kg/day
 - (c) Frequent ophthalmologic examinations required
 - (3) Gold salts—given IM or as oral compound
 - (a) Toxicities are hematologic, renal, hepatic
 - (b) Must be constantly monitored during treatment

- (4) Glucocorticoid drugs
 - (a) Indicated for resistant or life-threatening disease
 - (b) Ophthalmic administration for chronic uveitis
 - (c) Toxicities, e.g., Cushing's syndrome and growth retardation
- (5) Sulfasalazine—sulfa drug + salicylate
 - (a) Indicated for JIA
 - (b) May take 6 weeks to work
- (6) Biologic agents
 - (a) Synthetic proteins that block high levels of inflammatory proteins
 - (b) Indicated for moderate to severe arthritis that has not responded well to other therapies
3. Maintenance of function and prevention of deformity
 - a. Prescriptions for physical and occupational therapies
 - b. Balanced program of rest and activity
 - c. Selective splinting
 - d. Encourage normal play/activity
 - e. Avoid high levels of stress on inflamed weight-bearing joints
4. Counsel parents about course of chronic disease with exacerbations
5. Refer to pediatric rheumatologist

■ SYSTEMIC LUPUS ERYTHEMATOSUS

- Definition: Multisystem autoimmune disorder that is characterized by widespread inflammatory involvement of connective tissues with immune complex vasculitis
- Etiology/Incidence
 1. Unknown, but many factors implicated
 - a. Excessive sun exposure
 - b. Drug reaction
 - c. Infection
 - d. Hereditary
 - e. Immunogenetic
 2. Can develop at any age
 - a. Usually after 5 years of age
 - b. More common during adolescent years in females
 3. Females affected 8 times more often after 5 years of age
- Signs and Symptoms
 1. Fever
 2. Malaise
 3. Weight loss
 4. Malar facial rash
 5. Arthralgias
- Differential Diagnosis
 1. Juvenile rheumatoid arthritis (JRA)
 2. Other forms of acute glomerulonephritis
 3. Hemolytic anemia
 4. Leukemia
 5. Allergic or contact dermatitis
 6. Idiopathic seizure disorder
 7. Mononucleosis
 8. Acute rheumatic fever with carditis
 9. Septicemia
 10. Toxic exposure
- Physical Findings
 1. Onset is usually acute—three quarters of children usually diagnosed within 6 months of symptoms
 2. Diagnosis delayed for others by 4 to 5 years
 3. Early diagnostic suspicion based on:
 - a. Episodic, multisystem constellation of clinical disease
 - b. Associated with persistent antinuclear antibody (ANA) seropositivity
 4. Severity of manifestations variable
 - a. Rapidly fatal illness
 - b. Insidious chronic disability with multi-system exacerbation
 5. Each exacerbation of disease tends to mimic previous episodes
 6. Rash
 - a. Characteristic of acute onset or exacerbation
 - b. Malar erythematous
 - c. Butterfly distribution across bridge of nose and over each cheek
 - d. Discoid rash over sun-exposed areas
 7. Arthritis
 - a. Affects majority of children
 - b. Involves small joints
 - c. Transient and migratory
 - d. Never erosive
 - e. Possible avascular necrosis of bone in 25% children
 8. Pericarditis is most common manifestation of cardiac involvement—murmurs associated with endocarditis
 9. Central and peripheral nervous system manifestations
 - a. Recurrent headaches
 - b. Seizures
 - c. Chorea
 - d. Frank psychosis
 10. Kidney involvement in all children
 - a. Proteinuria (> 500 mg/d) or evidence of nephritis in urinalysis
 - b. Hypertension

- Diagnostic Tests/Findings
 1. Leukopenia—otherwise unexplained, common at onset
 2. ANA, anti-double-stranded DNA, anti-Smith antibody, lupus anticoagulant, and antiphospholipid antibody panel—positive in most children
 3. Coombs test—often positive
 4. Rheumatoid factor (RF) and other antitissue antibodies—often positive
 5. ECG and chest x-ray
 6. MRI/CT of brain
 7. Renal ultrasound
 8. Tissue biopsy to confirm diagnosis and distinguish disease severity
- Management/Treatment
 1. Long-term supportive care
 - a. Maintain adequate nutrition
 - b. Maintain fluid and electrolyte balance
 - c. Early recognition and treatment of infections
 - d. Control of hypertension
 - e. Exercise is important to maintain bone density
 2. Anti-inflammatory drugs useful for minor manifestations, e.g., myalgia and arthralgia
 3. Counsel parents and child about chronic nature of disease with repeated exacerbations, remissions often prolonged over many years

■ OSTEOMYELITIS

- Definition: Inflammation of bone caused by a pyogenic organism
- Etiology/Incidence
 1. Causative agents
 - a. In all age groups, *Staphylococcus aureus*
 - b. Consider also *Streptococcus pneumoniae* and *Haemophilus influenzae*
 2. Peak ages—infancy (less than 1 year) and preadolescence (9 to 11 years)
 3. More frequent in males
- Signs and Symptoms
 1. May appear well
 2. Systemic involvement ranging from malaise to shock
 3. Neonates usually afebrile, swollen or motionless limb early sign
 4. Earliest symptom in child may be refusal to bear weight or flexion of hip in comfortable position
- Differential Diagnosis
 1. Neoplasm
 2. Contusion

3. Nondisplaced fracture
4. Sickle cell crisis
5. JIA
6. Transient synovitis

- Physical Findings
 1. Early signs
 - a. Fever
 - b. Local bone tenderness
 2. If subperiosteal or soft tissue abscess develops, fluctuant mass present
- Diagnostic Tests/Findings
 1. WBC, ESR, and CRP—elevated, but not diagnostic
 2. Radiographs—at earliest stage may show soft tissue swelling
 3. Bone scan
 - a. May be initially normal
 - b. Repeated after 48 hours, may show cold/photopenic areas (indicating avascular sites)
 4. Aspiration—always indicated to identify pathogen
- Management/Treatment
 1. Refer to physician
 2. Delivery of systemic antibiotic
 - a. Broad spectrum antibiotic initially
 - (1) Most effective against isolated organism
 - (2) Least toxic antibiotic
 - (3) Used for 4 to 6 weeks
 3. Surgery reserved for:
 - a. Child with systemic illness
 - b. Worsening symptoms under medical treatment
 - c. Abscess present

■ DUCHENNE MUSCULAR DYSTROPHY (DMD)

- Definition: Progressive genetic disorder that affects muscles in lower extremities and eventually muscles of upper extremities, chest wall, and heart
- Etiology/Incidence
 1. X-linked recessive genetic disorder which results in absence or severe deficiency of cytoskeletal protein known as dystrophin
 2. Most commonly inherited neuromuscular disease in children
 3. Affects 1:3500 males; 1:1750 females are carriers
 4. Average age of diagnosis is 3 to 5 years of age

- Signs and Symptoms
 1. At birth—rarely affected clinically
 2. Becomes clinically evident by 3 to 5 years of age
 - a. Abnormalities of gait and posture
 - b. History of delayed developmental milestones
 - c. Large “muscular” looking calves
 - d. Inability to keep up with peers when running
 3. Progresses over next 2 decades—weakness more evident in proximal muscles
 4. Wheelchair dependent by 10 to 12 years of age
 - a. Muscles decrease in size
 - b. Contractures progress with loss of joint mobility
 - c. Kyphoscoliosis develops with respiratory function problems
 5. Complications from:
 - a. Cardiac involvement
 - b. Nervous system involvement
 - c. Musculoskeletal deformities
 - d. Compromised respiratory function
 6. Eventual death from cardiac or respiratory failure

- Differential Diagnosis
 1. Hypothyroidism
 2. Carnitine deficiency
 3. Spinal muscular atrophy
 4. Fascioscapulohumeral dystrophy
 5. Other types of muscular dystrophies of childhood

- Physical Findings
 1. Preschooler—3 to 5 years of age
 - a. Increasing lumbar lordosis
 - b. Pelvic waddling
 - c. Gowers’ maneuver
 - (1) Child may “walk” hands up legs to attain a standing position when arising from floor
 - (2) Indication of pelvic girdle weakness
 - (3) Distinctive in DMD, but seen in other conditions as well
 - d. Proximal muscle strength and ankle reflexes may be depressed
 - e. Calf hypertrophy present and the enlarged muscle tissue is eventually replaced with fat and connective tissue (pseudohypertrophy)
 2. Cardiac involvement in all patients—cardiomyopathy by adolescence
 3. Contractures develop before ambulation is compromised
 - a. Iliotibial bands
 - b. Hip flexors
 - c. Heel cords

- Diagnostic Tests/Findings
 1. Obtain 3-generation family history
 - a. May be positive history of muscle disorders, weakness, DMD
 - b. Carrier females have symptoms of weakness or cramping of muscles
 2. Laboratory studies
 - a. Creatine kinase (CK)—will be markedly elevated in affected males (15,000 to 35,000 IU/L)
 - b. EMG—distinctively myopathic
 - c. ECG—changes are distinctive
 - (1) Tall right precordial R waves
 - (2) Deep Q waves in left precordial and limb leads
 - d. Muscle biopsy—histopathologic findings
 - (1) Groups of necrotic degenerating fibers most prominent
 - (2) Variation in fiber size evident
 - (3) Dystrophic immunoreactivity confirms DMD
 - e. Genetic testing
 - (1) Utilizes WBC from blood specimen
 - (2) DNA analysis of DMD gene confirms diagnosis

- Management/Treatment
 1. No cure available at present
 2. Goal of treatment is essentially symptomatic, aimed at delay of progression and supportive care
 3. Maintenance of strength and mobility
 - a. Exercise
 - b. Use of ankle/foot orthoses, bracing, spinal support measures, and wheelchair as needed
 4. Consultation with neuromuscular disease specialty team for diagnosis and for periodic evaluations
 5. Refer family for genetic testing/counseling
 6. Counsel family regarding course of disease
 7. Refer parents
 - a. To other families who have children with DMD
 - b. To other community resources for support

■ QUESTIONS

Select the best answer

1. Which of the following disorders is usually associated with adduction of the forefoot?
 - a. Internal femoral torsion
 - b. Talipes equinovarus congenita
 - c. Genu valgum
 - d. Internal tibial torsion

2. The most common rheumatoid disease of childhood is:
 - a. Systemic lupus erythematosus
 - b. Kawasaki disease
 - c. JRA
 - d. Legg-Calvé-Perthes disease
3. Radiographic findings of disease progression and sphericity of femoral head is helpful in the diagnosis and follow-up of:
 - a. Transient synovitis of the hip
 - b. Osgood-Schlatter disease
 - c. Legg-Calvé-Perthes disease
 - d. Slipped capital femoral epiphysis
4. A 4-year-old boy is brought in by his mother, concerned about the sudden onset of a painful limp in his right leg 2 days ago. Today he has a low-grade fever. Which of the following diagnoses is most likely?
 - a. Osgood-Schlatter
 - b. JRA
 - c. Osteomyelitis
 - d. Transient synovitis of the hip
5. Which of the following would be the most appropriate initial management of a newborn diagnosed with developmental dysplasia of the hip?
 - a. Observe and reexamine at 2-week well-child visit
 - b. Triple diapering in nursery
 - c. Pavlik harness
 - d. Surgical reduction
6. A physical finding not usually associated with talipes equinovarus congenita is:
 - a. Contracture of the iliotibial bands
 - b. Deep crease on medial border of foot
 - c. Atrophy of calf muscles
 - d. Small foot with limited dorsiflexion
7. A characteristic feature of polyarticular JIA disease is:
 - a. The involvement of 5 or more inflamed joints
 - b. Confinement to lower extremity joints, knees, and ankles
 - c. Asymmetric involvement
 - d. High, daily intermittent spiking fevers
8. ANA seropositivity for antibodies is:
 - a. A valuable diagnostic marker for JIA
 - b. Is not positive in any other childhood diseases
 - c. More commonly found in older boys or in systemic disease
 - d. Present in over 75% of cases
9. Dislocation in the hip of a child six months or older may typically present with:
 - a. Asymmetry of skin folds
 - b. Atrophied hip muscles
 - c. Positive Galeazzi sign
 - d. Negative Trendelenburg sign
10. For a newborn, the correct management of hip dislocation should include:
 - a. Use of flexion-abduction device such as Pavlik harness to stabilize hip
 - b. Follow and observe closely for 3 to 4 weeks, then refer to orthopedist
 - c. Surgical reduction
 - d. Traction for 6 weeks
11. Duchenne muscular dystrophy is characterized by which of the following signs and symptoms?
 - a. At birth, affected infants are notably hypotonic, "floppy" babies
 - b. Earliest symptom is often refusal to bear weight
 - c. Abnormalities of gait and posture become evident during preschool years
 - d. Unable to keep up with peers when running by school age
12. Most children with Duchenne muscular dystrophy become wheel-chair dependent by what age?
 - a. 7 to 9 years of age
 - b. 10 to 12 years of age
 - c. 14 to 16 years of age
 - d. Highly variable depending on response to treatment
13. School-aged children and young adolescents involved in athletic activities may not be at increased risk for:
 - a. Osgood-Schlatter disease
 - b. Chondromalacia
 - c. Spondylolysis
 - d. Slipped capital femoral epiphysis
14. Management of scoliosis depends on the severity of curve as well as the age of the child. Which of the following would require surgical intervention?
 - a. Curves of 15 degrees in a child who is still growing
 - b. Thoracic and/or lumbar curve greater than 25 degrees, even if growth is complete
 - c. Thoracic curve greater than 30 degrees or lumbar curve greater than 40 degrees that has not progressed while in brace
 - d. Thoracic curve greater than 50 degrees or lumbar curve greater than 40 degrees

15. In performing a diagnostic workup and management plan for a child with osteomyelitis, which of the following is not accurate or recommended?
 - a. Elevated ESR confirms diagnosis
 - b. Aspiration is usually indicated
 - c. Antibiotic treatment for 4 to 6 weeks is recommended
 - d. Surgery is recommended if abscess is present
16. A 6-year-old child presents with a limp and knee pain. The PNP finds limited passive internal rotation and abduction of the hip joint on physical examination. The most likely diagnosis is:
 - a. Slipped capital femoral epiphysis
 - b. Osgood-Schlatter disease
 - c. Transient synovitis of the hip
 - d. Legg-Calvé-Perthes disease
17. Which of the following statements is true about acute osteomyelitis?
 - a. Occurs more frequently in females than males
 - b. Peak ages are infancy (less than 1 year) and preadolescence (9 to 11 years)
 - c. Most common sites are radius and ulna
 - d. A self-limiting disorder
18. Which of the following statements is not true of slipped capital femoral epiphysis?
 - a. Thought to be precipitated by hormone changes during puberty
 - b. Unilateral involvement is more common than bilateral
 - c. More common among males and African-Americans
 - d. Thought to be caused by repetitive stresses in young athletes prior to growth spurt
19. Genu varum is considered an abnormal condition when:
 - a. Extreme knock-knees continues after 7 years of age
 - b. Extreme bowing continues after 2 years of age
 - c. Parents are concerned about their child's appearance
 - d. Evident before 2 years of age
20. Tibial torsion is commonly associated with:
 - a. Pain
 - b. Restricted ROM
 - c. Internal rotation of lower extremities
 - d. Occurrence in adolescents 13 to 16 years of age
21. Which of the following diagnoses is associated with contracture of one of the sternocleidomastoid muscles?
 - a. Lordosis
 - b. Torticollis
 - c. Scoliosis
 - d. Kyphosis
22. Sports injuries are commonly associated with:
 - a. Improper training
 - b. Higher frequency in females
 - c. Scoliosis
 - d. Low socioeconomic status
23. Initial treatment of a sprain includes which of the following?
 - a. Rest, ice, compression, elevation, and NSAIDs
 - b. Heat, ROM exercise, compression, elevation, and NSAIDs
 - c. Rest, heat, compression, elevation, and NSAIDs
 - d. Rest, ice, ibuprofen, compression, and NSAIDs
24. The most definitive feature(s) for a diagnosis of "growing pains" includes:
 - a. Exclusion of other causes of lower extremity pain
 - b. Pain, swelling, erythema
 - c. Loss of ambulation
 - d. Decreased ROM
25. Systemic-onset JIA is most commonly associated with:
 - a. High, daily intermittent spiking fevers and rash
 - b. Single joint involvement
 - c. Positive RF factor
 - d. Painless joint involvement
26. Signs and symptoms associated with Duchenne muscular dystrophy are:
 - a. History of delayed developmental milestones
 - b. Visual-motor disturbance, calf hypertrophy
 - c. Delayed motor development, positive Ortolani maneuver
 - d. History of "clumsiness," visual-motor disturbance
27. Complications of SLE commonly include which of the following?
 - a. Pericarditis, arthritis, nephritis
 - b. Encephalitis, nephritis, pericarditis
 - c. Nephritis, arthritis, rheumatic fever
 - d. Nephritis, hemolytic anemia, contact dermatitis

28. Which of the following children need an orthopedic referral?
- A 6-year-old with mild bowing of the lower legs
 - A 6-month-old with internal tibial torsion
 - A three-week-old with equinovarus of feet
 - A newborn with a positive Pavlik sign
29. Antonio is a newborn and the PNP notes on physical assessment that both his feet turn in. When attempting range of motion, she finds that both feet move relatively freely in all directions. Antonio has:
- Clubfoot
 - Syndactyly
 - Metatarsus adductus
 - Fracture in his feet
30. Which of the following is an appropriate goal for a child being treated for osteomyelitis?
- Prohibiting activities
 - Complete course of antibiotic therapy
 - Encouraging a low fat diet
 - Restricting visitors
31. In a newborn, a diagnosis of hip dislocation is suspected when:
- Positive Galeazzi, Barlow, and Ortolani
 - Wide hip abduction that is symmetric
 - Flaccidity of the left leg following extension of both legs with return to flexion
 - Tonic neck reflex in which the left leg is flexed
32. Which of the following statements is true regarding slipped capital femoral epiphysis?
- More common in females
 - Generally occurs following severe sudden trauma
 - Incidence more common in athletes
 - The goal of treatment is to stabilize or improve the position of the femoral head
33. In Legg-Calvé-Perthes disease, which of the following signs and symptoms are seen?
- Insidious onset of limp with knee and groin pain
 - Sudden onset of limp and pain in lateral hip
 - Fever and insidious onset of limp
 - Afebrile and sudden onset of limp
34. Which of the following is true for idiopathic scoliosis, which occurs primarily in adolescents?
- Mild curves occur equally between the sexes
 - Generally there is no family history

- Back pain is usually associated with curves of 35 degrees or greater
- Bracing is indicated for thoracic curves of 10 to 25 degrees

■ ANSWERS

- | | |
|--------------|--------------|
| 1. b | 18. d |
| 2. c | 19. b |
| 3. c | 20. c |
| 4. d | 21. b |
| 5. c | 22. a |
| 6. a | 23. a |
| 7. a | 24. a |
| 8. a | 25. a |
| 9. c | 26. a |
| 10. a | 27. a |
| 11. c | 28. c |
| 12. b | 29. c |
| 13. d | 30. b |
| 14. d | 31. a |
| 15. a | 32. d |
| 16. d | 33. a |
| 17. b | 34. a |

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Neurological Disorders

Maureen Maguire

■ SEIZURE DISORDERS/EPILEPSY

- Definition
 1. Seizures—disturbances of normal nerve cell function characterized by uncontrolled, spontaneous electrical activity in the brain, that may result in loss of consciousness, altered body movements, or disturbances of sensation and behavior
 2. Epilepsy—condition of recurrent seizures
 3. Status epilepticus—a seizure lasting for 30 minutes or a series of 3 seizures without any periods of consciousness between them
- Etiology/Incidence
 1. Caused by any event with potential to produce insult to the brain
 2. Multiple causes; specific etiologies remain uncertain for 50% of cases
 - a. Genetic component or familial predisposition
 - b. Genetic disorders—tuberous sclerosis; neurofibromatosis
 - c. Hemorrhage (intracranial)
 - d. CNS infection (encephalitis, meningitis)
 - e. Head trauma
 - f. Developmental defects of brain
 - g. Biochemical factors (inborn metabolic errors, electrolyte imbalance)
 - h. Intracranial tumors
 - i. Toxic ingestions—e.g., alcohol
 - j. Poor drug compliance or altered drug metabolism because of illness
 3. Incidence varies greatly with age
 - a. 1:1000 during first year of life
 - b. 50% of all cases of epilepsy occur before age 25
 4. Variation in clinical manifestations due to location of brain involved
- Types of Seizures
 1. Partial seizures—begin with an electrical discharge in one limited area of the brain
 - a. Simple partial seizures
 - (1) Characterized by seizure activity restricted to one side of body but may spread to other parts of the body
 - (2) Usually last less than 2 minutes; no loss of consciousness and no postictal state
 - (3) Motor—part of body or entire side (e.g., arm, leg, Jacksonian march, postural, vocalizations)
 - (4) Sensory—visual, auditory, olfactory, paresthesias, visual hallucinations
 - (5) Autonomic—result in changes in parts of the nervous system that automatically control bodily functions, e.g., tachycardia, pallor, sweating, flushing
 - (6) Psychic—changes in how the person thinks, feels, or experiences things, e.g., memory problems, fearful, or feelings of déjà vu
 - b. Complex partial seizures—start in a limited area of the temporal or frontal lobes

- and spread to other parts of the brain with a variety of clinical expressions
- (1) Impairment of consciousness for 30 seconds to approximately 2 minutes or longer
 - (2) Cognitive symptomatology
 - (a) Abrupt alteration in mental state
 - (b) Involves disruption of time relationships and memory
 - (3) Affective symptomatology—inexplicable feelings, e.g., fear or dread without obvious cause
 - (4) Somatosensory disturbances
 - (a) Distortions of perception or hallucinations
 - (b) May involve taste, smell, vision
 - (5) Automatism
 - (a) Occur in 50% to 75% of cases
 - (b) Semipurposeful perseverative movements
 - (c) May involve walking, sucking, lip-pursing, or picking at clothing
2. Generalized seizures—begin with a widespread electrical discharge of both hemispheres of the brain, usually with some loss of consciousness
- a. Absence seizures (petit mal)
 - (1) Onset between 4 and 8 years of age, higher incidence in girls
 - (2) Brief, generalized, nonconvulsive episode with no aura and no postictal state
 - (3) Characterized by interruption of activity, staring and unresponsiveness
 - (4) Usually lasts less than 10 seconds but may be as long as 20 seconds
 - (5) Episode begins and ends abruptly
 - (6) Child may be unaware of episode and is fully alert afterwards
 - b. Tonic-clonic (grand mal)
 - (1) Consists of motor manifestations with loss of consciousness
 - (2) May begin with aura
 - (3) Tonic phase
 - (a) Sustained contraction of muscles
 - (b) Person falls to ground
 - (c) Extensor posturing and tonic contraction
 - (d) Lasts less than 1 minute
 - (4) Clonic phase
 - (a) Bilateral and rhythmic jerking
 - (b) May bite tongue
 - (c) Bowel or bladder incontinence may occur
 - (d) Stops after several minutes
 - (5) Postictal phase
 - (a) Period of cortical inhibition
 - (b) Vomiting may occur
 - c. Confusion and lethargy
 - (d) Gradual recovery of consciousness
- c. Myoclonic seizures
 - (1) Brief (1 to 2 seconds), sudden muscle contractions/jerks
 - (2) Both sides of the body are involved but may involve only one area of body
 - (3) May occur in clusters
 - (4) There may be no alteration in consciousness
 - d. Infantile spasms (West syndrome)
 - (1) Onset during first year of life; usually between 3 and 12 months
 - (2) Sudden jerk followed by stiffening with arm extension and knees pulled up to body (“jackknife” posture)
 - (3) Approximately 60% have some brain disorder before seizures start
 - (4) Often in clusters up to 100 individual spasms
 - (5) Classified in two groups—infants with spasms but no basic neurological disorder have good prognosis whereas those with symptomatic type have a specific etiological factor identified and an 80% to 90% risk of mental retardation
 - e. Atonic
 - (1) Also termed “drop” attacks
 - (2) Characterized by sudden loss of muscle tone, which may result in head nodding, or falling to the ground; usually less than 15 seconds
 - (3) Usually no alteration in consciousness
 - (4) May injure themselves so, if frequent, may require protective helmet
 - f. Lennox-Gastaut syndrome (rare)
 - (1) Severe epileptic encephalopathy
 - (2) Characterized by variety of primary, generalized seizures
 - (3) Onset between 2 and 6 years of age
 - (4) Have a preexisting brain disorder or injury
 - (5) About one third previously had infantile spasms
 - g. Tonic seizures
 - (1) Sudden significant increase in muscle tone lasting less than 20 seconds
 - (2) Often occur during sleep, but if it happens while awake the person will fall
 - (3) Frequently seen with Lennox-Gastaut syndrome
 - h. Clonic seizures
 - (1) Rhythmic jerking of arms and legs which may involve both sides of the body
 - (2) Length varies
 - (3) May progress to tonic-clonic seizure

- Differential Diagnosis
 1. Breathholding spells—usually under 6 years of age
 2. Sleep disorders, e.g., narcolepsy
 3. Tics
 4. Complicated migraine headaches
 5. Syncope—more common in adolescence
 6. Gastroesophageal reflux
 7. Benign paroxysmal vertigo
 8. Pseudoseizures
- Diagnostic Tests/Findings
 1. Clinical and laboratory diagnosis
 2. Complete history
 - a. Detailed description of seizure—onset, type, duration
 - b. Previous seizures and frequency
 - c. Evidence of infection
 - d. Abnormal behavior
 - e. Previous static or progressive neurologic/developmental dysfunction
 - f. Pica; possible ingestion of lead
 - g. Trauma
 - h. Birth history
 - i. Current medication including anticonvulsants
 - j. Family history of febrile and nonfebrile seizure
 3. Laboratory studies—to identify possible underlying etiology
 - a. Serum fasting glucose, calcium, magnesium, and serum electrolyte levels
 - b. Seizures and mental retardation suggestive of metabolic problem
 - (1) Plasma amino acids
 - (2) Blood ammonia
 - (3) Blood lactate and pyruvate
 - (4) Urinary organic acids
 - (5) Cytogenetic analysis
 - c. Other studies as determined by history/physical findings
 4. Lumbar puncture—not routine
 5. Neuroimaging studies
 - a. Skull radiographs—rarely indicated, but helpful to rule out skull fracture
 - b. Cranial CT and MRI
 - (1) For detection of structural abnormalities
 - (2) Indicated for:
 - (a) A changing seizure pattern
 - (b) Focal or lateralized abnormalities
 - (c) History of trauma
 - (d) Focally abnormal EEG
 - (e) Evidence of increased intracranial pressure
 - (f) Known or suspected specific white or gray matter disease
 - (g) First seizure in all adolescents
- 6. EEG—measures physiologic function of brain
 - a. Performed on all children, at least initially, to evaluate seizures
 - b. Interpret in context of child's age, history, and physical findings
 - c. Recorded for 1 hour during periods of wakefulness and sleeping
 - d. Helps to define seizure type
 - e. Epileptiform EEG supports diagnosis; normal EEG does not exclude diagnosis
 - f. Twenty-four hour ambulatory EEG often used if regular EEG is inconclusive
- Physical Findings
 1. Physical examination can be abnormal with underlying cerebral pathology
 2. Transient abnormal neurologic signs common during and after seizures
 - a. Intention tremor
 - b. Incoordination
 - c. Weakness of one or more extremities
 - d. Pathologic exaggeration of reflexes
 - e. Confusion
 3. Warning signs of serious neurologic problems
 - a. Asymmetric pupils
 - b. Signs of increased intracranial pressure
 - c. Focal deficits
 - d. Unusual physical features, e.g., lesions seen in tuberous sclerosis that may indicate underlying genetic disorder
 - e. Change in seizure pattern
- Management/Treatment: Referral to neurologist for initial management
 1. Antiepileptic drugs
 - a. Principles
 - (1) Utilize least number of medications
 - (2) Maintain maximum level of alertness with fewest number of seizures
 - (3) Select drug effective for specific seizure type (see **Table 11-1**)
 - (4) Use least toxic, least expensive, requiring least amount of laboratory monitoring
 - (5) Begin with single drug, easier to assess side effects
 - (6) Obtain baseline of child's physical status
 - (a) Complete blood count
 - (b) Liver function tests
 - (c) Blood urea nitrogen and urinalysis

■ **Table 11-1** Commonly Used Antiepileptic Drugs and Type of Seizures

Type of Seizure	Drug(s) of Choice	Drugs also Used
Generalized tonic-clonic	Carbamazepine Valproic acid	Phenobarbital Phenytoin
Simple/complex partial	Carbamazepine Valproic acid	Phenobarbital Phenytoin
Absence	Valproic acid Ethosuximide	Clonazepam
Myoclonic	Valproic acid	Clonazepam
Lennox-Gastaut	Valproic acid	Ethosuximide
Infantile spasm	Corticosteroids	Valproic acid

Adapted from: Heighway, S. (2004). *Pediatric nurse practitioner certification review guide* (4th ed.). (Virginia Layne Millonig and Caryl Mobley, Eds.). Sudbury, MA: Health Leadership Associates, Inc.

- b. Factors to consider in choice of drug (see **Table 11-2**)
 - (1) Type of seizure
 - (2) Dosage
 - (3) Potential side effects
 - (4) Half-life
 - (5) Age, sex, weight, and physical condition of child
2. Anticipatory guidance
 - a. First aid measures
 - (1) Protect child from injury, but do not restrain
 - (2) Assess for adequate airway, breathing, and circulatory status
 - (3) Do not insert items in mouth
 - (4) Note time, duration, and activity
 - (5) If seizure persists beyond 10 to 15 minutes, seek medical assistance
 - b. Observe for side effects of drugs
 - c. Possible drug interactions
 - d. Guidelines for activities/activity restrictions
 - (1) Avoid activities where seizures may cause dangerous fall (e.g., high diving, rope climbing)
 - (2) Supervised swimming only
 - (3) Discuss other sports participation with parents and child
 - e. Importance of compliance with drug therapy
3. Intractable seizures—15% children with epilepsy
 - a. Refer to epilepsy center with interdisciplinary team
 - b. Alternative treatments
 - (1) Ketogenic diet—very restricted, high fat/low carbohydrate diet

■ **Table 11-2** Considerations with Antiepileptic Drugs

Drug	Oral Dosage (mg/kg/d)	Half-life Hours	Side Effects
Carbamazepine	20–30	5–23	Allergic rashes, nausea, lethargy, blurry vision, nystagmus, dizziness, bone marrow suppression, decreased liver function
Valproic acid	30–60	6–16	Nausea, weight gain, tremor, transient alopecia, hepatotoxicity, leukopenia, thrombocytopenia, rash
Ethosuximide	20–40	15–68	Abdominal discomfort, nausea, rash, hiccups, behavioral problems, drowsiness, dystonias, blood dyscrasias
Phenytoin	5–7	7–42	Rash, hirsutism, nausea, psychomotor slowing, neuropathy, ataxia, gingival hyperplasia, folate deficiency
Phenobarbital	4–6	36–120	Sedation: paradoxical; hyperexcitability (rare)
Clonazepam	0.05–0.2	18	Ataxia, sedation, drooling, inattention, withdrawal seizures
ACTH	20–40 IU	—	Cushing syndrome, hypertension, susceptibility to infections, GI bleeding, hyperglycemia

Adapted from: Heighway, S. (2004). *Pediatric nurse practitioner certification review guide* (4th ed.). (Virginia Layne Millonig and Caryl Mobley, Eds.). Sudbury, MA: Health Leadership Associates, Inc.

- (2) Epilepsy surgery—approximately half of children with intractable seizures are appropriate candidates for surgical procedures

■ FEBRILE SEIZURES

- Definition: Generalized tonic or tonic-clonic seizures which occur as a consequence of an abrupt and steep rise in body temperature in young children
- Etiology/Incidence
 1. 3% to 4% of otherwise healthy children between 9 months and 5 years of age have one or more febrile seizures; peak onset between 18 and 24 months of age
 2. Exact cause unknown; hereditary influence suspected
 3. Most common nonepileptic seizure disorder and usually benign in nature
 4. Cooling the child's body will not prevent a febrile seizure
- Signs and Symptoms
 1. Two groups based on clinical features
 - a. Simple febrile seizures
 - (1) Duration < 15 minutes
 - (2) No focal features
 - (3) If occur in series, total duration of less than 30 minutes
 - b. Complex febrile seizures
 - (1) Duration > 15 minutes
 - (2) Focal features or postictal paresis present
 - (3) Occur in series with total duration greater than 30 minutes
- Differential Diagnosis
 1. Underlying meningitis or encephalitis
 2. Chills
 3. Underlying metabolic disorder
 4. Epilepsy
- Physical Findings
 1. Often associated with viral or bacterial illness
 2. Occur as the fever is rising
 3. Usually fever above 102°F (39°C)
- Diagnostic Tests/Findings
 1. Detailed description of seizure
 2. Complete physical assessment with careful neurological examination
 3. Lumbar punctures not routine, but indicated for:
 - a. Infants < 1 year of age
 - b. When seizure occurs after second day of illness
 - c. To rule out meningitis

4. Skull radiographs not routine
5. Blood laboratory studies, EEG, CT, or MRI not routine but performed if indicated by history or physical findings

- Management/Treatment
 1. Urgent treatment for unabated seizures of ≥ 10 minutes
 2. Fever control for temperature > 101°F to make the child feel better
 - a. Sponge baths with tepid water are not recommended because they are ineffective and risk the possibility of increasing temperature through shivering
 - b. Antipyretic use, e.g., acetaminophen for child's comfort
 3. A comprehensive search for cause of fever and treatment of underlying illness
 4. Reassurance to parents regarding excellent prognosis
 5. Antiepileptic drugs
 - a. Short-term or prolonged anticonvulsant prophylaxis not recommended
 - b. On rare occasions rectal diazepam may be used when parents have significant anxiety or for prolonged seizure; the side effects of diazepam (drowsiness, lethargy, and ataxia) could mask signs of a CNS infection
 - c. Parents should be taught the basic first aid measures for seizures

■ HEADACHES

- Definition: Head pain
- Etiology/Incidence
 1. Exact etiology often unknown; pain may be extracranial and/or intracranial
 - a. Possible causes
 - (1) Genetic predisposition
 - (2) Head trauma
 - (3) Environmental factors
 - (4) Illness or infection
 - (5) Emotional factors
 - (6) Certain foods and beverages, e.g., MSG and caffeine
 2. Difficult to distinguish tension headaches from migraine headaches in many children
 3. Chronic and recurrent headaches are most common neurologic complaints; occur in 3% to 20% of children and adolescents
 - a. Migraines—account for 50% of chronic or recurrent headaches; 60% of affected children are male

- b. Cluster headaches—least common in children
 - (1) Briefer in duration and more frequent than migraines
 - (2) Begin in teenage years
 - (3) More frequent in males
 - (4) Sharp, stabbing pain on one side of the head which is disabling
 - c. Tension headaches
 - 4. Headache pain is produced by several factors which activate pain fibers
 - a. Inflammation
 - b. Stretching
 - c. Torsion
 - d. Contraction of innervated structures
 - (1) Large intracranial arteries and veins
 - (2) Dural sinuses
 - (3) Periosteum of bone
 - (4) Muscle/skin of scalp
 - (5) Teeth and gums
- Signs and Symptoms
 1. Migraine headaches—recurrent headaches with varied frequency, intensity, and duration
 - a. Diagnostic criteria—repeated episodes of headache accompanied by at least 3 of the following symptoms:
 - (1) Recurrent abdominal pain (with or without headache), nausea, vomiting, extreme sensitivity to light or sound
 - (2) Often have an aura, usually visual, but may be sensory, motor, or vertiginous
 - (3) Throbbing or pounding pain
 - (4) Pain restricted to one side of head (although may shift sides from one headache to next); bilateral before puberty
 - (5) Relief of pain by brief periods of sleep
 - (6) Family history of migraine in one or more immediate relatives
 2. Tension headaches—headaches occurring with most frequency and intensity during periods of increased stress
 - a. Tend to involve occipital or temporal regions bilaterally and often extend to the neck, or may be diffuse
 - b. Nausea and vomiting may occur, but not as often as with migraine
 3. Intracranial headaches—may have certain distinguishing factors
 - a. Severe occipital headache
 - b. Exacerbated by straining, sneezing, or coughing
 - c. Awaken child from sleep
 - d. Exacerbated or improved markedly by position changes
 - e. Associated with projectile vomiting or vomiting without nausea
 - f. Associated with history of focal seizures
 - g. Increase in intensity and severity if not treated
 - 4. Sinus headache—chronic or recurrent headaches which occur in about 15% of children with chronic sinusitis
 - a. Often occur same time each day, build slowly, with throbbing quality
 - b. Accompanied by rhinorrhea, postnasal drip, persistent cough, recurrent ear infections; afebrile
 - c. Pain or pressure over frontal or maxillary sinuses
- Differential Diagnosis
 1. Acute, severe headache—distinguish between intracranial and extracranial cause by thorough history and physical examination
 - a. Intracranial causes
 - (1) Intracranial mass
 - (2) Infection—meningitis, encephalitis
 - (3) Intracranial hemorrhage
 - (4) Post-head injury
 - (5) Post-seizure
 - b. Extracranial causes
 - (1) Sinusitis—rare
 - (2) Temporomandibular joint problem
 2. Chronic headaches—stress, anxiety, or tension with resulting muscle contractions
 - Physical Findings
 1. Chronic migraine or tension headaches—physical examination generally normal
 2. Acute headache—critical indicators of possible intracranial cause
 - a. Meningismus
 - b. Focal neurologic signs
 - c. Papilledema
 - d. Split sutures
 - e. Evidence of cranial trauma (including blood in or behind ear)
 - f. Depressed level of consciousness
 - Diagnostic Tests/Findings:
 1. Unnecessary unless history/physical examination suggest intracranial etiology, e.g., infection, bleeding, or tumors, in which case, CT, MRI, or lumbar puncture
 2. Psychological evaluation may be needed with frequent tension headaches
 - Management/Treatment
 1. Specific treatment determined by etiology
 2. Headaches with suspected intracranial etiology need evaluation by physician/neurologist

3. Cautious use of medications for pain relief
 - a. Initial treatment with simple analgesics
 - b. Acetaminophen
 - (1) Under 1 year—60 mg orally every 4 to 6 hours
 - (2) 1 to 3 years—60 to 120 mg orally every 4 to 6 hours
 - (3) 3 to 6 years—120 to 180 mg orally every 4 to 6 hours
 - (4) 6 to 12 years—240 mg orally every 4 to 6 hours
 - (5) Older than 12 years—325 to 650 mg orally every 4 to 6 hours
 - c. If mild analgesics ineffective in children older than 11 years, may progress with caution to:
 - (1) Midrin
 - (a) Combination of isometheptene mucate 65 mg (constricts blood vessels); acetaminophen 325 mg and dichloralphenazone 100 mg (mild sedative)
 - (b) 1 to 2 capsules immediately; repeat 1 capsule every hour until relieved (a maximum of 3/day or 5/week)
 - (2) Ergotamine tartrate—2 mg sublingual
 - d. Prophylactic treatment
 - (1) With headaches more than once/week and those that interfere with routine activities, e.g., school attendance
 - (2) Unresponsive to symptomatic treatment
 - (3) Must consider side effects
 - (4) Medications used
 - (a) Propranolol—0.5 to 3.0 mg/kg/day \geq 11 years of age
 - (b) Cyproheptadine hydrochloride—0.25 mg/kg/day \geq 3 years of age
 - (c) Phenobarbital—3 to 5 mg/kg/day every night (3 to 10 years)
 - (d) Phenytoin—5 mg/kg/day in 2 doses
 - (e) Amitriptyline hydrochloride—1 to 2 mg/kg/day \geq 11 years of age
4. Stress management and relaxation techniques
5. Eliminate foods which may be triggers, e.g., chocolate, cheese, MSG
6. Refer to specialists for further evaluation if headaches are recurrent, unresponsive (including allergist, psychiatrist, neurologist, or pain clinic)

■ NEUROFIBROMATOSIS (NF1) (VON RECKLINGHAUSEN DISEASE)

- Definition
 1. A genetic disorder which causes tumors to grow around nerves
 2. Most common type of NF
 3. Always a progressive disorder
 4. Intelligence is usually normal
- Etiology/Incidence
 1. Inherited autosomal dominant disorder; associated with chromosome # 17, but 30 to 50% result from a spontaneous mutation of unknown cause
 2. Occurs 1:3000 to 1:4000 persons
 3. High variability and severity among individuals and within families
 4. Penetrance (likelihood that mutant gene will express itself at all, if it is present) is virtually 100%
- Signs and Symptoms—2 or more of the following:
 1. Six or more light brown spots on the skin (“café au lait spots”—CAL) measuring more than 5 mm in children or more than 15 mm in adolescents
 2. Freckling in armpits or groin
 3. Two or more neurofibromas
 4. Two or more growths on the iris of the eye (Lisch nodules)
 5. Optic glioma
 6. Learning disabilities are common
 7. Seizures are possible
 8. Distinctive osseous lesions (e.g., sphenoid wing dysplasia or thinning of long bone cortex, with or without pseudoarthritis)
 9. First degree relative (parent, sibling, child) with NF1
- Differential Diagnosis
 1. Benign condition with < 5 or 6 CLSs
 2. McCune-Albright syndrome
 3. Neurolemmoma
 4. Proteus syndrome
- Diagnostic Tests/Findings: Additional tests to confirm diagnosis and identify complications
 1. Slit-lamp ocular examination to identify ocular abnormalities (e.g., iris Lisch nodules and signs of optic pathway glioma)
 2. Cranial MRI—to identify gliomas
 3. Radiographs of spine, chest, skull—to identify neurofibromas and other tumors
 4. EEG—to detect epileptogenic foci

5. Psychological evaluation of school-age child for school—to determine abilities and deficiencies
 6. Audiogram/brain stem evoked response testing—baseline for hearing and detection of acoustic neuromas
- Management/Treatment
 1. Problems associated with NF1 most likely to require medical treatment
 - a. Constipation
 - b. Seizures
 - c. Headaches
 - d. Hyperactivity
 - e. Learning disabilities
 - f. Anxiety
 - g. Renovascular hypertension
 2. Surgery
 - a. Removing or debulking tumors
 - b. Treating skeletal dysplasia
 - c. Correcting scoliosis
 - d. Treatment of vascular compromise
 3. Frequent consultation with medical and surgical specialists, social workers, and other health care specialists
 4. Genetic counseling
 5. Referral to support group for NF

■ TUBEROUS SCLEROSIS (TS)

- Definition
 1. Neurocutaneous syndrome with a combination of skin abnormalities, seizures, and cognitive deficits
 2. Progressive disorder with deterioration over time, including new lesions and complications with increasing patient age
 3. Majority have ongoing debilitation from TS throughout their lives
 4. Mental retardation and seizures are most common problems
- Etiology/Incidence
 1. Autosomal dominant mutant gene but many cases result from new mutations
 2. Previously negative family history does not exclude diagnosis
 3. Incidence—1:6000 to 1:50,000 in United States
 4. Most are diagnosed between 2 and 6 years
 5. Risk for affected person's children is 50%
- Signs and Symptoms
 1. Hypopigmented skin lesions often noted at birth
 2. Developmental delay
 3. Abnormal movements, particularly myoclonic jerking movements

- Physical Findings
 1. Establishing diagnosis of TS depends on detecting presence of two major features or one major plus two minor features
 - a. Skin
 - (1) Hypopigmented macules, of elliptical shape (ash-leaf spots)
 - (2) Fibroadenomas—adenoma sebaceum
 - (3) Distinctive brown patch on forehead
 - (4) Shagreen patch (characteristic)—raised lesion in lumbosacral region
 - b. Teeth—characteristic pitting of enamel
 - c. Eye
 - (1) Choroidal hamartomas
 - (2) Hypopigmented defects of iris
 - d. CNS
 - (1) Periventricular tubers
 - (2) Cerebral astrocytomas
 - (3) Nonspecific EEG abnormalities, including hypsarrhythmia
 - e. Cardiovascular
 - (1) Cardiac rhabdomyomas
 - (2) Aortic and major artery constrictions
 - f. Kidney—renal angiomyolipomas
 - g. Lungs—diffuse interstitial fibrosis
 2. Seizures of all types, but myoclonic jerks and mental retardation most common symptoms

- Differential Diagnosis: Seizures or mental retardation

• Diagnostic Tests/Findings

1. MRI scan of brain—virtually diagnostic and must be used in suspected TS
2. Once diagnosis is made, close follow-up surveillance is essential using appropriate clinical laboratory and radiographic techniques to monitor progression and identify new lesions

• Management/Treatment

1. No specific medical treatment available
2. Treat seizures and other complications (e.g., heart failure, renal failure) the same as if TS were not present
3. Surgery on primary lesions may be indicated
4. Genetic counseling
5. Psychometric testing and psychoeducational techniques for cognitive needs
6. Family and individual support and counseling
7. Refer to tertiary care center for specialized treatment, information, and support

■ TIC DISORDERS

• Definition

1. Tic—repetitive, brief, involuntary, stereotypic muscle movement or vocalizations

2. Symptoms appear before 18 years of age and cannot be caused by the ingestion of substances such as stimulants or be part of another medical disease such as Huntington's disease
 3. Severity ranges, in terms of disruption and impairment, from mild and transient to more severe and permanent
 4. Types
 - a. Transient tic disorder—duration less than 1 year
 - b. Chronic tic disorder—duration more than 1 year, simple or complex; are either motor or vocal, but not both
 - c. Tourette syndrome—most severe chronic tic disorder; symptoms include multiple motor and vocal tics, varying in nature and severity over time
 - (1) Frequently have additional problems such as aggressiveness, social withdrawal, self-harming acts, and sleep disorders
 - (2) Symptoms become more unpredictable during adolescence and may result in school refusal
- Etiology/Incidence
 1. Uncertain etiology; possible genetic central nervous system disturbance
 2. Believed that abnormal neurotransmitters in the brain contribute
 3. Transient tics
 - a. Occur in 25% of normal children
 - b. Often begin in school-age children and can be intensified by anxiety, fatigue, or excitement
 4. Tourette syndrome
 - a. Onset between 2 and 15 years of age with mean age 6 to 7 years
 - b. Incidence—1:10,000 persons
 - c. Males 3 times more frequently affected
 - Signs and Symptoms
 1. Characteristics of tics
 - a. Variable expression—frequency, intensity, and severity
 - b. Exacerbated by stress
 - c. Some degree of voluntary control may be present
 - d. Typically subside during sleep
 2. Simple tics
 - a. Movements present that resemble nervous habits
 - b. Facial “twitches,” head shaking, eye blinking, shoulder shrugging, or throat clearing
 - 3. Tourette syndrome
 - a. Simple tics
 - b. Complex sequences of coordinated movements (e.g., bizarre gait, kicking, jumping, body gyrations, scratching, and seductive or obscene gestures)
 - c. Involuntary vocalizations occur, ranging from simple to complex noises
 - d. Expression is gender influenced
 - (1) Motor and vocal manifestations more prevalent in boys
 - (2) Behavioral problems, such as obsessive-compulsive disorder, more common in girls
 - Differential Diagnosis
 1. Chronic tic disorder
 2. Stuttering
 3. Seizures
 4. Medication-induced tics
 5. Pervasive developmental disorder
 6. Psychiatric disorders, e.g., schizophrenia
 7. Medical conditions with associated abnormal movements, e.g., post-viral encephalitis, multiple sclerosis
 - Physical Findings
 1. Usually normal
 2. Some degree of voluntary ability to suppress tics is often present
 - Diagnostic Tests/Findings: None
 - Management/Treatment
 1. Transient tics—support and education for child and family
 2. Chronic tics/Tourette syndrome
 - a. Referral to mental health specialist
 - b. May involve psychotherapy, behavior management, stress management
 - c. Pharmacologic management
 - (1) Consultation and/or referral—pediatric neurologist
 - (2) Medications useful to suppress behavioral symptoms but interfere with daily functioning
 - (3) Pimozide—beginning dose of 1 mg at bedtime
 - (a) Gradual increases (every 5 to 7 days) until symptoms subside
 - (b) Maximum dose of 10 mg/day in children; 20 mg/day in adolescents
 - (c) Side effects—sedation, lethargy, acute dystonic reactions, tardive dyskinesia
 - (d) Less frequent side effects than haloperidol

- (4) Haloperidol—beginning dose of 0.25 to 0.50 mg at bedtime
 - (a) Gradual increases (every 4 to 5 days) until symptoms subside
 - (b) Side effects similar to pimozide; tardive dyskinesia more common than with pimozide
- (5) Clonidine—less effective but with fewer side effects; is sometimes used

■ HEAD INJURIES

- Definition: Any injury to the meninges, scalp, skull, or any part of the brain; severity of injury ranges from very mild to severe brain-damaging injury
 - Etiology/Incidence
 1. Injury resulting from external physical force to the cranium and internal brain structures
 2. Approximately 1 in 25 children receives medical attention each year for head injuries
 3. Twice as frequent in boys
 4. Greatest incidence in children < 1 year and > 15 years
 5. Very young children are especially vulnerable because of relatively larger heads with less musculoskeletal support and large-surfaced vascular scalps
 6. Age specific etiology
 - a. Infant—child abuse, falls
 - b. One to 4 years of age—falls
 - c. School age—falls, motor vehicle accidents, sports/recreation
 - d. Adolescents—motor vehicle accidents, athletic injuries
 - Signs and Symptoms: Important to determine specific circumstances associated with head trauma
 1. Scalp injuries—contusions and lacerations are most frequent
 2. Concussion
 - a. Head injury sufficient to cause brief loss of consciousness and/or amnesia for event
 - b. Presence and duration of amnesia is indicative of severity
 - c. Usually brief loss of consciousness is followed in minutes by complete return to normal mental status and behavior
 - d. Recovery usually complete without complications
 - e. Few children experience postconcussional syndrome several hours after concussion, characterized by:
 - (1) Headache
 - (2) Drowsiness
 - (3) Confusion or irritability
 - (4) Symptoms may last for several days
3. Diffuse axonal injury (DAI)
 - a. Diffuse injury, usually resulting from violent motion, e.g., motor vehicle accidents
 - b. Diagnosis of exclusion associated with:
 - (1) Unconsciousness \geq 6 or more hours
 - (2) No other cause for symptoms, such as seizures or hematoma
 - c. More serious diagnosis with less favorable prognosis than concussion
 - d. Other symptoms depend on severity of injury
 - (1) Abnormal movements with abnormal pupillary reaction
 - (2) Difficulty regulating respirations and blood pressure
 4. Contusion
 - a. Bruising or tearing of cerebral structures
 - b. Common locations include frontal, temporal lobes and orbital area
 - c. Focal motor signs related to increased intracranial pressure (ICP) may be diagnostic
 5. Hematomas
 - a. Epidural hematoma—blood clot formation between skull and dura of the brain
 - (1) Most treatable, but potentially most lethal type of head injury
 - (2) Classic clinical sign is delayed onset of symptoms
 - (3) Initial neurological symptoms may be minimal or absent
 - (4) Secondary injury from increased ICP as hematoma enlarges, may result in neurological symptoms
 - (a) Headache
 - (b) Confusion
 - (c) Vomiting
 - (d) One-sided weakness
 - (e) Agitation
 - (5) Symptoms may progress to lethargy, coma, and even death if left untreated
 - (6) Prognosis is favorable if surgery is performed before secondary injury becomes irreversible, but this may not happen due to delayed presentation of symptoms
 - b. Subdural hematoma—blood clot formation on brain surface beneath the dura mater
 - (1) Acute presentation
 - (a) Symptoms appear within 48 hours of head injury
 - (b) Signs of intracranial hypertension including irritability or lethargy, vomiting, headache
 - (2) Subacute
 - (a) Symptoms appear between 2 and 21 days

- (b) Chronic hematoma with symptoms appearing after 21 days
 - (c) Signs include seizures, motor abnormalities (hypertonicity and agitation), systemic (irritability, vomiting, fever, anemia, poor weight gain)
- 6. Skull fracture
 - a. Brain damage may not be present, and conversely, brain can be injured without skull fracture present
 - b. Type, severity, symptomatology depend on area involved, age of child, and force of impact
 - c. Linear
 - (1) Fracture in temporoparietal region
 - (2) Most common
 - (3) Frequent in children less than 2 years old who fall from low heights
 - (4) Outcome is usually excellent
 - d. Depressed
 - (1) Skull is disrupted/depressed at point of impact
 - (2) Underlying structures may be bruised or lacerated
 - e. Basal
 - (1) Diagnosis dependent on recognition of signs including hemorrhage in nose, nasopharynx, middle ear
 - (2) Bruising over mastoid bone (Battle sign) or around eyes (raccoon's eye sign)
 - (3) Cranial nerve palsy
 - (4) Cerebral spinal fluid from ears or nose
- Differential Diagnosis
 - 1. Seizure disorder
 - 2. Child maltreatment
- Physical Findings
 - 1. Alterations in vital signs may indicate shock
 - 2. Headache, irritability, and/or crying may indicate acute pain
 - 3. Tense anterior fontanel in young child
 - 4. Alterations in level of consciousness (LOC) vary from irritability, agitation, restlessness to confusion and/or coma
 - 5. Skull fractures may or may not show bony displacement
 - 6. Unilateral swelling may present from a possible hematoma
 - 7. Decreased score on Glasgow Coma Scale
 - 8. Other signs
 - a. Asymmetry of head
 - b. Focal neurologic deficits
 - c. Cranial nerve injuries
 - d. CSF rhinorrhea or otorrhea
 - e. Ecchymosis
 - f. Hearing impairments
- 9. With cerebral edema and increasing ICP, may observe:
 - a. Changes in level of consciousness
 - b. Abnormal respiratory patterns
 - c. Loss of protective reflexes (e.g., cough, gag, or corneal)
 - d. Changes in motor function or posturing
 - e. Nausea and projectile vomiting
- Diagnostic Tests/Findings
 - 1. Skull radiography
 - a. Indicated when severity of head trauma includes:
 - (1) Loss of consciousness
 - (2) Presence of focal neurologic signs
 - b. Only 20% of basal skull fractures can be recognized on standard skull radiographs
 - c. If depressed skull fracture is suspected, obtain tangential views in addition to standard views
 - 2. CT scan—useful in detection of intracranial hemorrhage; investigation of localizing neurologic signs
 - 3. MRI—most useful in evaluation of subacute or chronic, rather than acute injuries
- Management/Treatment
 - 1. For acute injury, rapid, accurate assessment of primary injury and level of severity
 - 2. Most head trauma is minor, with momentary unconsciousness, then child resumes activity and does not require treatment
 - 3. Hospital observation may be required for:
 - a. More than momentary loss of consciousness
 - b. Lethargy, confusion, or irritability
 - c. Severe headache
 - d. Changes in speech or movements in arms and legs
 - e. Significant bleeding from wound
 - f. Vomiting 1 to 2 hours following injury
 - 4. Instructions for home observation
 - a. Need to return to healthcare provider if child exhibits:
 - (1) Excessive prolonged sleepiness
 - (2) Disorientation
 - (3) Confusion
 - (4) Persistent vomiting
 - (5) Seizure
 - (6) Change in swelling of scalp
 - 5. Anticipatory guidance
 - a. Use of safety equipment for recreation and sports
 - b. Never leave infant unattended on bed/furniture

- c. Evaluate play areas for risk factors
- d. Injury prevention instruction
- e. Drivers education/use of seat belts

■ MENINGITIS

- Definition—Inflammation of the meninges
- Etiology/Incidence of bacterial meningitis—related to age
 1. Most frequently a result of hematogenous dissemination
 2. Bacterial pathogens—related to age
 - a. Newborn
 - (1) *Escherichia coli*
 - (2) Group B streptococci
 - (3) *Listeria monocytogenes* (less frequent)
 - (4) Enterobacteriaceae
 - b. Etiologic agents responsible for 95% of cases that occur in children over 2 months of age have been:
 - (1) *Haemophilus influenzae* type b (dramatic decline in incidence with *H. influenzae* immunization)
 - (2) *Streptococcus pneumoniae*
 - (3) *Neisseria meningitidis*
 3. Other etiologic agents
 - a. Mycobacteria
 - b. Fungal infections
 - c. Viral (aseptic)—most common cause of meningitis
 - d. Protozoa
 4. Highest risk—infants between 6 and 12 months of age; 90% of all cases occur in children between 1 month and 5 years
- Signs and Symptoms
 1. Newborn
 - a. Often nonspecific and indistinguishable from those of septicemia
 - b. Most frequent signs
 - (1) Temperature instability
 - (2) Respiratory distress
 - (3) Irritability, lethargy
 - (4) Poor feeding
 - (5) Vomiting
 - c. Seizures present in 40%
 2. Older infants and children
 - a. Nausea and vomiting
 - b. Irritability, confusion
 - c. Anorexia
 - d. Headache, back pain, nuchal rigidity
 - e. Hyperesthesia, cranial nerve palsy, ataxia
 - f. Photophobia
- Differential Diagnosis
 1. Brain abscess
 2. Spinal, epidural, or intracranial abscess
 3. Bacterial endocarditis with embolism
 4. Subdural empyema with or without thrombophlebitis
 5. Brain tumor
- Physical Findings
 1. Newborn
 - a. Bulging fontanel (with or without suture diastasis)
 - b. Increased ICP
 - c. Cardinal signs of meningitis in older children are usually absent in infants
 - (1) No stiff neck
 - (2) No evidence of Kernig's and Brudzinski's signs
 2. Older infants and children
 - a. Common clinical signs associated with meningeal irritation are:
 - (1) Kernig's sign
 - (a) Flexion of the leg 90° at hip
 - (b) Pain on extension of leg
 - (2) Brudzinski's sign—involuntary flexion of legs when neck is flexed
 - b. Headache is frequent sign of increased intracranial pressure
- Diagnostic Tests/Findings
 1. Diagnosis based on examination and culture of cerebrospinal fluid (CSF)
 2. Lumbar puncture for CSF analysis
 - a. Contraindicated in following conditions
 - (1) Cardiopulmonary compromise
 - (2) Signs of increased ICP—papilledema
 - (3) Infection in area overlying lumbar puncture location
 - b. Adverse reactions—pain, headache, bleeding
 3. CSF analysis
 - a. Cloudy
 - b. Increased white blood cell count (predominantly neutrophils)
 - c. Increased protein (> 150 mg/dL)
 - d. Decreased glucose (< 30% of serum glucose)
 - e. Gram stain
 - f. Culture
 4. Polymerase chain reaction analysis possibly
- Management/Treatment
 1. Hospitalization for bacterial meningitis—first 3 to 4 days are critical
 2. Initiate antibiotic therapy once diagnosis is confirmed by clinical findings while awaiting specific CSF and blood culture results

3. Neonates
 - a. Fatality rate is 15% to 30% in neonates
 - b. Prognosis depends upon:
 - (1) Causative pathogen
 - (2) Predisposing risk factors
 - (3) Availability of intensive care facilities
 - c. Follow-up
 - (1) Group B streptococcus
 - (a) 15% to 20% have sequelae—spastic quadriplegia, profound mental retardation, hemiparesis, hearing/visual deficits
 - (b) 11% have hydrocephalus
 - (c) 13% have seizure disorder
 - (2) Gram negative meningitis
 - (a) 10% have severe sequelae of developmental delays
 - (b) About 25% to 35% have mild to moderate sequelae, which may not interfere with normal development
 - (c) Hydrocephalus develops in $\frac{1}{3}$ of infants
 - d. Neonatal survivors without major sequelae typically develop normally
 4. Older infants and children
 - a. Prognosis depends on several factors
 - (1) Patient's age at onset
 - (2) Duration of disease before appropriate antibiotic therapy initiated
 - (3) Specific microorganism involved and number of organisms
 - (4) Whether disorder is compromising host response to infection
 - b. Mortality rate 1% to 5% beyond the neonatal period
 - c. Up to 50% of survivors have some sequelae
 - (1) 10% have hearing deficits
 - (2) 15% have language disorder/delay
 - (3) 2% to 4% have vision impairment
 - (4) 10% to 11% have mental retardation
 - (5) 3% to 7% have motor problems
 - (6) 2% to 8% have seizures
 5. Immunizations for prevention of meningitis
 - a. Hib
 - b. PCV7
 - c. MCV4
2. The primary tumors seen in children
 - a. Astrocytomas
 - (1) Usually slow growing, noncancerous cysts
 - (2) Often develop between 5 and 8 years
 - b. Medulloblastoma
 - (1) The most common brain cancer in children
 - (2) Usually develop around age 5
 - (3) Occur more often in boys
 - c. Brain stem gliomas
 - (1) Occur almost exclusively in children
 - (2) Often grow very large before symptoms develop
 - (3) Average age of development is 5 years
 - (4) Five-year survival rate is low
 - d. Ependymomas
 - (1) Are located in the ventricles
 - (2) Block the flow of CSF
 3. In children between 4 and 11 years, infratentorial (posterior fossa; posterior third of the brain) tumors predominate (60%), including cerebellar and brain stem tumors; the others are supratentorial in the anterior two thirds of the brain
- Signs and Symptoms
 1. In infants with open sutures
 - a. Increased head circumference
 - b. Irritability
 - c. Head tilt
 - d. Loss of developmental milestones
 - e. Bulging fontanelles
 - f. No red reflex in the eye
 2. Older children
 - a. Headache; symptoms usually increase in frequency, becoming more severe in the morning followed by vomiting
 - b. 85% of children with malignant tumors have abnormal neurologic or ocular examinations within 2 to 4 months of onset of headaches
 - c. Certain specific neurologic symptoms may occur later and suggest localization of the CNS tumor
 - (a) Ataxia, hemiparesis, cranial nerve palsies
 - (b) Somnolence
 - (c) Seizures
 - (d) Head tilt
 - (e) Diencephalic syndrome—failure to thrive (FTT), emaciation
 - (f) Diabetes insipidus

■ BRAIN TUMORS

- Definition: A group (mass) of expanding intracranial abnormal cells; the most common tumors in childhood
- Etiology/Incidence
 1. Etiology is unknown; may be genetic predisposition, congenital factors, environmental exposures

- Differential Diagnosis
 1. Brain abscess
 2. Intracranial hemorrhage

3. Nonneoplastic hydrocephalus
 4. Arteriovenous malformations or aneurysm
 5. Indolent viral infections
 6. Subdural hematoma
- Physical Findings: Directly related to anatomic location, size, and to some extent the age of the child. May include:
 1. Tense bulging fontanel at rest (in infant)
 2. Cranial enlargement (present in infants and young children)
 3. Papilledema once sutures are fused (edema of optic nerve)
 4. Nuchal rigidity
 5. Poor coordination or clumsiness
 6. Poor fine motor control
 7. Hypoflexia or hyperflexia
 8. Positive Babinski sign
 9. Spasticity/paralysis
 10. Behavioral changes—may be earliest symptom in child/adolescent
 - Diagnostic Tests/Findings
 1. MRI—superior, sensitive neuroimaging technology
 2. CT scan
 - a. Without contrast, can detect:
 - (1) Whether lesion is cystic or solid
 - (2) Presence of calcifications, hemorrhage, edema, and hydrocephalus
 - b. With contrast can detect:
 - (1) Small tumors
 - (2) Differentiation of areas of edema
 3. Angiography—determines blood supply to affected structures
 4. CT—may include guided biopsy
 5. Lumbar puncture contraindicated in presence of increased ICP
 - Management/Treatment: Therapy selected depends on type and site of tumor
 1. Surgery (usually treatment of choice, although some tumors cannot be removed entirely and can only be debulked)
 2. Radiation therapy
 3. Chemotherapy
 4. Follow-up
 - a. EEG for seizures
 - b. VER for visual problems
 - c. BAER for hearing problems
 - d. Multidisciplinary approach for comprehensive health and developmental needs of child/family

■ QUESTIONS

Select the best answer

1. A typical febrile seizure is most likely to be the problem in which of the following children?
 - a. A 1-year-old with otitis media and a fever of 104°F
 - b. A 3-month-old with unequal pupils and bulging fontanels
 - c. An 11-year-old with fever of 101°F who is on valproic acid for seizure disorder
 - d. A 5-year-old with bacterial meningitis
2. Which of the following responses during a tonic-clonic seizure is most important to teach family members of a child who has these seizures?
 - a. Restrain the child
 - b. Insert an airway into the mouth to prevent tongue biting
 - c. Note the time, duration, and activity of the seizure
 - d. Protect child from injury
3. Which of the following is an appropriate strategy to instruct the parents of an 18-month-old child who has just been diagnosed with a febrile seizure and who has a fever of 104°F?
 - a. Give aspirin every 4 hours for temperature over 101°F
 - b. Bathe with tepid water
 - c. Dress warmly to avoid chills
 - d. First aid management during a seizure
4. What is the primary public health measure to decrease the incidence of bacterial meningitis in infants and children?
 - a. Keeping infants and young children away from crowded places
 - b. Set up screening measures in schools
 - c. Routinely giving immunizations such as Hib and PCV7
 - d. Provide written information about meningitis to high school students
5. The type of seizure that first presents during infancy in which the infant goes into a “jack-knife” posture is:
 - a. Febrile seizures
 - b. Infantile spasm
 - c. Absence seizure
 - d. Atonic “drop” attacks

6. Which of the following would not be associated with an uncomplicated concussion?
 - a. Focal motor signs
 - b. Brief loss of consciousness
 - c. Headache
 - d. Confusion or amnesia for the event
7. Which of the following factors is not usually present in a 4-year-old with migraine headaches?
 - a. Complaints of pain in one side of the head only
 - b. Positive family history in immediate relatives
 - c. Pain relieved by brief sleep
 - d. Recurrent abdominal pain
8. The parents of Sarah, a 13-month-old, are very concerned after she experienced a second simple febrile seizure. At today's visit, they ask you about the use of medications to prevent any further seizures. Which of the following statements is true regarding febrile seizure prophylaxis?
 - a. Short-term anticonvulsant prophylaxis with valproic acid is indicated after 3 febrile seizures
 - b. Prolonged anticonvulsant prophylaxis with phenobarbital for preventing recurrent febrile convulsions is now recommended
 - c. Diazepam may be indicated for children < 12 months who have recurrent, complex febrile seizures
 - d. Fever management with acetaminophen to keep the fever below 102°F (39°C) will prevent another seizure
9. Problems associated with NF1 most likely to require medical treatment include which of following?
 - a. Learning disabilities
 - b. Transient tics
 - c. Obsessive-compulsive behavior
 - d. Progressive mental retardation
10. An 11-year-old girl is brought in by her mother complaining of severe headaches associated with nausea and vomiting. Which of the following signs and symptoms would lead you to consider a brain tumor as part of your differential diagnoses?
 - a. Throbbing pain accompanied by severe light sensitivity
 - b. Bilateral throbbing pain
 - c. Preceded by a visual aura
 - d. More severe in the morning followed by vomiting
11. The National Institutes of Health Consensus conference on NF describes guidelines for diagnosis of NF. Which of the following is not among the diagnostic criteria?
 - a. Family history of first degree relative with NF1
 - b. 6 or more "café au lait" spots > 15 mm diameter in prepubertal child
 - c. Freckling in armpits or groin
 - d. Optic glioma
12. Although meningitis related mortality decreases dramatically after the neonatal period, as many as 50% of survivors experience some sequelae. Which of the following is the most frequent post-meningitis sequela in older infants and children?
 - a. Motor deficits
 - b. Seizures
 - c. Language delays/disorders
 - d. Visual impairments
13. A neonate is being worked up for meningitis after experiencing a seizure preceded by fever, irritability, and poor feeding for one day. On physical examination, which of the following findings would be most consistent with a diagnosis of meningitis?
 - a. Bulging fontanel
 - b. Positive Brudzinski's sign
 - c. Nuchal rigidity
 - d. Positive Kernig's sign
14. Diagnostic criteria for Tourette syndrome include which of the following?
 - a. Must have both motor and vocal tics
 - b. Steadily increasing pattern of motor tics
 - c. Presence of any tic for 6 months
 - d. Multiple motor tics with significant voluntary control
15. Which of the following statements about Tourette syndrome is not true?
 - a. It is the most severe type of tic disorder
 - b. It is more common among males than females
 - c. Mean age at onset is between 6 and 7 years
 - d. Symptoms decrease after puberty
16. A prominent feature in simple partial seizures is:
 - a. Very short postictal drowsiness
 - b. Microcephaly
 - c. Bilateral tremors
 - d. No loss of consciousness

17. Absence seizures:
 - a. Often begin between 1 and 2 years of age
 - b. Appear as altered awareness and blank stare for brief period
 - c. More commonly occur in first-born children
 - d. Usually progress to a more severe seizure disorder beyond childhood
18. The differential diagnosis for seizures includes which of the following:
 - a. Poliomyelitis without paralysis
 - b. Schizophrenia
 - c. Complicated migraine headaches
 - d. Multiple sclerosis
19. The most useful diagnostic tool for diagnosing epilepsy is:
 - a. MRI
 - b. EMG
 - c. EEG
 - d. CT scan
20. Which of the following is a primary drug used for treatment of generalized tonic-clonic seizures?
 - a. Ethosuximide
 - b. Valproic acid
 - c. Clonazepam
 - d. Corticosteroids
21. Which of the following is included in the diagnostic criteria for migraine headache in postpubertal children?
 - a. Recurrent abdominal pain, throbbing head pain not relieved by sleep
 - b. Nausea and vomiting, positive family history, aura
 - c. Positive family history, vomiting without nausea, pain not relieved by sleep
 - d. Aura, positive family history, pain in both hemispheres of the brain relieved by sleep
22. The type of head injury that is easily treatable but potentially the most lethal is:
 - a. Concussion
 - b. Epidural hematoma
 - c. Acute subdural hematoma
 - d. Linear skull fracture
23. Tuberous sclerosis is a progressive, neurocutaneous syndrome in which there is a combination of:
 - a. Skin abnormalities, blindness, and cognitive deficits
 - b. Skin abnormalities, seizures, cognitive deficits
 - c. Cerebral palsy, cognitive deficits, and skin problems
 - d. Skin abnormalities, blindness, and cognitive deficit
24. Inheritance of tuberous sclerosis is by:
 - a. Autosomal dominant mutant gene
 - b. Fragile X chromosome
 - c. Autosomal recessive mutant gene
 - d. Trisomy 23
25. The most important diagnostic measure for identifying tuberous sclerosis is:
 - a. EEG
 - b. Thorough neurological physical exam
 - c. MRI
 - d. Skin biopsy
26. For children 1 to 4 years of age, the most common cause of head injury is:
 - a. Motor vehicle accidents
 - b. Falls
 - c. Child abuse
 - d. Tricycle accidents
27. Dwayne, a senior in high school, suffers a concussion playing football. Immediately following the event, a typical clinical picture would include:
 - a. Brief loss of consciousness or/and amnesia
 - b. Confusion and lethargy lasting about 2 hours
 - c. Slowed and sometime unequal pupil response
 - d. Associated Battle sign
28. A head injury in which bruising or tearing of the cerebral structures occurs is a:
 - a. Contusion
 - b. Concussion
 - c. Hematoma
 - d. Diffuse axonal injury
29. The most common type of skull fracture in children with an excellent prognosis is:
 - a. Depressed
 - b. Basilar
 - c. Linear
 - d. Simple
30. Which of the following is most likely to be the etiologic cause of meningitis in the newborn?
 - a. Neisseria meningitidis
 - b. Escherichia coli
 - c. Streptococcus pneumoniae
 - d. Haemophilus influenzae B

31. Drug therapy commonly used to treat absence seizures, in addition to valproic acid, includes:
- Phenytoin
 - Phenobarbital
 - Carbamazepine
 - Benzodiazepines

■ ANSWERS

- | | |
|-------|-------|
| 1. a | 17. b |
| 2. d | 18. c |
| 3. d | 19. c |
| 4. c | 20. b |
| 5. b | 21. b |
| 6. a | 22. b |
| 7. a | 23. b |
| 8. c | 24. a |
| 9. a | 25. c |
| 10. d | 26. b |
| 11. b | 27. a |
| 12. c | 28. a |
| 13. a | 29. c |
| 14. a | 30. b |
| 15. d | 31. c |
| 16. d | |

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Genitourinary/Gynecological Disorders/Adolescent Pregnancy

Marlo Eldridge

MaryLou Rosenblatt

■ URINARY TRACT INFECTION (UTI)

- Definition: A generic term referring to the presence of bacterial infection of the urinary tract including the bladder (cystitis), urethra (urethritis), or kidney (pyelonephritis)
- Etiology/Incidence
 1. Multifactorial etiology including agent virulence and host predisposing factors
 - a. Agent virulence
 - (1) *Escherichia coli*—pathogenic agent in 80% to 90% of all childhood UTIs
 - (2) Other enteric bacteria agents—*Klebsiella*, *Enterobacter* sp.
 - (3) *Staphylococcus saprophyticus*—common in males
 - (4) Viral agents rare with exception of adenovirus
 - b. Host predisposing factors
 - (1) Immature kidneys associated with premature and low birth weight infants
 - (2) Congenital urologic abnormalities, reflux, neurogenic bladder
 - (3) Gender differences in anatomy of urinary tract predisposes females, e.g., short urethra and close proximity to anus
 - (4) Dysfunctional voiding—urinary stasis/incomplete bladder emptying
 - (5) Functional obstruction—constipation, pregnancy
 - (6) Trauma/irritants—catheterization, bubble baths, sexual intercourse, sexual abuse, pinworms
 2. Incidence—most common pediatric urinary tract problem
 - a. Newborns—2 per 100 live births
 - b. Accounts for 4.1 to 7.5% of infant febrile episodes
 - c. Infancy—increased incidence in males (2.7% of males and 0.7% of females have experienced an episode of bacteriuria by the time they reach one year of age)
 - d. Increased incidence among females after infancy and through adolescence, affecting less than 1% of school age boys and 1 to 3% of school-age girls
- Signs and Symptoms
 1. May be asymptomatic or with nonspecific symptoms, especially in infancy
 2. Symptom clusters by age group
 - a. Newborns—irritability, poor feeding, diarrhea, fever, vomiting
 - b. Infants/preschoolers—diarrhea, vomiting, fever, poor feeding, strong/foul-smelling urine
 - c. School age/adolescents—fever, vomiting, strong/foul-smelling urine, suprapubic or urethral pain, frequency, dysuria, and incontinence
- Differential Diagnosis
 1. Acute abdomen—appendicitis, sexually transmitted diseases, ectopic pregnancy

2. Chemical irritation—soaps, bubble baths
 3. Vulvovaginitis
 4. Dysfunctional voiding—enuresis
 5. Sexual abuse
 6. Foreign body
 7. Pelvic inflammatory disease (PID)
 8. Dysfunctional elimination syndrome (DES) —dysfunctional voiding accompanied by constipation
- Physical Findings
 1. May be normal
 2. Infancy—weight loss, poor feeding, diarrhea
 3. Fever and irritability
 4. Blood pressure may be elevated—reflux nephropathy
 5. Abdominal examination—pain, tenderness, guarding
 6. Urethral or vaginal irritation/discharge—with vulvovaginitis due to irritation or STD
 - Diagnostic Tests/Findings
 1. Urine analysis—presence of urinary leukocyte esterase, nitrate, and blood suggestive of UTI, but it is not diagnostic
 2. Urine culture mandatory for accurate diagnosis—technique for specimen collection depends on age and developmental status of child, severity of condition, and urgency of need for unequivocal results
 - a. Random voids/bagged urine—minimal usefulness due to high potential for contamination from external genitalia; 93% false positives; most useful for exclusion of the diagnosis of bacteriuria but not appropriate for culture and sensitivity
 - b. Clean-catch midstream—often contaminated from external genitalia, especially in young girls; more reliable from circumcised males and older girls
 - (1) Appropriate for mild symptoms or follow-up
 - (2) Positive with colonies > 100,000 colony forming units (CFU)/mL of single organism
 - c. Straight catheterization—used with infants/children who cannot void voluntarily; lower risk for nosocomial infection than indwelling catheters
 - (1) Appropriate for moderate or severe symptoms
 - (2) Positive with colonies > 10,000 CFU/mL of single or multiple organisms
 - d. Suprapubic aspiration—used with infants/children unable to void voluntarily when culture is urgently needed due to severity or equivocal results from alternative techniques
 - (1) Appropriate for moderate or severe symptoms
 - (2) Positive with colonies > 1000 CFU/mL of single or multiple organisms
 - Management/Treatment
 1. Antibiotic treatment
 - a. Parenteral antibiotics—newborns, infants, or older children with vomiting or severe symptoms, systemic illness, fever, or unable to take fluids
 - b. Oral antibiotic
 - (1) Generally treated for 10-day regimen
 - (2) Can be extended to 14-day regimen in complicated infections
 - (3) Single dose regimen remains controversial
 - c. First-line drugs of choice
 - (1) Trimethoprim-sulfamethoxazole (TMP/SMX)—infants > 2 months of age
 - (a) TMP 6 to 10 mg/kg/day + SMX 30 to 60 mg/kg/day bid

- (b) Recommended until sensitivities are available since most UTIs are caused by *E. coli*
- (2) Amoxicillin—30 to 50 mg/kg/day tid
- (3) Amoxicillin/clavulanate—40 mg/kg/day tid
- (4) Sulfisoxazole—150 mg/kg/day qid
- (5) Cephalexin—50 mg/kg/day tid
- (6) Nitrofurantoin—5 to 7 mg/kg/day in divided doses; ideal for treatment of bladder infections and it is highly concentrated in the urine, but less effective for systemic/renal infections as it does not concentrate well in the blood
- 2. Follow-up urine cultures
 - a. Second culture at 72 hours after initiating treatment if symptoms are not resolving
 - b. Culture one week after completion of treatment when “test of cure” is indicated
 - c. Close monitoring of periodic urine cultures with recurrent infections and/or unexplained fevers
- 3. Prophylactic antibiotics—vesicoureteral reflux (VUR); trimethoprim-sulfamethoxazole, one half daily dose, usually at bedtime until reflux resolves spontaneously, or via endoscopic/surgical intervention and is proven with a negative VCUg
 - a. Most VUR grades I to III resolve as child grows if there is no underlying dysfunctional voiding or dysfunctional elimination syndrome
 - b. VCUg to assess status of reflux every 12 to 18 months
- 4. Education/prevention
 - a. Increased fluid intake
 - b. Frequent voiding with complete emptying of bladder
 - c. Good perineal hygiene with front-to-back wiping
 - d. Avoid bubble baths and other urethral irritants

■ ENURESIS

- Definition: Involuntary urination after child has reached age when bladder control is usually attained; may occur during daytime (diurnal) or at night, especially while sleeping (nocturnal); usually resolves by 5 to 7 years of age
 - 1. Primary enuresis—child has never attained control
 - 2. Secondary enuresis—recurrence of incontinence following a period of at least 6 months of dryness
- Etiology/Incidence
 - 1. Many causes suggested
 - a. Primary—small bladder capacity; toilet-training problems; delayed maturation of voiding inhibitory reflex; sleep problems (“deep sleeper”); lack of inhibition of antidiuretic hormone (ADH); ingestion of increased amounts of fluid; dysfunctional voiding (inattention/too busy to void)
 - b. Secondary
 - (1) Diseases—UTI, diabetes, GU abnormalities
 - (2) Medications—e.g., theophylline, diuretics
 - (3) Family disruptions, stress
 - 2. Primary enuresis most common form in children—75% to 80%
 - 3. Over 12 years of age—50% have secondary enuresis
 - 4. Familial predisposition
 - a. One parent—44% increased risk
 - b. Both parents—77% increased risk
- Signs and Symptoms
 - 1. Bedwetting or daytime urine leakage
 - 2. Odor of urine in clothing
 - 3. May have withdrawal/isolation from peers, diminished self-esteem
- Differential Diagnosis
 - 1. UTI
 - 2. GU anomalies—ectopic ureter
 - 3. Mechanical obstruction
 - 4. Dysfunctional voiding
 - 5. Dysfunctional elimination syndrome—constipation
- Physical Findings: Genitalia
 - 1. Hypospadias, epispadias
 - 2. Labial fusion
 - 3. Dribbling of urine during examination
- Diagnostic Tests/Findings
 - 1. Urinalysis/urine culture—to rule out UTI or hypercalciuria
 - 2. Renal ultrasound/vesicoureterogram—with abnormal urine studies; GU anomaly on examination
- Management/Treatment
 - 1. Primary nocturnal
 - a. Limit fluid intake after dinner
 - b. Double voiding before bedtime
 - c. Avoid punishment/criticism
 - d. Usually self-limited; spontaneous resolution of 10% per year after 5 years of age

2. Motivational therapy
 - a. May be unsuccessful as exclusive treatment
 - b. Verbal praise for dryness
 - c. Reward system
 - d. Dryness calendar
3. Conditioning therapy—enuresis alarm
 - a. Triggered by urine
 - b. Children awakened by alarm
 - c. Alarm sensitizes child to sensation of full bladder
 - d. Restrictions
 - (1) Expensive—not covered by all insurance plans
 - (2) Treatment takes 2 to 3 months, often up to 6 months for greater success rate; relapse rate is high following discontinuation
 - (3) May awaken other family members
 - (4) Child's age, motivation, cooperation, family support important success factors
 - e. Pharmacologic treatment
 - (1) Desmopressin acetate
 - (a) Synthetic analog of antidiuretic hormone vasopressin
 - (b) Dose—oral tablet (0.2 mg to 0.6 mg, 1 hour before bedtime)
 - (c) Rapid response—1 to 2 weeks once initiated
 - (d) Increase or decrease according to response—dose is not weight based
 - (e) Relapse 90% once discontinued if ADH production has not increased with child's maturation
 - (f) Side effects—headache, congestion, nasal irritation, epistaxis
 - (2) Imipramine (use is controversial due to the potentially serious, dose-related adverse effects, including depression, seizures, cardiac arrhythmias. An EKG is recommended before beginning therapy.)
 - (a) Tricyclic antidepressant; unclear mechanism of action; may depress bladder contractions
 - (b) Used in children over 6 years of age
 - (c) Dose—0.9 to 1.5 mg/kg/day, 1 to 2 hours before bedtime
 - (d) Results seen within a few weeks
 - (e) Treat 3 to 6 months, then taper
 - (f) Relapse after treatment—75% to 90%
 - (g) Side effects—arrhythmias, sedation, dry mouth

4. Secondary enuresis
 - a. Evaluation of underlying etiology—disease process, medication, aggressive/interdisciplinary treatment of dysfunctional voiding/DES
 - b. Therapeutic intervention for individual/family stress

■ CRYPTORCHIDISM (UNDESCENDED TESTES)

- Definition: Absence of one or both testes in scrotal sac due to failure of normal descent from abdomen during fetal development
- Etiology/Incidence
 1. Normal fetal descent of testes
 - a. Hormonal mediation of normal testicular descent
 - (1) Abdominal descent to inguinal ring—12 to 14 weeks gestation
 - (2) Inguinal descent into scrotum—28 to 36 weeks
 - b. Increased incidence among premature infants secondary to gestational development
 2. Failure of normal descent associated with hormonal imbalance, chromosomal abnormalities, structural disorders
 3. May be unilateral (usually right-sided) or bilateral
 4. Incidence
 - a. Common (20% to 30%) among premature male births with birth weight < 1500 g
 - b. Lower incidence (3% to 5%) among full-term male infants
 - c. Incidence decreases to approximately 1% by one year due to spontaneous descent in approximately 75% of full-term and up to 95% of preterm cases
- Signs and Symptoms
 1. May be asymptomatic
 2. Family history of undescended testes
 3. Testes may be palpable or nonpalpable
- Differential Diagnosis
 1. Retractable testes
 2. Ectopic testes
 3. Anorchia
 4. Chromosomal disorders
- Physical Findings
 1. Palpable testes—may be retractile or ectopic
 2. Nonpalpable testes—may be abdominal or absent
 3. Presence or absence of testes should always be documented

- Diagnostic Tests/Findings
 1. Unilateral—usually none
 2. Bilateral nonpalpable testes
 - a. Karyotyping for chromosomal abnormalities
 - b. Follicle-stimulating and luteinizing hormones may suggest anorchia
 - c. Imaging studies occasionally utilized
- Management/Treatment
 1. Routine assessment at each well-child visit during first year of life; most spontaneous descents occur by 6 months
 2. Refer to urologist if undescended by one year
 - a. Hormonal therapy—human chorionic gonadotropin (hCG); gonadotropin-releasing hormone (GnRH) currently used in Europe but not yet approved for use in U.S. (response rates = hCG 19%, GnRH 21%, compared to 4% in placebo)
 - b. Surgical intervention—orchidopexy usually recommended when child is between 12 and 18 months
 3. Family education and support regarding potential complications
 - a. Infertility—greater risk in bilateral cryptorchidism
 - b. Testicular malignancy—up to 40 times increased risk
 - c. Hernia

■ HYDROCELE

- Definition: Painless scrotal swelling due to collection of peritoneal fluid within the tunica vaginalis surrounding the scrotum
 1. Noncommunicating type—tunica vaginalis is closed, limiting fluid collection to scrotum; size of hydrocele is constant
 2. Communicating type—tunica vaginalis remains open, allowing fluid to flow between peritoneum and hydrocele sac; often associated with hernia
- Etiology/Incidence
 1. Incomplete closure of processus vaginalis, which usually isolates tunica vaginalis from peritoneum
 2. Most common cause of painless scrotal swelling; uncertain incidence
- Signs and Symptoms
 1. Swelling in scrotum—alternating or fixed
 - a. Usually asymptomatic
 - b. May become painful if full or tense, secondary to coughing or straining
 - c. Variable size with child's state; larger when active/distressed, decreases with rest

- d. Smaller on awakening—enlarges as day progresses

- Differential Diagnosis
 1. Cryptorchidism
 2. Retractable testes
 3. Hernia
 4. Inguinal lymphadenopathy
 5. Patent processus vaginalis
- Physical Findings
 1. Scrotal swelling or asymmetry—tense appearance; scrotal skin normal, nontender
 2. Fluctuance
 3. Translucent with transillumination
- Diagnostic Tests/Findings: Abdominal ultrasound to differentiate hydrocele from hernia
- Management/Treatment
 1. Noncommunicating
 - a. Most resolve spontaneously without intervention
 - b. Refer for evaluation
 - (1) Persists beyond one year
 - (2) Significant increase in size
 - (3) Causes discomfort
 2. Communicating
 - a. Occasional spontaneous resolution
 - b. Frequently develops into hernia requiring surgical intervention
 - c. Refer for surgical evaluation if persists beyond one year

■ HYPOSPADIAS

- Definition: Congenital defect with urethral meatus on ventral surface of penis
- Etiology/Incidence
 1. Urethral folds along midline fail to fuse
 2. Common disorder—1 in 300 male births
 3. Occurs more frequently in Caucasians
 4. Both genetic and environmental factors have been implicated; a positive family history increases risk to 1 in 100 to 1 in 80 births
- Signs and Symptoms: Deflected urinary stream
- Differential Diagnosis: Ambiguous genitalia; female masculinization
- Physical Findings
 1. Ectopic opening of the urethral meatus
 - a. Anterior—glans, corona, anterior shaft
 - b. Midshaft
 - c. Posterior—scrotal, penoscrotal junction, posterior shaft

2. Inguinal hernia, undescended tests, incomplete foreskin
 3. Chordee—ventral curvature of penis due to fibrous band of tissue
- Diagnostic Tests/Findings
 1. Radiography if meatus in perineum (severe)
 2. Karyotype for chromosomal analysis
 - Management/Treatment
 1. Avoid circumcision—foreskin used for repair
 2. Refer to pediatric urology for evaluation and planning
 2. Mild cases—primarily cosmetic surgery
 3. Increasing severity—functional, psychological, and cosmetic surgery
 - a. Repair early—6 to 18 months
 - b. Family preparation for procedure and expected results

■ PHIMOSIS AND PARAPHIMOSIS

- Definition
 1. Phimosis—narrow, nonretractile foreskin of childhood; not fully retractable to expose glans
 - a. Newborns normally have adhesions, glans to foreskin
 - b. May not be fully retractable until 10 years of age or older
 2. Paraphimosis—inability to replace foreskin over glans after retraction
- Etiology/Incidence
 1. Primary phimosis physiologically normal
 2. Secondary phimosis due to a scarring process such as balanitis xerotica obliterans
 3. Phimosis may be due to congenital narrowing/tightness
 4. Phimosis may be due to inflammation/infection under foreskin
 5. Paraphimosis may be due to forcible retraction of foreskin for “cleaning” purposes
- Signs and Symptoms
 1. May be asymptomatic
 2. Painful urination
 3. Weak urine stream
 4. Pain/tenderness with paraphimosis
 5. Ballooning of foreskin when urinating; may be normal if voiding uncompromised
- Differential Diagnosis
 1. Balanitis (inflammation of glans penis)
 2. Balanoposthitis (inflammation of glans penis and prepuce)

- Physical Findings
 1. Phimosis—unretractable foreskin
 2. Paraphimosis—edema/discoloration of foreskin and glans
- Diagnostic Tests/Findings—none indicated
- Management/Treatment
 1. Maintain good hygiene
 2. Gentle stretch of foreskin during bath—advise family against forceful retraction; scarring and balanitis may occur
 3. Paraphimosis—goal is reduction of swelling to reduce foreskin; may be accomplished with ice, application of granulated sugar to the penis, or wrapping distal penis in saline soaked gauze and applying pressure for 5 to 10 minutes; will occasionally inject hyaluronidase beneath the band to release it; rarely a surgical emergency
 4. Surgery—circumcision in phimosis with urinary obstruction

■ MEATAL STENOSIS

- Definition: Narrowing of distal end of urethra
- Etiology/Incidence
 1. Post-circumcision—11%
 - a. Mechanical irritation by diaper
 - b. Ischemia from frenular artery damage during procedure
 - c. Inflammation secondary to dermatitis
 2. Almost never seen in uncircumcised males
- Signs and Symptoms
 1. Penile pain/discomfort with urination
 2. Narrow, dorsally diverted urine stream
 3. High velocity urine stream
 4. Occasional bleeding following void
- Differential Diagnosis
 1. Hypospadias
 2. Chordee
- Physical Findings
 1. Inflammation of glans
 2. Slit-like or narrowed meatus—best to observe urination; appearance alone may be misleading
- Diagnostic Tests/Findings: None
- Management/Treatment
 1. Air exposure
 2. Warm soaks/baths

3. Frequent diaper changes
4. Meatotomy may be necessary in some cases
5. Prevention
 - a. Care exercised at circumcision to avoid damage to frenular artery
 - b. Cover glans following procedure—petrolatum gauze commonly used
 - c. Observe for early sign of irritation/inflammation

▣ TESTICULAR TORSION

- Definition: Torsion of the spermatic cord; can result in gangrene of testes (emergency)
- Etiology/Incidence
 1. Abnormal fixation of testis to scrotum—permits testis to twist/rotate; impedes lymphatic and blood flow
 2. Not unusual to awaken with pain, but can also develop after scrotal trauma or increased activity
 3. Most common in adolescent males
- Signs and Symptoms
 1. Acute, painful swelling of scrotum
 2. Nausea, anorexia, vomiting
 3. Minimal fever, if any
 4. Lack of urinary symptoms is the norm
- Differential Diagnosis
 1. Trauma
 2. Orchitis
 3. Acute epididymitis
 4. Hydrocele
- Physical Findings
 1. Enlarged, highly tender testis
 2. Scrotum on involved side edematous, warm, erythematous
 3. Anxious patient, resistant to movement
 4. Lifting testis does not relieve pain (Prehn's sign)
 5. Solid mass may be visualized with transillumination
- Diagnostic Tests/Findings
 1. Complete blood count (CBC)—may see slight increase in white blood count
 2. Doppler ultrasound—reveals diminished blood flow
 3. U/A—often normal, but leukocytosis may develop rapidly
- Management/Treatment: Immediate referral for surgery

1. Emergently performed within first 6 hours—preservation of fertility great concern; prevention of atrophy and abscess
2. Untreated torsion can lead to testicular loss

▣ LABIAL ADHESIONS (LABIAL FUSION, SYNECHIA VULVAE, LABIAL AGGLUTINATION)

- Definition: Generally benign fusion of labial minora
- Etiology/Incidence
 1. Results from tissue irritation/inflammation and hypoestrogenization of labia minora
 2. Potential sources of irritation—trauma, superficial infection, poor hygiene (damp skin), sexual abuse
 3. Incidence—rarely present at birth (newborns are spared due to maternal estrogen); usually occurs after 2 months of age
 - a. Estimated incidence is 10% to 20% of all girls in first year
 - b. Highest incidence between 2 and 6 years of age, but may occur any time up to menarche
- Signs and Symptoms
 1. Generally asymptomatic
 2. Parental concern regarding potential anatomic abnormality
 3. Difficulty voiding, general discomfort
 4. Enuresis—primarily diurnal
 - a. Pooling of urine behind adhesion after voiding may occur depending upon degree of meatal obstruction
 - b. Results in dribbling of urine throughout the day
- Differential Diagnosis
 1. Intersex anomalies
 2. Imperforate hymen
 3. Genital scarring
- Physical Findings: Thin, flat, membrane of variable length found midline extending from clitoris to posterior fourchette when labia majora are gently separated
 1. Complete fusion—entire vestibule covered; may see pinpoint opening
 2. Partial fusion—much of genital structures visible
- Diagnostic Tests: None indicated

- Management/Treatment
 1. In most cases, parental reassurance and observation for resolution without intervention
 2. Previous practice of mechanical lysis no longer recommended due to high frequency of refusion
 3. Observation for UTI symptoms
 4. Topical application of conjugated estrogen cream twice a day for 2 to 3 weeks results in separation within 8 weeks in 90% of cases
 - a. Overuse may stimulate signs of precocious puberty, which resolve when cream is discontinued
 - b. Transient hyperpigmentation of labia may occur during treatment
 - c. Following separation
 - (1) Maintain good hygiene
 - (2) Topical applications of bland creams or petroleum jelly
 5. Inspection of vulvae on routine well-child visits to monitor baseline anatomy, hygiene, sexual development, and detect problems

■ VULVOVAGINITIS

- Definition: Perineal inflammation and/or infection of the vulva (vulvitis) or vagina (vaginitis); often associated with vaginal discharge, vaginal odor, vaginal itching/irritation
- Etiology/Incidence
 1. Sources of vulvovaginitis may be noninfectious or infectious
 2. Noninfectious vulvovaginitis
 - a. Chemical irritation—bubble bath, powder, detergents, soaps, over the counter (OTC) douches
 - b. Mechanical irritation—tight clothing, nylon underwear
 - c. Foreign body irritation—toilet tissue, retained tampon
 - d. Trauma/sexual abuse
 - e. Masturbation
 - f. Allergy to latex condoms
 3. Infectious vulvovaginitis
 - a. Nonspecific—bacterial overgrowth due to poor hygiene
 - b. Specific
 - (1) Bacterial—Group A beta hemolytic streptococcus, pneumococcus, enterococcus, *Shigella flexneri*/sonnie, *G. vaginalis*, *M. hominus*, *N. gonorrhoea*, *Chlamydia trachomatis*
 - (2) Viral—Herpes simplex virus (HSV), human papillomavirus (HPV)
 - (3) Parasitic—*Enterobius vermicularis* (pinworms); *Trichomonas vaginalis*
- (4) Mycotic/fungal—candidiasis (common following antibiotic use)
- 4. Uncertain incidence; 25% to 75% are noninfectious/nonspecific inflammation with normal flora
- Signs and Symptoms
 1. Vaginal discharge
 2. Genital discomfort/itching
 3. Dysuria/burning
 4. Erythema/edema of vulva or vagina
 5. Vaginal odor
- Differential Diagnosis
 1. Physiologic leukorrhea—thin, clear, or white discharge
 2. UTI
 3. Dermatologic disorders—psoriasis, seborrheic dermatitis, atopic dermatitis
- Physical Findings
 1. May have no physical findings
 2. Discharge
 - a. White to yellow—chemical, mechanical, *Chlamydia trachomatis*
 - b. Pale yellow to gray green—trichomoniasis
 - c. White, thick, cheesy—candidiasis (“yeast”)
 - d. Thin, white, frothy—bacterial vaginosis
 - e. Brown, bloody, foul odor—foreign body
 3. Genital erythema
 4. Lesions
 5. Perianal soiling
 6. Examination techniques—sensitive, gentle
 - a. Prepubertal female
 - (1) Pelvic examination usually deferred; visual inspection only
 - (2) Exploratory procedure under anesthesia may be needed for vaginal bleeding and should be referred
 - b. Pubescent female
 - (1) Pelvic examination—especially if sexually active
 - (2) Cervix best site for culture
 - (3) Wet mount and KOH may be obtained from vaginal pool secretions
- Diagnostic Tests/Findings
 1. Urinalysis for presence of WBC, trichomonads
 2. Tape test for pinworms
 3. Saline preparation for wet mount
 - a. Clue cells—bacterial vaginosis
 - b. Presence of WBC—may indicate bacterial vaginosis in the absence of clue cells/trichomonads
 - c. Trichomonads
 4. Potassium hydroxide (KOH) 10% preparation
 - a. Hyphae—candidiasis

- b. “Whiff test”—positive (fishy odor of bacterial vaginosis)
- 5. pH testing of vaginal secretions
 - a. pH of < 4.5 —normal or candidiasis
 - b. pH of > 4.5 —bacterial vaginosis, trichomonas
- Management/Treatment (see section on sexually transmitted diseases for specific management)
 1. Foundation of treating childhood vulvovaginitis is improvement of local perineal hygiene
 2. Nonspecific vaginitis often resolves without intervention
 3. Discontinue genital irritants—bubble bath, harsh bath soap and laundry detergents
 4. Cotton or cotton-lined underwear—avoid tight fitting clothing (tights, pants, undergarments)
 5. Bacterial vaginosis
 - a. Metronidazole—1 g/day orally in 2 divided doses for 7 days (adolescents/adults) and 15 mg/kg per day (children < 45 kg) with maximum dose of 1 g/day; OR metronidazole gel, 0.75%, 5 g (1 applicator) intravaginally daily for 5 days (adolescents) or 15 mg/k per day
 - b. Clindamycin cream 2%, 5 g (1 applicator) intravaginally at night for 7 days
 - c. Alternative treatments are less effective
 - d. Pregnant women should be treated for bacterial vaginitis
 6. Parasitic
 - a. Pinworms—mebendazole, pyrantel pamoate, or albendazole in single dose repeated in 2 weeks
 - b. Trichomonas vaginalis—metronidazole 15 mg/kg per day orally in 3 divided doses for 7 days (< 45 kg) with maximum of 1 g/day; OR 2 g orally in single dose or 1 g/day in 2 divided doses for 7 days (> 45 kg); partner must be treated
 7. Fungal—candidiasis
 - a. Oral agent—fluconazole 150 mg single dose
 - b. Various topical agents include clotrimazole, miconazole, butoconazole nitrate, terconazole, which are more effective than nystatin
 8. Foreign body
 - a. Prepubertal—attempt irrigation; warm saline via small feeding tube
 - b. Postpubertal
 - (1) Pelvic examination to locate object
 - (2) Moistened cotton-tip applicator or forceps for removal
 - c. Unsuccessful irrigation/pelvic or anxious child—refer for examination under general anesthesia

9. Positive cultures suspicious for sexual abuse or sexual activity—see section on sexually transmitted diseases
10. Physiologic leukorrhea
 - a. Educate about normal pubertal changes
 - b. Some benefit from “mini-pads” in underwear to absorb moisture/prevent wetness from staining clothing
 - c. Avoid use of douches and creams
11. Nonlatex condoms for suspected latex allergy

■ DYSMENORRHEA

- Definition: Pain during menstrual cycle; usually first 1 to 2 days; cramping discomfort felt mid-to-lower abdomen
 1. Primary dysmenorrhea—no pelvic abnormality, common in adolescents, usually develops 6 to 12 months after menarche, ovulation is necessary component
 2. Secondary dysmenorrhea—underlying pelvic pathology
 - a. Congenital anomalies (septate uterus)
 - b. Cervical stenosis or strictures
 - c. Cysts, tumors of ovary or uterus
 - d. Endometriosis
 - e. Pelvic inflammatory disease
- Etiology/Incidence (Primary Dysmenorrhea)
 1. Increased production of uterine prostaglandins; uterine contractions, ischemia
 2. Ovulation is required for development of primary dysmenorrhea
 3. Most common gynecological complaint
 4. 20% to 90% of adolescent women report dysmenorrhea
 - a. Significant limitation for 10% to 15% of females
 - b. School absenteeism—14% of those reporting dysmenorrhea frequently missed school
- Signs and Symptoms
 1. Pain usually starts with flow or several hours later, or may precede flow by several hours to 2 days
 2. Crampy/spasmodic pain, primarily lower abdominal area; may radiate to inner thighs, lower back
 3. Systemic symptoms
 - a. Nausea/vomiting/diarrhea
 - b. Lightheadedness/dizziness
 - c. Fatigue or general malaise
- Differential Diagnosis
 1. Reproductive system malformations
 2. Endometriosis

3. Pelvic Inflammatory Disease
 4. Psychogenic problems
- Physical Findings: May be none; defer pelvic examination only if adolescent is not sexually active
 1. Bimanual and rectovaginal exams indicated
 2. Cervical motion tenderness with PID
 - Diagnostic Tests/Findings
 1. Suspicion of PID—see section on sexually transmitted diseases
 2. Pelvic ultrasound for palpable masses or concern of GU abnormalities
 - Management/Treatment
 1. Primary dysmenorrhea—mild
 - a. Heat to abdomen
 - b. Exercise
 - c. Acetaminophen
 - d. Ibuprofen—400 mg orally immediately at onset of pain, then every 4 to 6 hours for 1 to 3 days; take with food, milk, antacid to avoid GI distress
 - e. Well balanced diet
 - f. Acknowledgment of symptoms, pain is real
 2. Primary dysmenorrhea—moderate to severe; unresponsive to treatment for mild disorder
 - a. Nonsteroidal anti-inflammatory drugs (NSAIDs)
 - (1) Inhibit prostaglandin synthesis
 - (2) Naproxen sodium—500 mg orally at onset, then 250 mg every 6 to 8 hours
 - (3) Mefenamic acid—500 mg orally at onset, then 250 mg every 6 to 8 hours
 - (4) Assess efficacy of NSAIDs after 3 to 4 cycles before using another medication
 - (5) NSAIDs contraindicated in clotting disorders, renal or peptic ulcer disease, pre-op patients, NSAID or aspirin allergy, aspirin induced asthma
 3. Severe dysmenorrhea—unresponsive to NSAIDs alone
 - a. Low-dose combination oral contraceptives (OC); effective in 90% of cases with severe pain
 - b. Minimum 3 to 4 cycles for symptom improvement
 - c. Continuous symptoms after 4 months
 - (1) OC used in conjunction with NSAIDs
 - (2) Consider gynecological referral
 4. Secondary dysmenorrhea
 - a. Begin PID treatment immediately, if indicated

- b. Gynecological referral if persistent dysmenorrhea after PID treatment or if pelvic pathology exists

■ PREMENSTRUAL SYNDROME

- Definition: Cluster of symptoms, physical, cognitive, and behavioral, that occur in second half of menstrual cycle (last week of luteal phase); usually resolve with onset of menses; symptoms exist over several cycles and cause disruption of normal activities.
- Etiology/Incidence
 1. Numerous mechanisms postulated
 - a. Vitamin deficiencies, inconsistent evidence
 - b. Fluid retention
 - c. Steroid hormone fluctuation
 - d. Alteration in serotonergic neuronal mechanisms
 - e. Inappropriate prostaglandin activity
 2. Studies note 14% to 61% adolescent women reporting at least one symptom
- Signs and Symptoms
 1. Onset of symptoms usually within 1 week of menses
 - a. Breast tenderness
 - b. Headache, muscle aches
 - c. Weight gain, bloating
 - d. Mood swings, lethargy, anxiety, irritability, depression
 - e. Fatigue
 - f. Appetite changes
 - g. Lower back pain
 - h. Loss of concentration
 - i. Acne
 - j. Constipation
 - k. Hot flashes, chills
- Differential Diagnosis
 1. Pregnancy
 2. Primary/secondary dysmenorrhea
 3. Premenstrual dysphoric disorder (PMDD), DSM-IV diagnosis for severe form of premenstrual syndrome (PMS); requires mental health referral
- Physical Findings: Pelvic examination normal
- Diagnostic Tests/Findings: None indicated
- Management/Treatment
 1. Diet/nutrition
 - a. Frequent small meals

- b. Increase intake—complex carbohydrates, protein, fresh fruits, vegetables, foods rich in pyridoxine (B₆)
 - c. Vitamin/mineral supplement—Vitamin B₆, magnesium
 - d. Limit intake—refined sugar, salt, red meat, alcohol, coffee, tea, chocolate
2. Lifestyle
 - a. Regular exercise (especially aerobic)
 - b. Stress management
 - c. Address psychosocial issues
 - d. Chart symptoms for 2 to 3 cycles to clarify/treat symptoms
 3. Pharmacological management
 - a. Diuretics
 - b. NSAID
 - c. Selective serotonin reuptake inhibitors (SSRIs), e.g., fluoxetine, sertraline, paroxetine—for adult women with diagnosis of PMDD
 - d. Oral contraceptive pills may reduce symptoms for some women; some women feel worse; a 24/4 formulation may improve symptoms more than a 21/7 formulation; OCPs may improve physical symptoms more than mental/emotional symptoms; close follow-up indicated

■ GENITOURINARY TRAUMA

- Definition: Injury to the genitourinary tract; refers to accidental injury (for nonaccidental trauma refer to section on sexual abuse)
- Etiology/Incidence
 1. Blunt insult generally from athletic activities, motor vehicle accidents, falls
 2. No specific incidence—commonly seen; over 50% associated with trauma to intraperitoneal organs
- Signs and Symptoms
 1. Frank urethral bleeding
 2. Hematuria
 3. Bluish-red mass in perineal area
- Differential Diagnosis
 1. Hemorrhagic cystitis
 2. Vaginitis
 3. Sexual abuse
- Physical Findings
 1. Hematomas—urethral/scrotal/perineal
 2. Periurethral lacerations
- Diagnostic Tests/Findings: Referral necessary if extensive injury suspected; radiographic evaluation of urinary tract is the cornerstone of treatment; usually done once referral to specialist has been made

- Management/Treatment
 1. Urethral/vulvar trauma
 - a. Mild bruising, superficial lacerations (symptomatic relief)—ice pack, sitz baths, analgesics
 - b. Blunt or penetrating trauma—surgical intervention
 2. Testicular trauma—surgical referral
 3. Suspected renal injury—referral
 4. Penetrating injury—immediate surgical exploration

■ GLOMERULONEPHRITIS (GN)

- Definition: Disease characterized by diffuse inflammatory changes in the glomeruli; immune-mediated response
 1. Primary acute form—poststreptococcal glomerulonephritis; most common form in children; true incidence unknown
 2. Primary chronic form
 - a. Primarily seen with IgA nephropathy
 - b. Other types—membranoproliferative glomerulonephritis (MPGN), mesangial proliferative glomerulonephritis
 3. Secondary forms—associated with other disorders, e.g., systemic lupus erythematosus, anaphylactoid purpura, vascular problems
- Etiology/Incidence
 1. Multifactorial etiology—not completely understood
 2. Combination of factors induce injury
 - a. Immune complex deposits in glomerular basement membrane
 - b. Coagulation factors—fibrin deposits
 - c. Exogenous nephrotoxins
 - (1) Penicillamine, trimethadione, captopril, probenecid
 - (2) Heavy metals—gold, mercury
 3. Uncertain incidence
- Signs and Symptoms
 1. Acute disease
 - a. Hematuria
 - b. Decreased urine output
 - c. Edema
 - d. Dark urine—acute poststreptococcal glomerulonephritis (APSGN)
 2. Chronic disease
 - a. Fatigue
 - b. Failure to thrive
- Differential Diagnosis
 1. Benign hematuria
 2. Hereditary nephropathy
 3. Systemic lupus erythematosus

4. Anaphylactoid purpura
 5. IgA nephropathy—most common GN leading to chronic renal failure worldwide
 6. Henoch-Schönlein purpura (HSP)
- Physical Findings
 1. May be asymptomatic or severely ill, depending upon extent of renal involvement
 2. Gross hematuria
 3. Edema—facial (especially periorbital) in the morning
 4. Hypertension—with or without renal insufficiency
 5. CVA tenderness
 - Diagnostic Tests/Findings
 1. Urinalysis
 - a. Casts—RBC, leukocytes, and/or casts indicate glomerular inflammation
 - b. Hematuria
 - c. Protein—correlates with degree of hematuria
 - d. pH—low
 - e. Specific gravity—increased
 2. Titers—serum ASO, AHT, anti-DNase B
 3. Cultures—throat, skin; may be negative by the time signs of nephritis appear
 4. Chest radiograph—assess pulmonary edema
 5. Serum complement
 - a. Returns to normal in APSGN
 - b. Chronic elevation in MPGN
 - Management/Treatment
 1. All treatment is supportive
 - a. Hypertension/relieve edema
 - (1) Fluid restriction
 - (2) Diuretics
 - (3) Vasodilators
 - b. Antibiotic (penicillin) if throat or skin infection persists

■ HYDRONEPHROSIS

- Definition: Unilateral or bilateral dilation of kidney(s)
- Etiology/Incidence
 1. Caused by anatomic block of urine flow from kidney
 2. Obstruction in 1 per 1000 births—slight male prevalence
 3. Ureteropelvic junction (UPJ)—most common site of obstruction
- Signs and Symptoms
 1. Nausea
 2. Abdominal or flank pain

3. Decreased urine output

- Differential Diagnosis
 1. Prune belly syndrome
 2. UPJ obstruction
 3. Ectopic ureterocele
 4. Urethral/ureterovesical obstructions
 5. Vesicoureteral reflux
 6. Posterior urethral valves
- Physical Findings
 1. Pain—abdominal/flank
 2. Failure to thrive
 3. May be asymptomatic in older children
- Diagnostic Tests/Findings
 1. May be detected during prenatal ultrasound
 2. IVP—late emptying of renal pelvis
 3. Renal scan—reveals impact of obstruction on total renal function
- Management/Treatment
 1. Surgery to relieve obstruction
 2. Obstruction will lead to destruction of renal parenchyma; early exploration and repair advocated
 3. Must follow-up long-term for continued assessment of renal function

■ RENAL TUBULAR ACIDOSIS (RTA)

- Definition: Defect in normal urine acidification with resulting persistent metabolic acidosis; primary RTA includes 2 types
 1. Type 1 (distal tube)—defect in distal tube secretion of hydrogen ions
 2. Type 2 (proximal tube)—defect in reabsorption of bicarbonate
- Etiology/Incidence
 1. Cellular basis of defect is unknown; distal RTA may have genetic transmission as autosomal dominant disorder
 2. Incidence of primary RTA is unknown
- Signs and Symptoms
 1. Growth failure
 2. Gastrointestinal complaints
 3. Muscle weakness
- Differential Diagnosis
 1. Diarrhea
 2. Diabetes mellitus
 3. Renal failure
 4. Lactic acidosis
- Physical Findings: Growth failure

- Diagnostic Tests/Findings
 1. Urine pH—first morning specimen; pH less than 5.5 supports diagnosis of proximal RTA; 5.8 or greater indicates distal RTA
 2. Serum electrolytes—serum bicarbonate less than 16 mEq; hyperkalemia
 3. Various low molecular weight proteins are used as markers
- Management/Treatment: Goals; achieve optimal growth and bone mineralization and prevent nephrocalcinosis and progression to renal failure
 1. Correction of acidosis; balance serum bicarbonate to normal level
 - a. Intravenous therapy for infants with severe hyperkalemia/acidosis
 - b. Oral therapy for most children
 2. Alkali administration as sodium bicarbonate or sodium citrate
 - a. Potassium supplement if needed
 - b. Sodium bicarbonate tablets—325 mg and 650 mg
 3. Mineralocorticoid deficiency corrected; diuretics reduce serum potassium
 4. Carnitine supplements if needed
 5. Risk of nephrocalcinosis, renal failure—continuous alkali therapy and long-term clinical monitoring
 6. Normal growth resumes with corrected acidosis

■ SEXUALLY TRANSMITTED INFECTIONS

Gonorrhea

- Definition: Acute infectious process primarily involving genital tract, anorectum, throat and ophthalmic epithelium
- Etiology/Incidence
 1. *Neisseria gonorrhoeae*—gram-negative diplococcus
 2. Reportable disease in all states; estimated 650,000 new cases per year in U.S.
 3. Women under 25 years of age are at the highest risk for gonorrhea infection
 4. Other risk factors include previous gonorrhea infection, other sexually transmitted infections, new or multiple sex partners, inconsistent condom use, commercial sex work, and drug use
 5. Associated with sexual abuse in children beyond newborn period and nonsexually active adolescents

- Signs and Symptoms
 1. Varies by site and gender; asymptomatic in 10% to 40% males and 50% to 80% females
 2. Vaginal or penile creamy discharge
 3. Perineal discomfort
 4. Menstrual irregularities
 5. Frequent, urgent, painful urination
 6. Rectal pain/itching
 7. Sore throat
 8. Fever, malaise, chills
- Differential Diagnosis
 1. Chlamydia trachomatis infection (may be concurrent)
 2. Genital mycoplasmas (may be concurrent)
 3. Bacterial vaginosis (may be concurrent)
 4. Trichomoniasis (may be concurrent)
 5. Pelvic Inflammatory Disease
- Physical Findings: Exam may be normal or include the following:
 1. External genitalia
 - a. Erythema, edema
 - b. Thick, purulent, greenish-yellow discharge (penile or vaginal)
 2. Female—pelvic examination
 - a. Cervical erythema, friability, exudate
 - b. Vaginal wall discharge/erythema
 - c. Cervical/adnexal tenderness
 3. Male
 - a. Thick, creamy penile discharge
 - b. Enlarged, tender prostate
 - c. Scrotal or groin pain (unilateral)
 - d. Tender swelling above testis
- Diagnostic Tests/Findings
 1. Gram stain for presence of gram-negative, intracellular diplococcus
 - a. Diagnostic in urethral specimen of symptomatic male; not recommended for asymptomatic men, or for endocervical, rectal, pharyngeal sites
 - b. Oldest and least expensive method
 - c. Basis of initiating treatment pending culture results
 2. Culture on selective media (Thayer-Martin) to confirm diagnosis
 - a. Specimen sites—vagina, endocervix, urethra, rectum, pharynx
 - b. Most sensitive and specific test
 3. Non-culture tests
 - a. Not recommended for use in prepubertal patients—false positive
 - b. Nucleic Acid Amplification Tests (NAATs) have high sensitivity and specificity for urine, male urethral, and cervical specimens (not rectal or pharyngeal)

4. No useful serologic test available to distinguish current from past infection
 5. Evaluate for possible concurrent syphilis, hepatitis B, HIV, and *Chlamydia trachomatis*
- Management/Treatment (see AAP *Red Book* for most current treatment guidelines)
 1. Uncomplicated infections
 - a. Children < 8 years weighing < 45 kg
 - (1) Ceftriaxone sodium, 125 mg, IM in single dose (drug of choice)
 - (2) Concomitant treatment for possible *Chlamydia trachomatis*; azithromycin, 20 mg/kg (maximum 1 g) orally in single dose; OR erythromycin, 50 mg/kg per day (maximum 2 g/day) orally in 4 doses for 14 days
 - b. Children/adolescents weighing > 45 kg and older than 8 years
 - (1) Ceftriaxone, 125 mg, IM in single dose (drug of choice); OR cefixime 400 mg oral, single dose
 - (2) Fluoroquinolones not recommended due to growing resistance
 - (3) Concomitant treatment for possible *Chlamydia trachomatis*—azithromycin, 1 g orally in single dose OR doxycycline, 100 mg orally bid for 7 days
 2. Complicated (disseminated) infections
 - a. Children < 8 years weighing < 45 kg
 - (1) Ceftriaxone, 50 mg/kg/day (maximum 1 g/day) in single dose (IV or IM) for 7 days
 - (2) Concomitant treatment for possible *Chlamydia trachomatis* with azithromycin or erythromycin
 - b. Children/adolescents weighing > 45 kg and older than 8 years
 - (1) Ceftriaxone, 1 g (IV or IM) in single dose for 7 days OR cefotaxime, 1 g (IV) every 8 hours for 7 days
 - (2) IV administration may change to oral antimicrobial agent such as ciprofloxacin (500 mg bid for 7 days) after 1 to 2 days of initial improvement
 - (3) Concomitant treatment for *Chlamydia trachomatis* with azithromycin, 1 g orally in single dose OR doxycycline, 100 mg, orally bid for 7 days
 3. Prophylaxis after sexual victimization
 - a. Antimicrobial prophylaxis no longer recommended for abused prepubertal victims
 - b. Prophylaxis for postpubertal females seen within 72 hours of sexual victimization
 - (1) Ceftriaxone, 125 mg IM in single dose
 - (2) Concomitant treatment for possible *Chlamydia trachomatis* with:
 - (a) Azithromycin, 20 mg/kg (maximum of 1 g) orally in single dose for patients weighing < 100 lb (45 kg)
 - (b) Azithromycin, 1 g orally in single dose for patients > 100 lb (45 kg)
 4. Prevention strategies
 - a. risk reduction education
 - b. condom use
 - c. routine screening for sexually active adolescents

Chlamydia

- Definition: Most common reported sexually transmitted infection in U.S. with primary sites of infection being genital tract, cornea, and respiratory system
- Etiology/Incidence
 1. *Chlamydia trachomatis*; obligate intracellular bacteria; at least eighteen variants
 2. Reportable disease most states
 3. Prevalence is consistently highest among adolescent females.
 4. Complications include endometritis, salpingitis, peri hepatitis, acute/chronic pelvic inflammatory disease (may result in ectopic pregnancy, infertility) for females; epididymitis, Reiter's syndrome for males
 5. Congenital chlamydia—See Chapter 15, Multi-system and Genetic Disorders
 - a. Perinatal transmission from infected mothers to infants estimated from 40% to 70%
 - b. Manifestation primarily as conjunctivitis or pneumonia
- Signs and Symptoms: Genital Tract Infection
 1. Often asymptomatic for months to years
 2. Abdominal/pelvic pain
 3. Dysuria/burning
- Differential Diagnosis
 1. Gonorrhea
 2. Genital mycoplasmas
 3. Trichomoniasis
 4. Bacterial vaginosis
 5. Pelvic Inflammatory Disease
- Physical Findings: Genital Tract Infection
 1. May be normal
 2. Erythema of external genitalia
 3. Vaginal/penile discharge—yellowish, watery
 4. Tenderness on bimanual examination

- Diagnostic Tests/Findings:
 1. Tissue culture—"gold standard" method for definitive diagnosis
 - a. Culture specimen must contain epithelial cells to be accurate
 - b. Only acceptable method for suspected child sexual abuse
 - c. Culture processing requires 2 to 3 days; results available in 3 to 7 days depending on laboratory
 2. Nucleic acid amplification tests—most sensitive screening tests for genital chlamydia infection
 - a. Include PCR (Polymerase chain reaction), TMA (transcription mediated amplification)
 - b. Specimens can be urine, urethral or cervical
 - c. Can detect gonorrhea as well
- Management/Treatment (See AAP, *Red Book* for additional treatment guidelines)
 1. Antibiotic treatment for uncomplicated genital tract infection
 - a. Adolescents—doxycycline, 100 mg bid for 7 days OR azithromycin 1 g in single dose
 - (1) Doxycycline is contraindicated in pregnancy
 - (2) Alternative treatment regimen in pregnancy
 - (a) Erythromycin base, 50 mg/kg/day qid for 7 days; may be given in half doses for 14 days if not tolerated well
 - (b) Amoxicillin 1.5 g/day in 3 divided doses for 7 days
 - b. Children (6 months to 12 years)—erythromycin, 50 mg/kg/day qid for 7 days OR azithromycin 20 mg/kg in single dose (not to exceed 1 gram)
 - c. Infants < 6 months of age
 - (1) Erythromycin, 50 mg/kg/day qid for 14 days
 - (2) Second course may be required
 - (3) Association between oral erythromycin and infantile hypertrophic pyloric stenosis (IHPS) has been reported in infants < 2 weeks of age which require informing parents of potential risks and careful follow-up
 - d. Identify, examine, test, and treat any sexual contacts
 2. Evaluate for other STDs, e.g., gonorrhea, syphilis, and treat as necessary
 3. No need for retest following treatment with doxycycline, azithromycin, unless symptoms persist or if possibility of reinfection

4. Retest may be recommended at 3 or more weeks following treatment with erythromycin, sulfisoxazole, or amoxicillin

Acquired Syphilis

- Definition: A contagious systemic infectious disease characterized by three progressive clinical stages
 1. Primary stage—painless chancres on skin or mucous membranes at site of exposure which may go unnoticed
 2. Secondary stage—begins between 1 to 2 months after inoculation, characterized by skin rash, cutaneous lesions, lymphadenopathy
 3. Tertiary stage—multisystem involvement that may occur from years after primary infection, including aortitis or gummatous changes of skin, bone, or viscera; neurosyphilis may occur at any stage of infection, particularly in HIV patients
- Etiology/Incidence
 1. Infectious agent—*Treponema pallidum*, a thin, motile spirochete
 2. Transmission—sexual contact, transplacental, direct contact with infected tissue (see Chapter 15, Multisystem and Genetic Disorders, for congenital syphilis)
 3. Reportable communicable disease in all states
 4. Increased incidence during later 1980s and early 1990s
 - a. Incidence has begun to decline except in large urban areas and rural south
 - b. 10% to 12% of all reported cases are among adolescents between 15 and 19 years of age
- Signs and Symptoms
 1. Primary stage—one or more painless lesions, usually on genitalia but may be on lips, tongue, or extremities
 2. Secondary stage—fever, malaise, sore throat, skin rash, hair loss
 3. Tertiary stage—symptoms recur years after initial infections; rarely seen among adolescents
 4. Neurosyphilis—may be asymptomatic; neurologic signs include fever, headache, photophobia, meningismus, cranial nerve palsies; less frequently confusion, delirium, and seizures
- Differential Diagnosis
 1. Primary syphilis
 - a. Genital herpes
 - b. Chancroid, Granuloma inguinale, lymphogranuloma venereum

- c. Traumatic lesion, excoriation
- d. Behcet's lesion
- 2. Secondary syphilis
 - a. Pityriasis rosea
 - b. Psoriasis
 - c. Condylomata acuminata
 - d. Drug sensitivity reactions
 - e. Infectious mononucleosis
 - f. Lupus erythematosus
- Physical Findings
 - 1. Primary stage
 - a. Chancre—one or more painless ulcers
 - b. Most common on genitalia but seen at other sites of inoculation
 - 2. Secondary stage
 - a. Generalized polymorphic maculopapular rash—classic if palms and soles included
 - b. Round to oval, reddish-brown, “copper-colored” lesions
 - c. Lymphadenopathy, arthralgia, fever, malaise, sore throat, headaches, splenomegaly
 - d. Hypertrophic papular lesions of vulva/anus—condyloma lata
 - 3. Latency period follows with recurrences of secondary rash
 - 4. Tertiary phase
 - a. 15 or more years after chancre
 - b. Aortitis, gummous changes of bone, skin, or viscera
 - c. Neurosyphilis, infection of the central nervous system
- 1. May occur at any stage
- 2. HIV positive persons at higher risk
- Diagnostic Tests/Findings
 - 1. Dark-field microscopic tests or direct fluorescent antibody tests (DFA)—presence of spirochetes from scrapings or washings of primary lesions; inexpensive, definitive diagnosis
 - 2. Serologic tests—presumptive diagnosis
 - a. Nontreponemal tests—rapid plasma reagin (RPR), venereal disease research laboratory (VDRL), automated reagin test (ART)
 - (1) Measure nonspecific antigens
 - (2) False-positive rate of 1% to 2%
 - (3) False-negatives with recently acquired infections prior to seroconversion, or latent syphilis of long duration
 - (4) Serial testing used to monitor response to treatment
 - b. Treponemal tests—fluorescent treponemal antibody absorption (FTA-ABS) and *Treponema pallidum* particle agglutination (TP-PA)
 - (1) Detect specific treponemal antigens
 - (2) Greater specificity than nontreponemal methods
- (3) More expensive and time-consuming
- (4) Useful to distinguish/confirm positive vs. false-positive nontreponemal results
- (5) May show false-positive results in presence of Lyme disease, acute infections, autoimmune disorders, and narcotic addiction
- c. High probability of infection in sexually active person with reactive nontreponemal and treponemal tests
- d. VDRL/RPR screening in early pregnancy and at delivery for all women to prevent/identify transplacental transmission
- Management/Treatment: See AAP *Red Book* for additional treatment guidelines
 - 1. Primary, secondary, and early latent syphilis (less than 1 year duration)
 - a. Children—penicillin G benzathine, 50,000 U/kg IM up to adult dose of 2.4 million units in single dose
 - b. Adults—penicillin G benzathine, 2.4 million units, IM in single dose
 - (1) If allergic to penicillin and NOT pregnant—doxycycline, 100 mg orally bid for 14 days OR
 - (2) Tetracycline, 500 mg orally qid for 14 days
 - 2. Late latent (more than 1-year duration) and tertiary syphilis
 - a. Children—penicillin G benzathine, 50,000 U/kg, IM up to adult dose of 2.4 million units, as 3 single doses administered at 1-week intervals
 - b. Adults—penicillin G benzathine 7.2 million units total administered as 3 doses of 2.4 million units IM per dose at 1-week intervals
 - (1) If allergic to penicillin and NOT pregnant—doxycycline, 100 mg orally bid for 4 weeks OR
 - (2) Tetracycline, 500 mg orally qid for 4 weeks
 - 3. Neurosyphilis
 - a. Children—aqueous crystalline penicillin G, 200,000 to 300,000 U/kg per day given every 4 to 6 hours for 10 to 14 days not to exceed adult dose
 - b. Adults—aqueous crystalline penicillin G, 18 to 24 million units per day in doses of 3 to 4 million units, IV every 4 hours for 10 to 14 days OR penicillin G procaine, 2.4 million units, IM once daily PLUS probencid, 500 mg, 4 times per day orally, both for 10 to 14 days
 - c. See *Red Book* for penicillin allergic regimens

4. Evaluation of patient and all recent sexual contacts for syphilis and other STIs
5. Prevention and Control
 - a. Patient education/discussion of sexuality, contraception, and STIs as part of adolescent well-child visits
 - b. Counseling regarding safe sexual practices including abstinence and proper use of condoms
 - c. Treatment of sexual contacts
 - d. Report each case to local health authorities for contact investigation
 - e. All women should be screened early in pregnancy and at delivery

Genital Herpes Simplex Virus (HSV)

- Definition: Most common HSV infection among adolescents characterized by clusters of painful lesions of the genital tract, perineum, mouth, lips, or pharynx
- Etiology/Incidence
 1. Agent (herpes simplex viruses)—large, DNA viruses of two major types
 - a. Type 2 (HSV-2)—primary source of genital herpes, usually, but not always, affecting skin below the waist
 - b. Type 1 (HSV-1)—less common source of genital herpes, usually, but not always, sites include face and skin above the waist
 2. Primary transmission through sexual contact and/or direct contact with open lesions; may be transmitted by autoinoculation of HSV-1 to genital area
 3. Transplacental transmission results in congenital herpes—see Chapter 15, Multisystem and Genetic Disorders
 4. Genital herpes is rare in prepubertal children except in cases of child abuse
 5. Estimated prevalence of 1.6% among sexually active adolescents
- Signs and Symptoms
 1. Painful genital lesions
 2. Burning with urination
 3. Tender, swollen lymph nodes
 4. Fever, malaise
- Differential Diagnosis
 1. Chancre of early syphilis
 2. Chancroid
 3. Lymphogranuloma venereum, granuloma inguinale
 4. Behcet's lesion
 5. Excoriation
 6. Allergic reaction
- Physical Findings
 1. Vesicular/ulcerated lesions—genital tract, perineum, mouth, lips, pharynx
 2. Genital, perianal erythema, and/or edema
 3. Cervical friability, discharge
 4. Lymphadenopathy
- Diagnostic Tests/Findings
 1. Tissue culture—standard, most reliable diagnostic test
 - a. Sensitivity varies by stage of disease—highest with vesicular lesions; lowest with recurrent infections and crusted lesions
 - b. Results available within 1 to 3 days
 2. Newer diagnostic techniques
 - a. PCR-DNA probe testing for CSF specimens—technique with good sensitivity and specificity; results available within 1 to 3 days
 - b. Direct fluorescent antibody/enzyme immunoassay—more rapid results than cultures, but less sensitive results
 - c. Polymerase chain reaction (PCR)—very sensitive; lower specificity resulting in false-positive results
 3. Serologic testing—may show rise in HSV antibodies; of limited value
 - a. May be used to confirm initial diagnosis
 - b. Often shows no rise in titers with recurrences
- Management/Treatment: See *AAP Red Book* for additional treatment guidelines
 1. Primary episode of genital infection
 - a. Acyclovir, 200 mg orally, 5 times/day for 7 to 10 days or 400 mg 3 times per day for 7 to 10 days
 - b. Initiation of treatment within 6 days of onset of lesions may reduce duration and severity of symptoms
 - c. Valacyclovir and famciclovir provide alternatives for adult (pediatric formulations not yet available) treatment with advantage of less frequent dosing
 2. Recurrent episodes—alternative doses/frequency
 - a. Acyclovir, 200 mg orally, 5 times/day for 5 days
 - b. Acyclovir less effective in treatment of recurrent vs. primary episodes
 - c. Valacyclovir and famciclovir are alternatives for recurrent episodes
 3. Topical acyclovir no longer recommended; limited benefit
 4. Suppressive therapy—with frequent recurrences of > 6 per year

- a. Acyclovir 400 mg, orally, bid; OR 200 mg, orally, 3 to 5 times/day
 - b. Discontinue after one year to reassess recurrences
5. Sitz baths may provide relief
 6. Education—recurrences, viral shedding, abstinence when lesions are present, use of condoms during sexual activities

Genital Warts (Condylomata Acuminata)

- Most common symptomatic viral reproductive tract infection in U.S.; characterized by epithelial warts/tumors of mucous membranes and skin
- Etiology/Incidence
 1. Causative agent—Human papillomavirus (HPV), a small DNA virus with more than 100 subtypes; types 6 and 11 usually cause genital warts; and types 16, 18, 31, 33, 35 cause vaginal, anal, and cervical dysplasia
 2. Primary mode of transmission is sexual contact; sexual abuse must be considered when present in prepubertal child
 3. Genital HPV infection may be as high as 40% among sexually active female adolescents
 4. Most infections are transient, and clear spontaneously
- Signs and Symptoms
 1. Firm bumps in anogenital area
 2. Occasional local symptoms—burning, pain, itching, bleeding
 3. Often asymptomatic
- Differential Diagnosis
 1. Chancre of secondary syphilis
 2. Molluscum contagiosum
- Physical Findings
 1. Firm, flesh-colored anogenital lesions resembling cauliflower in configuration
 2. Range in size from few millimeters to centimeters
 3. Males—warts on shaft of penis, meatus, scrotum, and perianal areas
 4. Females—warts usually seen on labia and perianal areas
- Diagnostic Tests/Findings
 1. Diagnosis usually based on clinical inspection; no culture is available
 2. Colposcopy to detect cervical lesions—application of 3% to 5% acetic acid (vinegar) causes lesion to blanch; not definitive
 3. Pelvic examination—Pap smear for cytological analysis may be diagnostic

4. Biopsy of lesion for histologic examination—may be diagnostic
5. DNA probe—may detect asymptomatic HPV infection

- Management/Treatment

1. No definitive treatment yet available to eradicate HPV virus; palliative treatment focusing on removal of lesions, symptom relief, and close follow-up for recurrences and sequelae
 2. Spontaneous resolution within 3 months in 25% cases; recurrences are common
 3. External visible lesions
 - a. Self-treatment
 - (1) Podophyllum resin solution or gel 0.5% (contraindicated in pregnancy and not yet tested for safety/efficacy in children)
 - (a) Topical application bid for 3 days; need not be washed off
 - (b) First application should be done in office to assure proper technique
 - (c) Treatment may be repeated up to 4 cycles with 4-day rest period between cycles
 - (2) Imiquimod
 - (a) 3 times per week \times 6 to 10 hours, then wash off
 - (b) Maximum is 16 weeks
 - b. Clinician applied
 - (1) Podophyllin, 10% to 25% in compound tincture of benzoin (contraindicated in pregnancy)
 - (a) Weekly treatment up to total of 6 applications
 - (b) Must be washed off in 1 to 4 hours
 - (2) Trichloroacetic acid (TCA 80% to 90%)
 - (a) Topical application followed by careful drying and application of talc or baking soda
 - (b) Weekly treatments up to total of 6 applications
 - (c) Causes more local discomfort than podophyllin
 - (3) Liquid nitrogen or cryotherapy
 - (4) Laser surgery, cryosurgery, excision, electrodesiccation—reserved for extensive, severe, and/or resistant cases
4. Gynecologic referral necessary for cervical warts
5. Monitoring of Pap smears for increased risk of cervical cancer
 - a. First Pap smear 3 years after first intercourse or 21 years old
 - b. Repeat annually if normal

- c. For immune competent women under 20 years old, ASCUS (atypical squamous cells of undetermined significance) and LSIL (low-grade squamous intraepithelial lesions), in one year; high-grade lesions (ASC-H and HSIL), refer for colposcopy; if remains ASCUS at 24 months, refer for colposcopy
 - d. For immune competent women 20 years and older and immune compromised women, repeat ASCUS without high-risk HPV in one year; if remains ASCUS at one year, refer to colonoscopy; refer ASCUS with high risk HPV, LSIL, HSIL, ASC-H for colposcopy
 - e. Annual Pap smears for all HPV positive asymptomatic females thereafter
 - f. Obtain Pap smear for suspicious cervical exam
6. Screening serology for syphilis; evaluate for any other concomitant STDs and treat accordingly
 7. Education regarding safer sexual practices
 8. Prevention—HPV vaccine (for types 6, 11, 16, 18) available for females aged 11 to 26 years, 3 injection series given over 6 months; no current vaccine recommendation for males

Trichomoniasis

- Definition: Common sexually transmitted infection of the genital tract
- Etiology/Incidence
 1. Causative agent—*Trichomonas vaginalis*, a flagellated protozoan
 2. Transmitted primarily through sexual contact; presence in prepubertal child should alert practitioner to possible sexual abuse
 3. Often associated with other STDs, e.g., gonorrhea, chlamydia
 4. Unknown incidence
- Signs and Symptoms
 1. Females—asymptomatic in 25% to 50% of all cases
 - a. Vaginal discharge
 - b. Vulvovaginal irritation and itching
 - c. Vaginal odor
 - d. Difficult urination
 2. Males—usually asymptomatic
 - a. Mild dysuria
 - b. Itching
- Differential Diagnosis
 1. Candidiasis
 2. Chemical vaginitis

3. Bacterial vaginosis
 4. UTI
 5. Poor hygiene
 6. Gonorrhea
- Physical Findings
 1. Vaginal discharge—frothy, light yellow to grey-green, musty odor
 2. Pelvic examination—evidence of vaginitis, cervicitis; erythema, edema, and pruritus of external genitalia may be present
 3. Males generally asymptomatic
 - Diagnostic Tests/Findings
 1. Wet mount of vaginal secretions or spun urine sediment—presence of motile trichomonads
 2. Motile trichomonads may also be seen on Pap smears and urine analysis
 3. Other tests—culture (trypticase yeast extract iron serum [TYI] medium); antibody tests using enzyme immunoassay; direct/indirect immunofluorescence techniques are available
 - a. More sensitive techniques than wet mount
 - b. Rarely needed to make diagnosis
 - Management/Treatment: See *AAP Red Book*, for additional treatment guidelines
 1. Metronidazole—treatment of choice
 - a. Prepubertal—15 mg/kg/day orally tid for 7 days (maximum 2 g for 7 days) or 40 mg/kg (maximum 2 g) orally in single dose
 - b. Adolescent—2 g orally in single dose or 500 mg 2 times per day \times 7 days
 - c. Avoid alcohol during treatment and for 48 hours after treatment
 - d. During pregnancy—2 g single dose of metronidazole or 7 day regimen
 2. No sexual activity during treatment
 3. Partners should receive concurrent therapy
 4. Evaluate for presence of other STI—treat accordingly

Bacterial Vaginosis (BV)

- Definition: Clinical syndrome characterized by vaginal symptoms, primarily in sexually active adolescents/adults
- Etiology/Incidence
 1. Not an actual infection but classified as a sexually transmitted disease
 2. Results from replacement of normal vaginal flora (*Lactobacillus*) with high concentrations of anaerobes—*Gardnerella vaginalis*, *Mycoplasma hominis*
 3. Transmission may be sexual or nonsexual
 4. Incidence unknown

- Signs and Symptoms
 1. Profuse vaginal discharge with “fishy” odor
 2. May be asymptomatic
 - Differential Diagnosis
 1. Foreign body
 2. Gonorrhea/chlamydia/trichomoniasis
 3. Vulvovaginitis—group A streptococci, shigella organisms
 4. Candidiasis
 5. Physiologic leukorrhea
 - Physical Findings
 1. Vaginal/cervical discharge—thin, white, malodorous; adherent to vaginal wall
 2. Itching, swelling, redness of external genitalia
 - Diagnostic Tests/Findings
 1. Vaginal secretions
 - a. pH greater than 4.5
 - b. KOH 10% mixed with vaginal discharge—releases amine, “fishy odor” (whiff test)
 - c. Saline wet mount—clue cells
 2. Culture available—rarely helpful, expensive
 - Management/Treatment (See *AAP Red Book* for additional treatment guidelines)
 1. Metronidazole
 - a. 500 mg, orally, bid for 7 days OR
 - b. 2 g orally, single dose with 2nd dose in 48 hours
 2. Alternative Treatment
 - a. Clindamycin cream 2%, one full applicator (5 g) intravaginally at bedtime for 7 days
 - b. Metronidazole gel 0.75%, one full applicator (5 g), intravaginally bid for 5 days
 - c. Clindamycin, 300 mg, orally, bid for 7 days
 3. Clindamycin recommended for pregnant women due to possible teratogenicity of metronidazole
 4. Education regarding possible complications
 - a. Avoid douching
 - b. Common recurrence
 - c. Treatment of male partner not recommended, does not decrease female recurrence
 - d. Increased risk for PID
 - e. Pregnancy risk for chorioamnionitis and premature delivery
 - Male Condoms—most effective barrier method
 1. Mechanism—mechanical barrier preventing semen from entering vagina; types of male condoms—over 100 brands currently on the market
 - a. Latex—recommended
 - b. Polyurethane (latex-free) if latex sensitive
 - c. Natural skin (lambskin)—not recommended (inadequate STD and HIV protection)
 2. Failure rates
 - a. Theoretical—3%
 - b. Actual—12%, primarily due to nonuse/misuse
 - c. Combined with spermicide—no more effective than lubricated condoms in protecting against STIs and HIV
 3. Risks/precautions
 - a. Breakage rate is approximately 1% to 2%
 - b. Only water-based lubricants should be used—latex breaks down in contact with petroleum-based products
 - c. Latex breakdown over time and/or when exposed to heat
 - Vaginal Spermicides—topical creams, jellies, foams, suppositories, and films to prevent pregnancy; used alone or in combination with condoms
 1. Active spermicidal agent—nonoxynol-9 or octoxynol-9
 2. Failure rate is approximately 6%
 3. Risks/precautions (CDC, 2006)
 - a. Not effective in preventing cervical gonorrhea, Chlamydia, or HIV
 - b. Frequent use may disrupt genital epithelium, may be associated with an increased risk of HIV transmission
 - c. Increased risk for UTIs in women using diaphragm/spermicide
 - d. Should not be used as a lubricant or microbicide for anal intercourse due to rectal cell damage
 - Diaphragm—female barrier contraceptive method
 1. Thin latex dome with flexible ring for vaginal insertion prior to intercourse; positioned with posterior rim on posterior fornix and anterior rim behind pubic bone
 2. Requires pelvic examination for proper fitting for appropriate size
 3. Failure rate among adolescents—10% to 25%
 4. Risks/precautions
 - a. Requires technical skill/comfort with body for correct placement
 - b. Must be kept in place for 6 hours post-intercourse
- CONTRACEPTION**
- Abstinence—most effective method
 1. Half of all adolescents select this option
 2. Should be discussed as viable option regardless of prior history

- c. Spermicidal agent must be used for subsequent intercourse
 - d. Side effects—UTI, vaginitis
- Combined Hormonal Contraceptives—oral contraceptives (OC), transdermal contraceptive patch, vaginal contraceptive ring
 1. Mechanisms
 - a. Prevents ovulation
 - b. Increases viscosity of cervical mucus inhibiting sperm penetration
 - c. Alters endometrium to resist implantation
 2. Health benefits
 - a. Decreased risk of endometrial and ovarian cancer
 - b. Improved androgen insensitivity
 - c. Decreased risk of hospitalization for GC-PID
 - d. Suppress endometriosis
 - e. Decrease iron deficiency anemia
 - f. Improve dysmenorrhea
 3. Oral contraceptive pills; estrogen-progestin combinations
 - a. Estrogens—ethinyl estradiol and mestranol
 - (1) Ethinyl estradiol—1.2 to 1.4 times stronger than mestranol
 - (2) Ethinyl estradiol effective at doses as low as 20 µg
 - (3) Usually select lowest effective estrogen dose (20 to 35 µg)
 - b. Basic progestins—norethindrone, norethindrone acetate, norethynodrel, ethynodiol diacetate, norgestrel
 - c. Newer progestins—norgestimate, desogestrel, gestodene
 - d. Monophasic, biphasic, or triphasic combinations—deliver constant or progressively increasing progestin during cycle
 4. Transdermal contraceptive patch
 - a. Ethinylestradiol/norelgestromin
 - b. Wear one patch × 1 week, repeat for 2 more weeks
 - c. Patch must be correctly applied and site rotated
 5. Vaginal contraceptive ring
 - a. Ethinylestradiol/etonorgestrel
 - b. Insert ring and remains in place × 3 weeks
 - c. Learn how to place and check for ring
 6. Failure rates—0.5% (theoretical) to 2% to 3% (actual)
 7. Risks/precautions
 - a. Drug interactions
 - (1) Drugs that may reduce OC effectiveness—antibiotics, antifungals, anticonvulsants, antacids
 - (2) Drugs that enhance OC effectiveness—ascorbic acid, co-trimoxazole
 - b. Major side effects—thrombosis
 - c. Minor side effects—vaginal spotting, nausea, bloating, irritability
 - d. Does not protect against HIV and other STIs
 - e. Absolute contraindications for use
 - (1) History of clotting disorder
 - (2) Impaired liver function
 - (3) Abnormal vaginal bleeding (undiagnosed)
 - (4) Pregnancy
 - (5) Estrogen-dependent carcinoma
 - f. Relative contraindications for use
 - (1) Severe hypertension
 - (2) Migraines
 - (3) Chronic diseases, e.g., diabetes, heart disease, sickle cell, etc.
 - (4) Rheumatologic disorders
 - Long-Acting Progestins (Injectable and Implantable to Inhibit Ovulation)
 1. Depo-medroxyprogesterone acetate (injectable)
 - a. Dosage and administration—150 mg/mL intramuscular injection every 3 months during first 5 days of normal period to assure nonpregnant status
 - b. Failure rate—0.25%
 - c. Precautions/risks
 - (1) If > 3 months between injections, pregnancy test as precaution
 - (2) Should not be given to postpartum, lactating adolescent mothers
 - (3) Spotting, weight gain, bloating, headaches, mood changes
 - (4) Risk of decreased bone density
 2. Implanon (Etonogestrel)
 - a. One rod implanted in subcutaneous tissue of upper, inner arm
 - b. Effective for 3 years
 - c. Vaginal bleeding and spotting are not predictable
 - d. Does not protect against STIs
 - IUD
 1. Safe, effective long-term contraception
 2. Copper T 380A increases uterine and tubal fluids that impair sperm function
 3. LNG-IUS (Mirena) releases levonorgestrel 20 mcg per day, which thickens cervical mucus, inhibits sperm, suppresses endometrium
 4. Reduces risk of ectopic pregnancy
 5. LNG-IUS decreases blood loss
 6. Fertility rebounds when discontinued
 7. Does not protect against STIs

- Emergency Contraception (EC)—Use of oral contraceptive within 72 hours of unprotected intercourse
 1. Evaluate for pregnancy risk
 2. EC will not terminate existing pregnancy
 3. Plan B is progesterone only (levonorgestrel), 2 tabs
 4. Schedule of doses from 20 OCPs brands, given in 2 doses 12 hours apart; may cause nausea, vomiting (see *Contraceptive Technology* 18th edition, p. 280)
 5. Meclizine HCl 25 to 50 mg may be given before first dose of OCP method to reduce nausea
 6. Provide counseling to prevent future need for EC
 7. Perform pregnancy test if no menses after 3 weeks
 8. Failure rate 1.6%
 9. Can give Plan B prescription in advance
- Contraceptive Methods Less Suitable for Adolescents
 1. Coitus interruptus—failure rate > 27%
 2. Natural family planning—high failure rate
 3. Cervical cap—difficulties with proper placement; risk of infection
 4. Progestin-only mini-pill—failure rates up to 13% among mature adults, increased risk of ectopic pregnancy when method fails
 5. Female condom—failure rates of 12% to 20% among adult users

■ ISSUES OF PREGNANCY AND BIRTH FOR THE ADOLESCENT

- Pregnancy: Diagnosis and Counseling
 1. Risk factors
 - a. Early onset of sexual activity; inadequate concept of fertility and contraception
 - b. Low socioeconomic level
 - c. Poor academic achievement
 - d. Low self-image; few options for future
 - e. Early pregnancy in mother or sister
 - f. Substance abuse
 - g. Physical/sexual abuse
 - h. Barriers to contraceptive use include misinformation, lack of health care, poor communication with partner
 - i. Exposure to irresponsible media portrayal of sex
 2. Incidence
 - a. U.S. has the highest rates of adolescent pregnancy and births among industrialized nations; estimates of 800,000 each year
 - b. Rates have shown a decline from 1991 to 2002 of 28% among 15- to 19-year-olds

- with largest decline among African-American teens
 - c. Birth rate among 15- to 19-year-old females—42.9:1000 (2002)
 - d. Subsequent pregnancy common—12% to 44% pregnant again within 1 year, 20% to 37% within 2 years
 - e. Teen mothers are more likely to drop out of school than teens without babies
3. Signs and Symptoms
 - a. First trimester
 - (1) Irregular menses/amenorrhea
 - (2) GI—nausea, vomiting
 - (3) Urinary frequency
 - (4) Breast tenderness/tingling
 - (5) Other—headache, vertigo, abdominal cramps
 - b. Second trimester
 - (1) Increased/darkening skin pigmentation
 - (2) Fetal movement—“quickening” 16 to 20 weeks
 - (3) Contractions—Braxton-Hicks 16 to 27 weeks
 - c. Third trimester—increased contractions
 4. Physical Findings
 - a. First trimester
 - (1) Breast—fullness/tenderness, nipple tingling/discharge/darkening areola
 - (2) Abdomen—uterine fundus at symphysis pubis at 12 weeks
 - (3) Pelvic examination
 - (a) Softening of uterine isthmus (Hegar sign) 6 to 8 weeks
 - (b) Bluish hue to cervix/vaginal epithelium (Chadwick sign) 6 to 8 weeks
 - (c) Cervical softening (Goodell sign) 6 to 8 weeks
 - (d) Increased leukorrhea
 - (4) Weight gain—2 pounds (1 kg)
 - (5) Fetal heart tones (FHT)—doppler 10 to 12 weeks
 - b. Second trimester
 - (1) Stretch marks “striae” on abdomen and breasts
 - (2) Fundus midway between symphysis pubis and umbilicus by 14 to 15 weeks, at umbilicus 20 to 22 weeks
 - (3) Fetal heart tones (FHT) by fetoscope at 20 weeks
 - (4) Weight gain—11 pounds (5 kg)
 - c. Third trimester
 - (1) Colostrum from breasts 28 to 40 weeks
 - (2) Fundus between umbilicus and xiphoid, 28 weeks; at xiphoid, 38 weeks

- (3) Weight gain—11 pounds (5 kg)
- (4) Bloody show—impending labor
- (5) Ruptured membranes
- d. Labor
 - (1) Stage 1—effacement and dilatation of cervix
 - (2) Stage 2—delivery of fetus
 - (3) Stage 3—separation and delivery of placenta
- 5. Differential Diagnosis
 - a. Ectopic pregnancy
 - b. Incomplete spontaneous abortion
 - c. Molar pregnancy
 - d. UTI
- 6. Diagnostic Tests/Findings
 - a. Urine for human chorionic gonadotropin (hCG)—positive 7 to 10 days after conception
 - b. Radioimmunoassay (RIA)—serology, more specific than urine, expensive, positive 7 to 10 days after conception
 - c. Cervical cultures—gonorrhea/chlamydia screen
 - d. Wet mount—saline, KOH
 - e. Papanicolaou smears
 - f. Serology—syphilis, hepatitis B surface antigen, blood type/Rh factor, CBC with indices, rubella, human immunodeficiency virus
 - g. Urinalysis/culture (if indicated)
 - h. Pelvic ultrasound
- 7. Counseling/Education
 - a. Impact on future plans, finances, family structure
 - b. Identity and age of father—anticipated involvement
 - c. Options
 - (1) Continue pregnancy and maintain custody
 - (2) Place child for adoption
 - (3) Termination of pregnancy
 - d. Prenatal care—examinations, adequate diet, vitamins (include folic acid), iron, and calcium
 - e. Avoidance of medications/drugs/alcohol/x-rays
- Prenatal Diagnosis: Identify potential inherited/acquired defects
 - 1. Variety of causes—genetic factors (25%); environmental (15%); combination of both genetic and environmental (30%); unaccountable (30%)
 - 2. Incidence—3% to 5% infants in U.S.
 - 3. Risk factors—maternal
 - a. Disease—diabetes, thyroid, immune deficiency or compromise
 - b. Age—young, especially less than 16 years; over 35 years
 - c. Previous child with Down syndrome, anencephaly, meningomyelocele
- 4. Screening tools
 - a. Family pedigree—graphic record, family medical history
 - b. Alpha-fetoprotein
 - (1) Screens for neural tube defects; Meckel syndrome
 - (2) Serum levels less accurate than amniotic fluid analysis but can be done earlier in pregnancy for initial screen
 - c. Amniocentesis—collection of amniotic fluid, 15 to 16 weeks gestation; karyotype/chromosomal analysis; inborn errors of metabolism; confirmatory test with abnormal serum alpha-fetoprotein
 - d. Chorionic villus sampling—tissue (villus) sample from fetal placenta at 9 to 11 weeks gestation; chromosomal abnormality; usually reserved for women over 35 years
 - e. Metabolic disease, hemaglobinopathies from DNA analysis
- Genetic Counseling: Communication process regarding risk/problems surrounding certain disorders
 - 1. Often initiated following birth of affected child
 - 2. Both parents included—family history, medical/psychological consequences for child/family, possibility of future children affected, options
 - 3. Utilizes results from prenatal diagnostic tools/tests
- Pregnancy Termination
 - 1. Spontaneous termination—miscarriage
 - a. Miscarriage/spontaneous abortion of pregnancy prior to fetal viability
 - b. May be complete or incomplete
 - c. Occurs in 20% of all pregnancies, including adolescents
 - d. Often associated with genetic abnormality in fetus
 - 2. Elective termination—induced abortion
 - a. Induced or elective abortion—35% of adolescent pregnancies
 - b. Procedural options—dependent on trimester of termination
 - c. First trimester abortion options—rule out STI prior to procedure
 - (1) Manual syringe evacuation/early suction curettage—4 to 6 weeks of pregnancy

- (a) Cannula positioned into uterus with aspiration or suction of conceptus (menstrual extraction)
 - (b) Low risk of genital injury or complications
 - (2) Suction curettage/vacuum aspiration—up to 12 to 14 weeks of pregnancy
 - (a) Cervical dilation 6+ hours prior to procedure using Laminaria (hydrophilic seaweed sticks)
 - (b) Cannula position into uterus with suction followed by curettage
 - (c) Considered safest in first trimester
 - (3) Medical abortion—induces abortion when administered in early pregnancy (49 days or less)
 - (a) Acts as antiprogesterone
 - (b) Initial dose of Mifepristone 600 mg followed with 400 mcg of misoprostol (a prostaglandin) two days later
 - (c) Risks include pain, excessive bleeding
 - d. Second trimester
 - (1) Dilatation and evacuation curettage (D&E)—20 to 24 weeks of pregnancy
 - (a) Cervical dilation prior to procedure with osmotic dilators
 - (b) Conceptus removed via curettage, aspiration, or ring forceps under general anesthesia; risk of cervical trauma
 - (c) Antibiotics recommended postprocedure
 - (2) Prostaglandin suppository technique—16 to 24 weeks of pregnancy; rarely used
 - (a) Cervical dilation with osmotic dilators followed by vaginal suppositories of prostaglandin E; has largely replaced more controversial intraamniotic instillation techniques
 - (b) 20 mg prostaglandin suppositories used every 3 to 4 hours, induces labor and subsequent abortion within 4 to 60 hours
 - (c) Complications include significant flu-like symptoms and possible delivery of live fetus
 - e. Referral and follow-up
 - (1) Referral to obstetrician/gynecologist or family planning clinic once decision to terminate pregnancy is made
 - (2) Continue supportive care and counseling options on follow-up
- Birthing Methods: May have greater risk of cesarean delivery due to immature pelvic skeletal development; cephalopelvic disproportion
 - Prematurity/Low Birth Weight
 - 1. Maternal risk factors—maternal age less than 16; poor prenatal care/poor nutrition
 - 2. Prevention aimed at prenatal management
 - a. Good nutrition; avoid dieting—daily calories 2500 to 2700 kcal/day
 - b. Supplements—vitamins (include folic acid), iron (anemia possible), calcium
 - 3. Early/consistent prenatal care
 - a. Avoid all medications unless specifically approved by healthcare provider
 - b. Potential for multisystem complications—infant
 - (1) Respiratory—respiratory distress syndrome, bronchopulmonary dysplasia, apnea
 - (2) Cardiovascular—patent ductus arteriosus, bradycardia, malformations
 - (3) Hematologic—hyperbilirubinemia, subcutaneous hemorrhage
 - (4) Gastrointestinal—poor motility
 - (5) Metabolic/endocrine—hypocalcemia, hypoglycemia or hyperglycemia, hypothermia
 - (6) Central nervous system—intraventricular hemorrhage, hypotonia
 - (7) Renal abnormalities
 - (8) Infections
 - Home Monitoring/follow-up care
 - 1. Frequent contact with mother/infant—early discharge follow-up recommended within 48 hours
 - a. Infant feeding patterns
 - b. Adaptation of mother to sleep changes
 - c. Social support—father of baby, mother's family
 - 2. Follow-up (well-child visits)
 - a. Adolescent problems—repeated pregnancy, STI
 - b. Optimal health of mother/infant
 - c. Future plans—completion of high school, college/technical training, career
 - Maternal Substance Abuse
 - 1. Risk of substance abuse among adolescents—requires assessment
 - 2. Drug/alcohol use
 - a. Counsel at initial prenatal visit
 - b. Reinforce impact of all substances on developing fetus—tobacco, alcohol, marijuana, inhalants, cocaine, heroin

- Screening for potentially abusive partner
 1. Increased risk of battering by partner when pregnant
 - a. Assess relationship between girl and partner
 - b. Some states require mandatory reporting of victims of domestic violence
 2. Risk factors for child abuse
 - a. Parental factors
 - (1) Young and/or immature adolescent parents
 - (2) Minimal education
 - (3) Financial stress
 - (4) Lack of social support
 - (5) Unplanned or unwanted pregnancy
 - (6) Unrealistic expectations of parenting
 - b. Infant factors
 - (1) Prematurity
 - (2) Perception as “different/bad;” difficult temperament
 - (3) Congenital defect/malformation
- Perinatal complications of neonate
 1. Low birth weight and prematurity—see earlier section
 2. Infant mortality, linked with low birth weight and prematurity
 3. Cognitive/social development—possible low self-esteem, less responsive/expressive, decreased ability to trust others
 4. Sudden Infant Death Syndrome (SIDS)

■ QUESTIONS

Select the best answer

1. Urinary tract infections (UTI) are the most common pediatric urinary tract problems seen in primary care. Which of the following statements is not true regarding UTIs?
 - a. Symptoms are often nonspecific, especially in infancy
 - b. Urine culture is required for definitive diagnosis
 - c. Trimethoprim-sulfamethoxazole is drug of choice for most children
 - d. Radiologic studies are rarely indicated with first infection
2. A 2-year-old girl presents with symptoms of painful urination, frequency, and occasional incontinence over the past week. When seen in your office, she has a temperature of 101.6°F. Which of the following would be your approach in establishing a definitive diagnosis?
 - a. Clean-catch midstream collection of specimen for urine analysis
 - b. Clean-catch midstream collection of specimen for urine culture
 - c. Straight catheterization collection of specimen for urine culture
 - d. Voiding cystourethrogram (VCUG)
3. The most likely organism to cause a UTI in the pediatric population is:
 - a. *Staphylococcus saprophyticus*
 - b. *Klebsiella*
 - c. *Chlamydia*
 - d. *E. coli*
4. One of the most commonly suggested reasons for primary enuresis is:
 - a. Certain medications, such as theophylline
 - b. Genitourinary abnormalities
 - c. Family disruptions and stress
 - d. Delayed maturation of voiding inhibitory reflex
5. Suzanne, a 7-year-old, comes to you for a physical examination prior to participation in soccer. Suzanne’s mother is concerned that the child “still has accidents at night.” You determine that Suzanne has primary nocturnal enuresis and your first recommendation to her mother is to:
 - a. Avoid use of criticism or punishment
 - b. Use a sticker/star chart
 - c. Treat with medication
 - d. Purchase an enuresis alarm
6. The incidence of cryptorchidism at one year of age is about 1%. The best explanation for this is:
 - a. Examination of the scrotum begins at this age
 - b. A child can usually stand, making palpation of the testes easier
 - c. Spontaneous resolution often occurs in first year
 - d. Surgical repair can now be done in neonatal period
7. Communicating hydrocele is best differentiated from the noncommunicating type by the fact that:
 - a. There is no association with hernia
 - b. It usually resolves on its own
 - c. The fluid is static in the scrotum
 - d. Frequently develops into hernia
8. In counseling parents when their child is diagnosed with mild hypospadias, suggest that the following may be part of the management:
 - a. Circumcision
 - b. Radiography
 - c. Consult with pediatric urology
 - d. Surgical correction at 2 years of age

9. On physical examination of a 2-year-old uncircumcised male, you note that the foreskin is retracted and discolored. There is swelling of the glans. The most likely diagnosis is:
 - a. Phimosis
 - b. Balanitis
 - c. UTI
 - d. Paraphimosis
10. Meatal stenosis, narrowing of the distal urethra, is seen following:
 - a. Orchiopexy
 - b. Circumcision
 - c. Epididymitis
 - d. Hypospadias repair
11. During a track meet, a 14-year-old male pole-vaulter falls to the ground screaming in pain. He complains of intense, searing pain in his right scrotum. He vomits twice while waiting for the ambulance. He most likely has:
 - a. Orchitis
 - b. Hydrocele
 - c. Acute epididymitis
 - d. Testicular torsion
12. Treatment for this disorder is primarily:
 - a. Scrotal elevation
 - b. Ice
 - c. Immediate surgical referral
 - d. Bedrest
13. Labial adhesions are a relatively common finding among infants and young girls. Which of the following statements about this condition is true?
 - a. Adhesions are usually present at birth but may be missed on examination
 - b. Highest incidence is from birth to 3 years
 - c. Simple lysis of adhesions is often recommended
 - d. Most cases resolve without intervention
14. Which of the following statements is true regarding the use of topical application of conjugated estrogen cream with labial adhesions?
 - a. It is highly successful in resolving most adhesions within 2 months
 - b. It is no longer recommended because it may stimulate precocious puberty
 - c. Topical applications of bland creams or petroleum jelly are equally effective
 - d. Mechanical lysis is preferred treatment today
15. Sheryl, a 12-year-old, complains of a vaginal discharge for the past 8 to 9 months. She tells you her underpants are frequently wet. When she wipes after urinating, there is "white stuff" on the tissue. Sheryl denies urinary problems, genital itching, or odor. She also denies sexual activity. Her menses have not yet started, but she reports she "started to develop" in her breasts at about age 10. Her vaginal discharge is most likely a result of:
 - a. A fungal infection
 - b. Poor hygiene
 - c. Retained foreign body
 - d. Physiologic leukorrhea
16. Your recommendations to Sheryl regarding management of the discharge includes which one of these?
 - a. Vinegar and water douche
 - b. Placing a sanitary "mini pad" in her underpants
 - c. A 10-day course of penicillin or erythromycin
 - d. Use of a monilial cream for 1 week
17. Which of the following is not true of dysmenorrhea?
 - a. Onset is usually within the first two to three months following menarche
 - b. A leading cause of school absenteeism in adolescent females
 - c. Systemic symptoms include vomiting and dizziness
 - d. Pain is from start of menses to about 24 to 48 hours later
18. Amy, a 16-year-old, has symptoms of premenstrual syndrome. She refuses to take NSAIDs, preferring instead more "natural" treatments. Her options include:
 - a. Eating foods rich in sodium and fat
 - b. Including more foods or supplements with vitamin C
 - c. Limited fluid intake to avoid "bloating"
 - d. Adequate rest, a healthy diet, and exercise
19. In cases of accidental genitourinary trauma, which of the following is not commonly seen?
 - a. Extensive tears of the vaginal wall
 - b. Hematuria
 - c. Hematoma of the urethra, scrotum, lower abdomen
 - d. Periurethral lacerations
20. The most common form of glomerulonephritis in children is:
 - a. Mesangial proliferative
 - b. Poststreptococcal
 - c. Membranoproliferative
 - d. Mesangio-streptococcal

21. Which of the following signs/symptoms is not associated with acute forms of glomerulonephritis?
 - a. Edema
 - b. Hematuria
 - c. Increased urine output
 - d. Dark urine
22. One of your male patients presents with weight loss, abdominal pain, and decreased urine output. On examination, you palpate a right-sided mass, noting tenderness in the abdomen and flank. Urinalysis reveals significant leukocytosis. An intravenous pyelogram is ordered, which shows marked delay of emptying from the renal pelvis. The most likely diagnosis is:
 - a. Glomerulonephritis
 - b. Pyelonephritis
 - c. Hydronephrosis
 - d. UTI
23. Which of the following is true of renal tubular acidosis type 1?
 - a. Genetically transmitted as autosomal recessive disorder
 - b. Distal tube defect affecting secretion of hydrogen ions
 - c. Distal tube defect affecting bicarbonate reabsorption
 - d. Most children remain short in stature in spite of early treatment
24. Vulvovaginitis may be caused by all of the following except:
 - a. Poor hygiene
 - b. Herpes simplex virus
 - c. Pinworms
 - d. Condylomata acuminata
25. Meatal stenosis can be identified by all of these with the exception of:
 - a. Crying with urination
 - b. Inflammation of glans penis
 - c. Slit-like meatus
 - d. Wide urinary stream
26. Trina, age 14, comes in with complaints of headache and nausea. She admits to having a boyfriend, whom she has sex with "once in a while," and sometimes he uses a condom. She had a period last month, but it lasted 2 days instead of the usual seven. Which of the following are not usually perceived as risk factors for adolescent pregnancy?
 - a. Early onset of sexual activity
 - b. Familiarity with fertility knowledge
 - c. Sporadic, if any, contraceptive use
 - d. Low self-image
27. Trina's urine pregnancy test is positive and confirmed by serology. Which of the following physical findings is not consistent with a gestation of less than 12 weeks?
 - a. Hegar sign
 - b. Goodell sign
 - c. Doppler auscultation of fetal heart sounds
 - d. 4 to 6 pound weight gain
28. Congenital defects are present in approximately 3% to 5% of infants born in this country. Which of the following factors is the lowest contributor to defects?
 - a. Genetic
 - b. Unknown
 - c. Environmental
 - d. Combination of environmental and genetic
29. Which of the following is the earliest screening test that you would use in managing Trina's care during her first trimester of pregnancy?
 - a. Amniocentesis
 - b. Chorionic villus sampling
 - c. Serum alpha-fetoprotein
 - d. Fetal radiograph
30. Trina expresses to you that she may want to terminate the pregnancy. She has heard that abortion is a simple procedure, without complications. Your discussion with her is based on the fact that:
 - a. Adolescents only choose to terminate pregnancy 10% of the time
 - b. Menstrual extraction could be done at this point
 - c. The risk is high for hemorrhage and fever
 - d. Cervical injury overall from induced abortion is rare
31. You know that if Trina delivers her baby, close follow-up of both is important. What is not a focus of care in the first few weeks after delivery?
 - a. Social support for the family
 - b. Mother's goals for education
 - c. Infant's continued weight gain
 - d. Adaptation of mother to sleep changes
32. While child abuse occurs across age barriers, the adolescent parent may be at greater risk for abusing their child because:
 - a. The pregnancy may be unplanned or unwanted
 - b. Adolescents continue substance abuse after the baby is born
 - c. The infant mortality rate is low
 - d. Education becomes more important than parenting

33. A thick, purulent vaginal discharge that is greenish-yellow is most likely an infection caused by:
- Chlamydia trachomatis
 - Herpes simplex virus
 - Neisseria gonorrhoea
 - Human papillomavirus
34. The primary treatment for this infection would be:
- Amoxicillin
 - Ceftriaxone
 - Ofloxacin
 - Penicillin
35. The most common sexually transmitted disease in the United States is:
- Gonorrhoea
 - Human immunodeficiency virus (HIV)
 - Chlamydia
 - Herpes
36. Which of the following tests provide the most definitive diagnosis for suspected syphilis?
- VDRL
 - ART
 - Dark-field microscopy
 - FTA-AB_s
37. Which of the following is not true regarding the use of acyclovir in the treatment of herpes simplex virus?
- Topical treatment not recommended
 - Treatment is equally effective for active primary and recurrent lesions
 - The focus of treatment is decrease in intensity symptom duration, and viral shedding
 - Therapy is best initiated within 6 days of onset of lesions
38. Your 15-year-old female patient presents with genital lesions. Your workup is based on the knowledge that:
- Human papillomavirus (HPV) is often associated with malignancy
 - HPV manifests itself as molluscum contagiosum
 - Most HPV infections cause genital itching and pain
 - Syphilis serology should be included to distinguish HPV from condylomata lata
39. After your examination, you determine that your patient has condylomata acuminata. Which of the following is not commonly used to treat these lesions?
- Topical podophyllum resin
 - Laser treatment for unresponsive lesions
 - Topical acyclovir
 - Treatment is often not needed; many lesions regress spontaneously
40. During your evaluation of Lisa, a 16-year-old female patient, she relates recent, first-time sexual intercourse with her boyfriend. Now she complains of a "frothy" substance in her underwear that "smells weird." She also has slight itching in her vaginal area. You suspect:
- Trichomoniasis
 - Gonorrhoea
 - Chlamydia
 - Herpes
41. What education needs to be included in prescribing metronidazole to treat trichomoniasis?
- It is safe for use during pregnancy
 - Sexual contact may resume after 48 hours of treatment
 - Sexual contacts do not require therapy
 - Alcohol should not be used during treatment
42. A patient with a thin, white, malodorous vaginal discharge most likely has:
- Trichomoniasis
 - Monilia
 - Bacterial vaginosis
 - Chlamydia
43. At which point of life is UTI more common in males?
- Toddler
 - Adolescent
 - School-age
 - Newborn/infant
44. The organism primarily responsible for UTI is:
- Proteus
 - E. coli
 - Enterobacter
 - Pseudomonas
45. Most patients with uncomplicated UTI can be treated on an outpatient basis. The first antibiotic you would consider using is:
- Amoxicillin
 - Trimethoprim-sulfamethoxazole
 - Cephalexin
 - Amoxicillin/clavulanate
46. Cryptorchidism is more prevalent in:
- Term infants
 - Premature infants
 - Babies at 1 year of age
 - Toddlers

47. A 3-year-old boy is in the process of toilet training. The parents come to see you because when the child urinates, all of the urine goes on the floor. You suspect hypospadias because:
- It is a rare disorder and would indicate a more serious problem
 - Normally, the urine stream is directed from the meatal opening at the tip of the penis, in a straight path
 - The child is circumcised
 - Fusion of the urethral folds has occurred
48. Which of the following is not used in the management of the child with phimosis?
- Ice packs
 - Gentle stretching when bathing
 - Circumcision in cases of urinary obstruction
 - Good hygiene
49. Jeanine, an 8-year-old girl, presents with a 2-week history of a brownish-red, very foul smelling, vaginal discharge. It is most likely caused by:
- Sexual abuse
 - Accidental genital trauma
 - Foreign body
 - Poor hygiene
50. Which of the following is not a proposed etiologic factor in premenstrual syndrome?
- Imbalance of water and sodium
 - Fluctuation in steroids
 - Vitamin B₁₂ deficiency
 - Synthesis of prostaglandins
51. Treatment of a child with glomerulonephritis would include:
- Vasoconstrictors for hypotension
 - Antibiotics for persistent infection
 - Increased fluids to maintain hydration
 - Avoidance of diuretics
52. Which of the following is not a frequent sign of gonorrhea infection:
- Diarrhea
 - Urinary symptoms
 - Menstrual problems
 - Fever
53. The most significant finding in the case of chlamydia infection is:
- The vaginal discharge is gray and frothy
 - Most examinations are normal
 - The external genitalia is markedly inflamed
 - Infection by *Chlamydia trachomatis* alone is the rule
54. One difference between the lesions of primary syphilis (chancre) and HSV genital lesions that may help in differentiating the two is:
- Syphilis chancre is painless while HSV lesions are painful
 - Syphilis lesions have a flat edge while HSV lesion edges are more raised
 - Syphilis chancres are more likely to ulcerate than HSV lesions
 - Erythema and edema is greater with syphilis chancre than HSV lesions
55. Education of sexually active adolescents concerning use of condoms to prevent STDs would include:
- Only latex or natural skin condoms should be used
 - Polyurethane condoms have inadequate STD protection
 - Condom use is only method to prevent STDs and AIDS
 - Only petroleum-based lubricants should be used
56. Which of the following contraceptive methods would be least suitable for adolescents?
- Condoms with spermicides
 - Progestin-only mini-pill
 - Long-acting progestins
 - Estrogen-progestin combination
57. Which of the following is not an absolute contraindication for oral contraceptives?
- Clotting disorder
 - Impaired liver function
 - Severe hypertension
 - Undiagnosed vaginal bleeding

ANSWERS

- | | |
|-------|-------|
| 1. d | 30. b |
| 2. c | 31. b |
| 3. d | 32. a |
| 4. d | 33. c |
| 5. a | 34. b |
| 6. c | 35. c |
| 7. d | 36. c |
| 8. c | 37. b |
| 9. d | 38. d |
| 10. b | 39. c |
| 11. d | 40. a |
| 12. c | 41. d |
| 13. d | 42. c |
| 14. a | 43. d |
| 15. d | 44. b |
| 16. b | 45. b |
| 17. a | 46. b |
| 18. d | 47. b |
| 19. a | 48. a |
| 20. b | 49. c |
| 21. c | 50. c |
| 22. c | 51. b |
| 23. b | 52. a |
| 24. d | 53. b |
| 25. d | 54. a |
| 26. b | 55. c |
| 27. d | 56. b |
| 28. b | 57. c |
| 29. c | |

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Hematologic/Oncologic/ Immunologic Disorders

M. Elizabeth Younger

Kathy Ruble

■ ABO INCOMPATIBILITY

- Definition: Incompatibility between the ABO blood group of the fetus and mother (mother is blood type O and baby is A, B, or AB)
- Etiology/Incidence
 1. Occurs in approximately 20% of pregnancies
 2. Approximately 33% of the infants in these pregnancies are Coombs test positive
 3. Approximately 20% of these infants develop significant hyperbilirubinemia and may also develop anemia in the first several weeks of life
- Signs and Symptoms
 1. Mild cases—asymptomatic
 2. Severe cases—yellow discoloration of skin, sclerae, and gums or inside of mouth
- Differential Diagnosis
 1. Physiologic jaundice—most common
 2. Infection
 3. Hyperbilirubinemia of prematurity
 4. Metabolic disorder
- Physical Findings
 1. Jaundice usually occurring within first 24 hours of life; may be present or appear up to a week later
 2. May have hepatosplenomegaly
- Diagnostic Tests/Findings
 1. Blood type—mother usually O; baby A, B, or AB

2. Coombs test (DAT)—positive in approximately $\frac{1}{3}$ of cases
3. Hemoglobin—moderately low; hemolysis occasionally occurs up to 2 to 3 months
4. Serum bilirubin—variably elevated

• Management/Treatment

1. Monitor unconjugated bilirubin levels
2. Phototherapy if indicated based on bilirubin, gestational, and post-delivery age of neonate
3. If anemia is severe, may require transfusion

■ RH INCOMPATIBILITY

- Definition: Incompatibility between the Rh blood group of the fetus and mother (Mother is Rh negative and baby is Rh positive)
- Etiology/Incidence: Relatively uncommon, 5% occurrence in first susceptible newborn, becomes more severe with each pregnancy if untreated
- Signs and Symptoms
 1. Causes hemolysis, resulting in anemia and hyperbilirubinemia
 2. In severe cases causes fetal death secondary to erythroblastosis fetalis
- Differential Diagnosis
 1. Physiologic jaundice—most common
 2. Anemia secondary to another etiology
 3. Infection
 4. Hyperbilirubinemia of prematurity
 5. Metabolic disorder

- Physical Findings
 1. Jaundice
- Diagnostic Tests/Findings
 1. Blood type—mother Rh negative, baby Rh positive
 2. Direct Coombs test—positive
 3. Hemoglobin—below normal, may be very low; hemolysis often continues up to 3 months
 4. Serum indirect bilirubin—markedly elevated
- Management/Treatment
 1. Mother
 - a. Rh isoimmunization screen at first prenatal visit
 - b. If mother Rh negative, test father; if father is Rh positive then the pregnancy is at risk
 - c. Risk for problems increases with each pregnancy as antibody levels rise
 - d. Administration of Rh immune globulin after any invasive procedure during pregnancy and after the termination of each pregnancy (including any miscarriage and/or abortion)
 2. Infant
 - a. Antenatal treatment—once diagnosis has been established, transfusion of fetus with Rh negative blood
 - b. Postpartum treatment
 - (1) Phototherapy, with exchange transfusion if indicated by bilirubin level
 - (2) Transfusion of packed red blood cells if indicated by hemoglobin level
 - (3) Multiple studies have reported on efficacy of administration of gamma globulin, but there is no recommendation for this treatment at this time

- (5) Higher incidence among preterm infants and LBW infants
 - (6) Occurs in approximately 13% of breastfed infants within the 1st week of life (breastfeeding jaundice)
- b. Breastmilk jaundice
 - (1) Occurs in approximately 2% of breastfed infants
 - (2) Develops after the 7th day of life with maximum bilirubin concentrations of 10 to 30 mg/dL reached during the 2nd to 3rd week of life
 - (3) Levels fall rapidly with discontinuation of breast feeding for 1 to 2 days, after which breast feeding can resume
 - c. Prolonged (nonphysiologic) neonatal hyperbilirubinemia—associated with breastfeeding, maternal diabetes, induced labor, prematurity, Asian ethnicity, and male gender
 - (1) Common in infancy
 - (2) Less common in older children but can be associated with hemoglobinopathies and some metabolic syndromes
2. Conjugated (direct) types—caused by range of pathologic conditions (rare in newborns) including biliary obstruction, infection, drugs (aspirin, acetaminophen), and other metabolic disorders
 - a. Direct bilirubin > 1.5 to 2 mg/dL
 - b. Jaundice in first day of life
 - c. Total bilirubin > 12.9 mg/dL (full-term); > 15 mg/dL (preterm)
 - d. Persistence > 1 week (full term); > 2 weeks (preterm)

- Signs and Symptoms: Yellow discoloration of skin, whites of eyes, gums, and oral mucosa

- Differential Diagnosis
 1. Transient neonatal hyperbilirubinemia
 - a. Physiologic
 - b. Breastfeeding associated
 - c. Breastmilk jaundice
 2. Infection
 3. Hepatic disease
 4. Intestinal obstruction

- Physical Findings
 1. Clinical jaundice varies based on bilirubin level—5 mg/dL appears first on head, progresses down chest/abdomen as bilirubin increases; usually at least 15 mg/dL when noted on distal extremities
 2. Hepatosplenomegaly
 3. Edema

■ NEONATAL HYPERBILIRUBINEMIA

- Definition: An increased amount bilirubin in the blood; if untreated and bilirubin levels continue to rise can cause encephalopathy (kernicterus)
- Etiology/Incidence
 1. Unconjugated (indirect) types—may be caused by overproduction of bilirubin, impaired conjugation, transport or uptake of bilirubin
 - a. Physiological hyperbilirubinemia
 - (1) Indirect bilirubin 5 to 7 mg/dL
 - (2) Usually peaks within 3 to 5 days of life
 - (3) Usually resolves within 10 days
 - (4) Occurs in approximately 50% of full-term infants

- Diagnostic Tests/Findings
 1. Evaluate for pathologic causes for jaundice—sepsis, polycythemia
 2. Coombs test (DAT)
 - a. Blood group incompatibilities—DAT usually positive
 - b. Membrane disorders, red cell enzyme disorders, bacterial or viral sepsis, drug toxin—DAT usually negative
 3. Bilirubin
 - a. Indirect hyperbilirubinemia—unconjugated bilirubin increased
 - b. Direct hyperbilirubinemia—conjugated bilirubin increased
 - (1) Reticulocyte count—may be increased with both indirect or direct
- Management/Treatment: Based on etiology, age of child, and bilirubin level
 1. Indirect (unconjugated) hyperbilirubinemia
 - a. Hydration and feeding
 - b. Phototherapy
 - c. Exchange transfusion
 2. Direct (conjugated) hyperbilirubinemia—treat underlying disease and/or refer as appropriate

■ HEMOGLOBINOPATHIES

- Definition: Production of abnormal hemoglobin due to inherited genetic mutation in globin genes
- Etiology/Incidence: Incidence associated with specific ethnic groups
 1. Beta (β) thalassemia
 - a. β -chain synthesis decreased in β -thalassemia intermedia; absent in β -thalassemia major (Cooley's anemia)
 - b. Increased but ineffective erythropoiesis
 - c. Shortened red cell life span
 2. Alpha (α) thalassemia
 - a. β -chain production is impaired
 - b. Increased but ineffective erythropoiesis
 - c. Shortened red cell life span
 3. Hemoglobin C and E
 - a. Amino acid substitution of lysine for glutamic acid
 - b. Hemoglobin C—carrier state in 2% of African-Americans
 - c. Hemoglobin E—prevalent in populations from Southeast Asia
 4. Ethnic groups—individuals of African, Asian, and Mediterranean descent are at highest risk
- Signs and Symptoms
 1. β -thalassemia major and intermedia
 - a. Pale skin or mucous membranes
 - b. Weakness
 - c. Characteristic facies
 2. β -thalassemia minor (trait), α -thalassemia trait, hemoglobin C, hemoglobin E—usually asymptomatic
- Differential Diagnosis
 1. Iron deficiency anemia
 2. Lead poisoning
 3. Chronic infection
 4. Sideroblastic anemia
 5. Malignancy
- Physical Findings
 1. β -thalassemia major and intermedia
 - a. Splenomegaly, occasional hepatosplenomegaly
 - b. Jaundice, usually mild
 - c. Abnormal facies secondary to extramedullary hematopoiesis—prominence of malar eminences, frontal bossing, depression of bridge of nose, exposure of upper incisors
 - d. Growth retardation
 2. β -thalassemia minor, α -thalassemia trait, and hemoglobin C trait—physical examination normal
 3. Hemoglobin C and E—splenomegaly
- Diagnostic Tests/Findings
 1. β -thalassemia
 - a. CBC with red cell indices
 - (1) Hemoglobin—decreased
 - (2) Hypochromia, microcytosis, low MCV, anisocytosis, target cells
 - (3) Reticulocyte count—increased
 - b. Hemoglobin electrophoresis
 - (1) β^+ thalassemia
 - (a) Hgb A—present
 - (b) Hgb A₂—increased (usually > 3.6%, depending on laboratory)
 - (c) Hgb F—normal or slightly increased
 - (2) β^0 thalassemia
 - (a) Hgb A—absent
 - (b) Hgb A₂—increased (usually > 3.6%, depending on laboratory)
 - (c) Hgb F—normal or slightly increased
 2. β -thalassemia (minor or trait)—usually discovered on routine examination or in family investigation
 - a. CBC with red cell indices
 - (1) Hemoglobin—slightly decreased (9 to 11 g/dL)
 - (2) Hypochromic, microcytic cells, target cells, anisocytosis, basophilic stippling, low MCV, low MCH, normal RDW

- b. Hemoglobin electrophoresis
 - (1) Hgb A₂—increased (usually > 3.6%, depending on laboratory)
 - (2) Hgb F—may be mildly elevated
- 3. α -thalassemia trait
 - a. CBC with red cell indices
 - (1) Hemoglobin—may be slightly decreased
 - (2) Microcytosis, hypochromia
 - b. Hemoglobin electrophoresis—Hgb Barts on newborn screen; disappears by 1 to 3 months of age (2 gene deletion approximately 2%, 3 gene deletion ~2% to 6%)
- 4. Hemoglobin C and Hemoglobin E
 - a. CBC with red cell indices
 - (1) Hemoglobin—may be decreased
 - (2) Hemoglobin C—target cells, spherocytes, increased reticulocytes
 - (3) Hemoglobin E—target cells, microcytosis
 - b. Hemoglobin electrophoresis
 - (1) Hemoglobin C (Hgb C)—present
 - (2) Hemoglobin E (Hgb E)—present
- Management/Treatment
 1. β -thalassemia major
 - a. Refer to hematologist
 - b. Chronic transfusion protocol—to maintain hemoglobin, support growth, and prevent extramedullary hematopoiesis
 - c. Chelation therapy (desferoxamine) after serum ferritin \geq 2000 ng/mL; to remove excessive intracellular iron
 - d. Prophylactic penicillin initiated by 2 months of age, until at least 5 years of age; recommendations for prophylaxis after the age of 5 are equivocal
 - e. Splenectomy—may reduce transfusion requirements
 - (1) Postsplenectomy care: All immunizations including pneumococcal, meningococcal, and annual influenza immunizations as recommended by the CDC; prophylactic penicillin or amoxicillin daily; because of the increased risk for infection with an encapsulated bacteria, any fever greater than 101.5°F (38.5°C) requires immediate evaluation with a physical exam, CBC, and blood culture; and if child looks septic, a dose of ceftriaxone
 2. β -thalassemia intermedia
 - a. Observe hemoglobin
 - b. Splenectomy may help
 3. β -thalassemia minor, α -thalassemia trait, hemoglobin C and E—no therapy
 4. Genetic counseling

■ IRON DEFICIENCY ANEMIA

- Definition: A microcytic, hypochromic anemia caused by inadequate supply of iron, associated with low reticulocyte count and elevated red cell distribution width (RDW)
- Etiology/Incidence
 1. Causative factors
 - a. Deficient dietary intake
 - b. Increased demand—growth (low birth weight, prematurity, adolescence, pregnancy), cyanotic congenital heart disease
 - c. Blood loss—GI tract is most common site
 - d. Malabsorption—rare except for after a bowel resection
 2. Incidence
 - a. Most common nutritional deficiency in children
 - b. Most common between 8 and 18 months of age and in adolescence, not during early infancy as infants are born with sufficient iron stores to prevent anemia for first 4 to 5 months of life
 - c. Higher incidence in lower socioeconomic groups
- Signs and Symptoms: Vary with severity of anemia
 1. Mild anemia—usually asymptomatic
 2. More severe anemia
 - a. Fatigue
 - b. Irritability
 - c. Delayed motor development
 - d. Eating nonnutrient substances such as ice, plaster, clay, paint, fabric (pica)
- Differential Diagnosis
 1. Hemoglobinopathy
 2. Lead poisoning
 3. Anemia of chronic disease or inflammation
- Physical Findings: Vary with severity of anemia
 1. Mild—normal physical examination
 2. More severe—pallor, tachycardia, systolic murmur, hepatomegaly, CHF
- Diagnostic Tests/Findings—see **Table 13-1**
 1. Hemoglobin— < 3rd percentile for age
 2. Peripheral blood smear—hypochromic, microcytic red cells, confirmed by RBC indices MCV < 3rd percentile for age
 3. Wide red cell distribution width (RDW) > 17
 4. Serum ferritin—decreased (this is the first value to fall)

■ **Table 13-1** Differential Diagnosis and Incidence of Sickle Cell Disease and Sickle Cell Trait in U.S. African-Americans

Syndrome (Genotype)	Incidence	Clinical Severity	Hgb (g/dL) ^a	MCV (fl) ^a	Reticulocytes (%) ^a	Newborn Screening ^b	Electrophoresis (%)
Sickle cell anemia (SS)	1:400	Usually marked	6.5–9.5	>80	5–20	FS	S 80–90 F 2–20 A ₂ < 3.6
Sickle hemoglobin C (SC)	1:1000	Mild to moderate	9.5–13.5	75–95	5–10	FSC	S 45–55 C 45–55
Sickle beta plus (Sβ ⁺) thalassemia	1:4000	Mild to moderate	8.5–12.5	< 75	5–10	FSA or FS ^c	S 65–90 A 5–30 F 2–20 A ₂ > 3.6
Sickle beta zero (Sβ ⁰) thalassemia	1:10000	Marked to moderate	6.5–9.5	< 80	5–20	FS	S 80–92 F 2–10 A ₂ > 3.6
Sickle cell trait (AS)	1:10	Asymptomatic	Normal	Normal	Normal	FAS	S 35–45% A 55–65%

^aHematologic values are approximate; results apply to older children

^bHemoglobins reported in order of quantity (e.g., FSA = F > S > A); F, fetal hemoglobin; S, sickle hemoglobin; C, hemoglobin C; A, hemoglobin A.

^cQuantity of Hb A at birth; sometimes insufficient to quantitate (AAP, 2002, NIH, 2002)

5. Serum iron and iron binding capacity
 - a. Decreased total serum iron
 - b. Increased iron binding capacity (TIBC)
 - c. Decreased iron saturation (16% or less)
 6. Free erythrocyte protoporphyrin (FEP)—elevated
- Management/Treatment: According to American Academy of Pediatrics guidelines, children should routinely be screened between the ages of 9 to 12 months, and additionally for child at risk, between 1 to 5 years of life
 1. Nutritional counseling—prevention
 - a. Maintain breastfeeding for 6 months if possible; supplemental iron drops or iron fortified cereal by 4 to 5 months of age
 - b. If not breastfed, use iron fortified infant formula until 1 year of age
 - c. Use iron fortified cereals from 6 to 12 months of age
 - d. No cow's milk before 1 year of age, then limit to 18 to 24 oz/day
 - e. Prescribe 2 to 3 mg/kg/day elemental iron in 1 to 2 doses/day for prophylaxis in low birth weight infants
 2. Oral iron medication
 - a. Prescribe as elemental iron—3 to 6 mg/kg/day in 1 to 3 doses until hemoglobin normal, then 2 to 3 mg/kg/day for 4 months to replace iron stores
 - b. Failure to respond—consider the following reasons in this order
 - (1) Failure or inconsistent administration of medication
 - (2) Persistent or unrecognized blood loss
 - (3) Incorrect diagnosis
 - (4) Impaired GI absorption
 3. Parenteral iron dextran—consider for use in cases of noncompliance, severe bowel disease, genuine intolerance, chronic hemorrhage, chronic diarrhea
 4. Packed red cell transfusion—is seldom necessary but may be considered in children who are debilitated and/or chronically ill and are symptomatic (especially with cardiac dysfunction), or when hemoglobin level is ≤4 g/dL

■ SICKLE CELL DISEASE

- Definition: A group of hemoglobinopathies that include inheritance of two hemoglobins that sickle; these hemoglobins include Hgb S, C, O_{Arab}, D, G_{Philadelphia}, and α-chain mutant; transmitted as an autosomal recessive gene
- Etiology/Incidence
 1. Hemoglobin S results from a single base pair substitution of valine for glutamic acid at the sixth position of the beta (β) globin gene; this results in red blood cells that become sickle

shaped when deoxygenated; sickled shape of the RBCs leads to hemolysis and intermittent episodes of vascular occlusion that can cause tissue ischemia and acute and chronic organ dysfunction

2. Sickle cell disease occurs in individuals of African, Mediterranean, Indian, and Middle Eastern descent; incidence and clinical severity of sickle cell disease and trait in U.S. African-American population is listed in **Table 13-2**
- Signs and Symptoms: Vary with associated problems (see Table 13-2)
 1. Infection (peak incidence between 1 and 3 years of age)
 - a. Fever
 - b. Malaise
 - c. Anorexia
 - d. Poor feeding
 2. Acute painful events—“pain crisis,” “vaso-occlusive crisis”
 - a. Pain—most often in bones, but can occur in any part of body (chest, stomach, hands, and/or feet, back, etc.); in children < 2 years of age usually in hands and/or feet (dactylitis)
 - b. Swelling—sometimes seen at site of pain
 - c. Low grade fever—sometimes occurs

■ **Table 13-2** Associated Problems of Sickle Cell Disease

Hemolysis
Chronic anemia
Jaundice
Aplastic crisis ^a
Cholelithiasis
Delayed growth and sexual maturity
Vaso-occlusion
Recurrent acute pain (e.g., dactylitis, musculoskeletal, abdominal)
Functional asplenia (bacterial infections ^a)
Splenic sequestration ^a
Acute chest syndrome ^a
Stroke ^a
Hyposthenuria and enuresis
Papillary necrosis of kidneys
Chronic nephropathy
Priapism
Avascular necrosis of humeral heads, femoral heads
Proliferative retinopathy
Leg ulcers

^a Potential cause of mortality. (AAP, 2002, NIH, 2002)

3. Splenic sequestration
 - a. Weakness
 - b. Irritability
 - c. Unusual sleepiness
 - d. Paleness
 - e. Large spleen
 - f. Fast heart rate
 - g. Pain in the left side of the abdomen—does not always occur
 4. Aplastic crisis (is associated with parvovirus B19)
 - a. Pale
 - b. Malaise
 - c. Headache
 - d. Fever
 - e. Mild upper respiratory infection symptoms
- Differential Diagnosis
 1. Infection
 - a. Septicemia
 - b. Meningitis
 - c. Pneumonia
 - d. Osteomyelitis
 - e. Viral illness
 2. Acute painful events
 - a. Bone pain
 - (1) Bone infarct
 - (2) Osteomyelitis
 - (3) Rheumatoid arthritis
 - (4) Leukemia
 - b. Abdominal pain
 - (1) Cholelithiasis—right upper quadrant
 - (2) Splenic infarct—left upper quadrant
 - (3) Functional abdominal pain
 - (4) Gas pain
 3. Splenic sequestration—chronic splenomegaly
 4. Aplastic crisis—other viral illnesses
 - Physical Findings: Presence variable
 1. Jaundice
 2. Cardiac murmur
 3. Splenomegaly
 4. Pallor
 - Diagnostic Tests/Findings
 1. Prenatal diagnosis—can be done by analysis of DNA obtained through chorionic villus sampling (9 to 11 weeks gestation) or amniocentesis (11 to 17 weeks gestation)
 2. Usual diagnostic hemoglobin electrophoresis and hematologic results in infants and adults with sickle cell disease are presented in Table 13-1
 3. “Sickle prep” (metabisulfite solution)—should not be used as diagnostic test as it does not distinguish among sickle cell anemia, other

forms of sickle cell disease, and sickle cell trait or common interacting hemoglobinopathies such as Hgb C or thalassemia

4. Splenic sequestration
 - a. Hemoglobin—below baseline (steady state)
 - b. Platelets—decreased
 5. Aplastic crisis
 - a. Hemoglobin—below steady state
 - b. Reticulocyte count— < 1.0%
- Management/Treatment
 1. Maintenance care—should be directed toward prevention of complications and crisis precipitating factors, and should include:
 - a. Should receive all immunizations as recommended by American Academy of Pediatrics including pneumococcal, meningococcal, and annual influenza vaccines
 - b. In addition to routine conjugate pneumococcal (PCV) vaccine received at 2, 4, 6, and 12 or 15 months should then receive polysaccharide pneumococcal vaccine (PPV) every 5–10 years as recommended by current guidelines
 - c. Prophylactic penicillin—initiate by 2 months of age and continue until at least 5 years of age
 - d. Family education regarding increased risk of infection due to functional asplenia and the need to seek medical attention promptly for evaluation of febrile illnesses
 - e. Because of the increased risk for infection with an encapsulated bacteria, any fever greater than 101.5°F (38.5°C) requires immediate evaluation with a physical exam, CBC and blood culture, and if child looks septic, a dose of ceftriaxone
 - f. For children with frequent complications, daily hydroxyurea has been reported to decrease hemolysis and painful crises
 2. Acute painful events
 - a. Prevention—education regarding factors that may precipitate painful events, i.e., dehydration, hypoxia, fever, exposure to extreme temperatures; how to manage mild to moderate pain; and how to recognize signs of serious problems
 - b. Home-based management
 - (1) Analgesia as prescribed
 - (2) Nonpharmacologic treatment such as heat, localized massage
 - c. Emergency department (ED) management—parenteral fluids and analgesics
 - d. Inpatient management—if pain not reduced by ED management,

hospitalization for hydration, parenteral analgesics, and concomitant NSAID

3. Acute exacerbations of anemia (such as aplastic crisis)
 - a. Packed red blood cell transfusions may be indicated
 - b. Exchange transfusions may be indicated for severe vaso-occlusive associated problems (e.g., organ failure, stroke)
4. Management of other problems associated with sickle cell disease—consultation with or referral to a pediatric hematologist
5. Frequent reinforcement of education regarding fever and pain management
6. Anticipatory guidance regarding physiological and psychological effects of chronic illness and sickle cell disease specifically, including delayed growth, enuresis, school attendance, physical endurance
7. Genetic counseling

▣ LEAD POISONING (PLUMBISM)

- Definition: A chronic disease caused by the accumulation of toxic amounts of lead in the body; the CDC defines lead poisoning as a whole blood lead level (BLL) $\geq 10 \mu\text{g}/\text{dL}$
- Etiology/Incidence
 1. Ingestion or inhalation of lead or lead compounds; transplacental transmission may also occur
 2. Sources of lead exposure
 - a. Lead-based paint in older homes built prior to 1978, especially housing built before 1950
 - b. Lead contaminated soil and dust from automobile emissions (decreasing with use of lead-free gasoline)
 - c. Lead contaminated drinking water (lead or lead-soldered pipes)
 - d. Certain Mexican and Indian lead-containing folk remedies
 - e. Lead based paint on imported items, including toys
 3. Highest prevalence among poor, inner-city children living in older, deteriorating housing
 4. Children between 1 and 3 years of age at greatest risk
 5. Lead toxicity may contribute to neurobehavioral, as well as cognitive, morbidities of childhood
- Signs and Symptoms: Vary with degree of exposure
 1. Low level exposure may be asymptomatic
 2. Mild acute lead poisoning—resembles gastroenteritis, e.g., anorexia, nausea, vomiting,

constipation or diarrhea, and abdominal pain; other possible symptoms are sleep disturbances, metallic taste in mouth, limb pain, and headaches

3. Severe lead poisoning—lethargy, difficulty walking, tingling, cognitive impairment, personality changes
 4. Approximately 90% of children with lead poisoning will have pica
- Differential Diagnosis
 1. Iron deficiency anemia
 2. α or β thalassemia
 3. Metabolic disorders
 4. Developmental or cognitive delay secondary to other causes
 - Physical Findings
 1. May see bluish discoloration of gingival border (Burtonian blue lines)
 2. Bradycardia
 3. Neuropathy
 4. Papilledema
 5. Ataxia
 - Diagnostic Tests/Findings
 1. CDC revised guidelines (2002) recommend universal screening for Medicaid children and those deemed at risk, such as those who live in homes built before 1978
 2. Whole blood lead level test (BLL) by categories of risk
 - a. Lead level $< 10 \mu\text{g/dL}$ —not considered blood poisoning
 - b. Lead level of 10 to $14 \mu\text{g/dL}$ —considered borderline (venipuncture confirmation within 3 month)
 - c. Lead level of 15 to $19 \mu\text{g/dL}$ (venipuncture confirmation within 3 months)
 - d. Lead level of 20 to $44 \mu\text{g/dL}$ (venipuncture confirmation within 1 week to 1 month)
 - e. Lead level of 45 to $69 \mu\text{g/dL}$ (diagnostic venous blood testing within 24 to 48 hours)
 - f. Lead level of $> 70 \mu\text{g/dL}$ (medical emergency, retest immediately)
 - Management/Treatment
 1. Follow-up BLL monitoring—frequency determined based on whether low or high risk and BLL obtained
 - a. Lead level $< 10 \mu\text{g/dL}$ —if high risk, retest in 6 months; if low risk, no further testing needed
 - b. Lead level of 10 to $14 \mu\text{g/dL}$ —at risk for lead poisoning, early follow-up within 3 months; if follow-up level is 15 to $19 \mu\text{g/dL}$ or higher 3 months apart, retest every 1 to 2 months until results $< 15 \mu\text{g/dL}$ for at least 6 months, then every 6 to 9 months
 - c. Lead level of 15 to $19 \mu\text{g/dL}$ —at risk for lead poisoning, early follow-up within 2 months; if follow-up level is > 15 , retest every 1 to 2 months until results $< 15 \mu\text{g/dL}$ for at least 6 months, then every 3 months until child is 36 months old; include educational and nutritional counseling
 - d. Lead level of 20 to $44 \mu\text{g/dL}$ —early follow-up in 1 to 2 months for BLLs 20 to $24 \mu\text{g/dL}$; 2 weeks to 1 month for BLLs 25 to $44 \mu\text{g/dL}$, retest every 1 to 2 months until results $< 15 \mu\text{g/dL}$ for at least 6 months, then every 3 months until child is 36 months old
 - e. Lead level of 45 to $69 \mu\text{g/dL}$ —early follow-up as soon as possible; retest every month until results $< 15 \mu\text{g/dL}$ for at least 6 months, then every 3 months until child is 36 months old
 - f. Lead level of $> 70 \mu\text{g/dL}$ —early follow-up as soon as possible, retest every month until results $< 15 \mu\text{g/dL}$ for at least 6 months, then every 3 months until child is 36 months old
 2. For blood lead level $> 20 \mu\text{g/dL}$, refer for further medical evaluation, interventions, and follow-up
 - a. Test for iron deficiency
 - b. Environmental assessment and removal of known sources of lead in environment
 - c. Medical evaluation for symptoms and possible pharmacologic management
 - (1) Lead level of 20 to $44 \mu\text{g/dL}$ may require pharmacologic management
 - (2) Lead level of $> 45 \mu\text{g/dL}$ will require chelation therapy with Succimer
 - (3) Parenteral chelation with dimercaprol and calcium edentate is used for cases of lead encephalopathy
 3. Primary prevention
 - a. Outreach education regarding nutrition and avoidance of exposure
 - b. Assessment of potential risk with specific environmental and health questions during routine well-child visits

■ GLUCOSE-6-PHOSPHATE DEHYDROGENASE (G-6-PD) DEFICIENCY

- Definition: X-linked autosomal recessive disorder in which activity of red cell enzyme G-6-PD is decreased or absent causing hemolytic anemia

- Etiology/Incidence
 1. Lack of G-6-PD decreases the ability to deal with oxidative stress and results in hemolysis; episodes of hemolysis may be induced by the following:
 - a. Drugs such as aspirin, sulfonamides, antimalarials
 - b. Fava beans, ingestion or exposure to pollen from the bean's flower (occurs in Mediterranean- and Canton-type deficiencies)
 - c. Infection (in more susceptible individuals)
 2. Many genetic variants described with altered enzyme levels which determines the severity of disorder
 - a. A type—enzyme activity decreases with age of cell; hemolyzes only old RBC; occurs in 11% African-American males, 2% African-American females
 - b. Mediterranean type—associated with severe deficiency of enzyme
 - c. Canton type—severe disease in Asians
 3. Most common among individuals of African, Mediterranean, or Asian descent
- Signs and Symptoms: Symptoms develop 24 to 48 hours after ingestion of substance having oxidant properties
 1. Weakness
 2. Pale appearance
 3. Severe case
 - a. Blood in the urine
 - b. Yellow discoloration of skin, whites of eyes, and gums or inside of mouth
- Differential Diagnosis
 1. Other causes of hemolytic anemia
 2. Hemoglobinopathies
- Physical Findings
 1. Hyperbilirubinemia in infants—usually associated with Mediterranean and Canton individuals
 2. Older children are asymptomatic between episodes of hemolysis
- Diagnostic Tests/Findings
 1. G-6-PD fluorescence-based screen (may give false negative)
 2. Red blood cell indices during or just after hemolytic episode
 - a. Heinz bodies present
 - b. Fragmented cells and blister cells
 3. Reticulocytosis
 4. Hemoglobin—usually normal between episodes of hemolysis; may be decreased in Mediterranean or Canton types

5. Acute self-limiting hemolytic anemia with hemoglobinuria (type A variant)
6. Acute life-endangering hemolysis often leads to acute renal failure (all other variants)

- Management/Treatment
 1. Generally mild symptoms require minimal intervention
 2. Identification and avoidance of foods and drugs that cause hemolysis
 3. Transfusion for severe hemolysis
 4. Genetic counseling; routine screening not generally recommended

■ BLEEDING DISORDERS

Hemophilia

- Definition: Bleeding disorder caused by congenital deficiency of clotting factor VIII or IX
- Etiology/Incidence
 1. Hemophilia A (factor VIII deficiency, classical hemophilia)
 - a. Third most common X-linked disorder, but 20% to 30% are caused by spontaneous mutation
 - b. Approximately 1:5000 male births
 - c. 10% to 30% of patients develop antibodies against the functional activity of factor VIII
 - d. Factor VIII deficiency is 4 times more common than factor IX deficiency
 2. Hemophilia B (factor IX deficiency, Christmas disease)—X-linked disorder, with 20% to 30% spontaneous mutation
- Signs and Symptoms: Vary based on severity of factor deficiency
 1. Easy bruising at injection sites
 2. Prolonged bleeding following circumcision—not all severe hemophiliacs bleed post circumcision
 3. Excessive bruising after child begins walking
 4. Prolonged bleeding in any part of the body
 5. May have pain at the site of the bleed
 6. Hemarthrosis
- Differential Diagnosis
 1. Thrombocytopenia
 2. von Willebrand disease
 3. Vitamin K deficiency
 4. Disseminated intravascular coagulation (DIC)
 5. Child abuse
- Physical Findings: Signs of bleeding including ecchymoses, swelling and pain in joints, and prolonged bleeding from lacerations or injections

- Diagnostic Tests/Findings
 1. Prenatal diagnosis—can be done by fetal blood sampling (periumbilical blood sampling [PUBS]) (18 to 20 weeks gestation) through fetoscopy or by analysis of DNA obtained through chorionic villus sampling (9 to 11 weeks gestation) or amniocentesis (11 to 17 weeks gestation)
 2. Diagnostic test—direct assay of plasma factor activity level for hemophilia A and B—see **Table 13-3**
 3. Screening tests
 - a. Activated partial thromboplastin time (APTT)—prolonged
 - b. Prothrombin time (PT)—normal
 - c. Bleeding time (not indicated)—normal
- Management/Treatment
 1. Collaborative interdisciplinary approach facilitated by regional hemophilia treatment center
 2. Factor replacement therapy
 - a. Hemophilia A—factor VIII concentrate intravenously
 - b. Hemophilia B—factor IX concentrate intravenously
 - c. Approximately 10% to 15% of patients develop anti-factor antibodies and may require other therapy
 3. Prophylaxis
 - a. Primary prophylaxis—regular infusions of factor VIII or IX given to prevent joint hemorrhage and bleeding episodes
 - b. Secondary prophylaxis—started after a joint has developed a pattern or repeated

■ **Table 13-3** Relationship of Factor Levels to Severity of Clinical Manifestations of Hemophilia A and B

Type	% Factor VIII/IX	Type of Hemorrhage
Severe	< 1	Spontaneous hemarthrosis and deep tissue hemorrhages
Moderate	1–5	Gross bleeding following moderate trauma; some hemarthrosis; rare spontaneous hemorrhage
Mild	6–49	Severe hemorrhage only following moderate to severe trauma or surgery

- bleeding; used to prevent further bleeding into the joint
- 4. Desmopressin intranasally or intravenously—for mild FVIII deficiency
- 5. Antifibrinolytic therapy—for oral mucosal bleeds
- 6. Physical therapy—for musculoskeletal bleeds
- 7. Surgery—synovectomy, arthroscopic or with use of isotopes
- 8. Anticipatory guidance regarding developmental issues such as discipline, child-care, and schooling
- 9. Genetic counseling

von Willebrand Disease (VWD)

- Definition: An inherited hemorrhagic disorder characterized by defective primary hemostasis and due to a quantitative or qualitative abnormality in von Willebrand factor (VWF)
- Etiology/Incidence
 1. The most common congenital bleeding disorder
 2. Six variant types
 - a. Type 1—previous Type I
 - b. Type 2A—previous Type I B
 - c. Type 2M—previous Type B
 - d. Type 2B—previous Type II B
 - e. Type 2N—previous Type Normandy
 - f. Type 3—previous Type III
 3. Affects at least 1% of the population, but only 10% are symptomatic
- Signs and Symptoms: Great variation in frequency, severity, and bleeding manifestations
 1. Nosebleeds
 2. Bleeding gums
 3. Heavy menstrual bleeding
 4. Prolonged oozing from cuts
 5. Increased bleeding after trauma or surgery
- Differential Diagnosis
 1. Thrombocytopenia
 2. Hemophilia
 3. Vitamin K deficiency
 4. Disseminated intravascular coagulation
- Physical Findings
 1. Easy bruising
 2. Multiple sites of bruising
 3. Oozing or bleeding at trauma or surgical site
- Diagnostic Tests/Findings
 1. Variation in findings by specific type of VWD, as well as within same patient over time
 2. Bleeding time—usually prolonged

3. von Willebrand factor—usually decreased or absent
 4. von Willebrand factor antigen—usually decreased or absent
 5. Blood group—VWF factor decreased in type O
- Management/Treatment
 1. Desmopressin acetate—used to treat bleeding complications or as preoperative preparation for Type 1; contraindicated in the treatment of Type 2B
 2. Alternative treatment if desmopressin not indicated or effective—treatment with factor concentrates with intact von Willebrand factor should be considered
 3. Antifibrinolytic agents—particularly effective in areas where fibrinolysis appears to contribute to bleeding, i.e., in mucous membranes (nose, mouth, throat, and menorrhagia), trauma, and dental procedures and surgery

Idiopathic Thrombocytopenia (ITP, Autoimmune Thrombocytopenia)

- Definition: Immune-mediated disorder characterized by production of antiplatelet antibodies
 1. Acute ITP—platelet count returns to normal ($> 150,000/\text{mm}^3$) within 6 months
 2. Chronic ITP—platelet count low beyond 6 months; approximately 10% of children with ITP
 3. Recurrent or relapsing ITP—the platelet count decreases again after having returned to normal levels (rare)
- Etiology/Incidence
 1. Autoimmune disorder, sometimes related to sensitization by viral infection
 2. 4 to 5.3 cases/100,000 children
 3. Most prevalent during early- to mid-childhood and in older adults, although it affects all ages
 4. Chronic ITP—more common in children > 7 years of age; more prevalent in females (3:1 ratio)
- Signs and Symptoms
 1. Bruising
 2. Nosebleeds
 3. Bleeding of gums and lips
 4. Petechiae
 5. Menorrhagia
 6. Child appears well, except for signs of bleeding
- Differential Diagnosis
 1. Thrombocytopenia of other causes
 - a. Bone marrow infiltration
 - b. Septicemia
 - c. Aplastic anemia
 2. Disseminated intravascular coagulation (DIC)
 3. Hemolytic uremic syndrome
 4. Acute leukemia
 5. Evan's syndrome (thrombocytopenia and hemolytic anemia)
 6. In young children
 - a. Congenital amegakaryocytic thrombocytopenia-absent radius syndrome
 - b. Wiskott-Aldrich syndrome (only in males)
 7. In older children, especially those with chronic symptoms
 - a. Systemic lupus erythematosus
 - b. HIV infection
 - c. Lymphoma
- Physical Findings
 1. Petechiae, purpura, and ecchymoses
 2. Hemorrhages in mucous membranes
 3. Pallor usually not present (unless there has been significant bleeding)
 4. Splenomegaly
- Diagnostic Tests/Findings
 1. CBC—generally the only required test
 2. Hemoglobin—normal or slightly reduced with prior bleeding; can drop with bruising only
 3. Platelet count
 - a. $\leq 20,000/\text{mm}^3$ (diagnostic for acute ITP); often $\leq 10,000/\text{mm}^3$
 - b. $< 100,000/\text{mm}^3$ for > 6 months (diagnostic for chronic ITP)
 4. WBC—normal; if active infection, may have increased neutrophils, lymphocytes, or atypical mononuclear cells
 5. Bleeding time test (unnecessary)—always abnormal if platelets $< 50,000/\text{mm}^3$
- Management/Treatment: Controversial
 1. Acute ITP
 - a. Treatment usually not indicated if platelet count $> 50,000/\text{mm}^3$
 - b. Treatment usually considered if platelet count $< 20,000/\text{mm}^3$, especially with extensive cutaneous (and especially mucosal) bleeding or if protective environment cannot be assured
 - c. Acute bleeds, especially intracranial or gastrointestinal, may require therapy
 - d. Pharmacologic management
 - (1) High-dose corticosteroids (prednisone 2 to 5 mg/kg/day orally for 1 to 3 weeks) with slow wean
 - (2) Intravenous gamma globulin (IVIG)—1 g/kg/day for 1 to 3 days is the treatment of choice for acute bleeds; platelets may also be given but are short-lived

2. Chronic ITP
 - a. Referral to a hematologist
 - b. Pharmacologic management—when thrombocytopenia worsens during viral illness or prior to elective surgery, prednisone or IVIG can be administered
 - c. Prolonged use of high dose steroids should be avoided, but low dose maintenance steroids may be necessary for a prolonged period of time
 - d. Antacids, H₂ blockers, and/or proton pump inhibitors need to be taken with steroids
 - e. Monoclonal antibody therapy (e.g., rituximab) may be effective in refractory autoimmune thrombocytopenia
 - f. Splenectomy—treatment of choice when disease is severe or symptomatic chronic ITP
 - g. Post-splenectomy care
 - (1) All immunizations, including meningococcal, pneumococcal polysaccharide, and annual influenza vaccines should be given as per CDC recommendations
 - (2) Prophylactic penicillin or amoxicillin daily
 - (3) Because of the increased risk for infection with an encapsulated bacteria, any fever greater than 101°F requires immediate evaluation with a physical exam, CBC, and blood culture; and if child looks septic, a dose of ceftriaxone
3. Patient/family education
 - a. Avoid all competitive contact sports that could result in head trauma or ruptured spleen
 - b. Avoid aspirin and aspirin-containing medications and NSAIDs
 - c. Monitor for signs of occult bleeds
 - d. Seek medical care if signs of bleeding are noted

■ CANCERS

Leukemias

- Definition: A malignant neoplasm of bone marrow characterized by proliferation of immature white cells
- Etiology/Incidence
 1. Etiology is usually unknown
 2. Accounts for 25% to 30% of all childhood cancers
- Acute leukemia constitutes 97% of all childhood leukemias and includes most common types
 - a. Acute lymphoblastic leukemia (ALL)—75%; survival rate is 60% to 80%
 - b. Acute myeloid leukemia (AML), also known as acute nonlymphocytic leukemia (ANLL)—20%; survival rate is 40% to 60%
- Chronic myelogenous leukemia (CML) constitutes 3% of childhood leukemia
- 1 per 25,000 of population up to 14 years of age
- Peak incidence between 2 and 5 years of age
- Signs and Symptoms
 1. Fatigue, headache
 2. Bruising
 3. Fever
 4. Nosebleeds
 5. Bone pain, limp
- Differential Diagnosis
 1. Chronic infections such as Epstein-Barr virus (EBV) or cytomegalovirus (CMV)
 2. ITP
 3. Transient erythroblastopenia of childhood
 4. Aplastic anemia
 5. Juvenile rheumatoid arthritis
- Physical Findings
 1. Pallor
 2. Purpura, ecchymosis
 3. Organomegaly (liver/spleen), adenopathy
- Diagnostic Tests/Findings
 1. CBC—presence of blast cells on peripheral blood smear highly suggestive; needs confirmatory bone marrow examination
 2. Bone marrow aspiration/biopsy (required for diagnosis)—bone marrow replaced by > 30% blasts, usually 80% to 100%
- Management/Treatment
 1. Combination chemotherapy (protocol length 108 to 130 weeks)
 2. CNS prophylaxis
 - a. Radiation therapy (for high-risk patients)
 - b. Combined intrathecal chemotherapy
 3. Patient/family education regarding side effects of therapy, anticipatory guidance on coping
 4. For relapse on therapy, bone marrow transplant is recommended; relapse after therapy is completed, a second course of chemotherapy may provide cure; for AML, bone marrow transplant is treatment of choice; if no suitable donors available standard chemotherapy is utilized

5. Primary care considerations—immunizations held while on therapy and resume 6 to 12 months after therapy completed (institutional specific policy); sibling immunizations not affected; fever on therapy requires CBC/blood cultures and hospitalization for neutropenia with fever or if patient has central line
6. Long-term follow-up for delayed effects of chemotherapy and/or radiation

Neuroblastoma

- Definition: Tumor mass along the neural pathway
- Etiology/Incidence
 1. Etiology—possible genetic factors; familial predisposition
 2. Most common malignancy in infancy, accounts for 7% of all childhood malignancies
 3. Metastases at onset in 70% to 75% of cases
 4. 10 per 1 million live births annually
 5. Survival rate for patients < 1 year of age, 82%; for 1 to 2 years of age, 32%; over 2 years, 10%
- Signs and Symptoms: Dependent on primary site, presence of metastases
 1. Listlessness
 2. Poor feeding
 3. Pale
 4. Weight loss
 5. Abdominal pain
 6. Weakness
 7. Irritability
- Differential Diagnosis
 1. Trauma
 2. Lymphadenopathy
 3. Leukemia
 4. Lymphoma
 5. Wilms tumor
- Physical Findings: Related to site of primary tumor
 1. Lymph node enlargement
 2. Hepatomegaly
 3. Abdominal or flank mass
 4. Periorbital ecchymoses (raccoon eyes)
 5. Scalp or skin nodules (often bluish coloration)
- Diagnostic Tests/Findings
 1. CT and/or MRI scan to determine site and location of any suspected masses
 2. Tissue biopsy to confirm diagnosis
 3. Bone marrow aspiration and biopsy to evaluate for infiltrating tumor
 4. Serum or urine catecholamine levels—may be increased and followed for tumor response/recurrence

- Management/Treatment: Based on stage and site of tumor
 1. Treatment of emergent symptoms
 2. Surgery
 - a. Staging excision of tumor
 - b. Evaluation of treatment (early stage/low risk disease may have observation only)
 3. Radiation therapy (for resistant/relapse disease)
 4. Combination chemotherapy
 5. Bone marrow transplant for high risk disease
 6. Patient/family education regarding side effects of therapy
 7. Anticipatory guidance on developmental issues such as sleep, toileting, discipline, and child-care
 8. Primary care considerations—immunizations held while on therapy and resume 6 to 12 months after therapy completed (institutional specific policy); sibling immunizations not affected; fever on therapy requires CBC/blood cultures and hospitalization for neutropenia with fever or if patient has central line
 9. Long-term follow-up for delayed effects of chemotherapy and/or radiation

Retinoblastoma

- Definition: Congenital malignant intraocular tumor
- Etiology/Incidence
 1. Abnormal fetal neural crest cell maturation
 - a. Hereditary—germinal mutation (40%); siblings and subsequent offspring at risk
 - b. Acquired—somatic mutations
 2. Most common intraocular childhood tumor
 - a. 1:10,000 live births
 - b. Slightly higher incidence in males
 - c. Accounts for 7% of all childhood cancers; 15% childhood cancer mortality
 3. Majority diagnosed before 5 years— $\frac{1}{3}$ are bilateral; usually diagnosed in first year
 4. Prognosis varies with age and tumor staging
 - a. Overall survival rates > 90%
 - b. Highest survival for children < 1 year and > 6 years
 - c. High incidence of secondary malignancies with hereditary form may limit survival into third or fourth decade of life
- Signs and Symptoms
 1. Squinting
 2. Eyes turning inward or outward
 3. Painful red eye

- Differential Diagnosis
 1. Cataract
 2. Retinal detachment
 3. Persistent hyperplastic primary vitreous
 4. Coloboma
 5. Retinopathy of prematurity
- Physical Findings
 1. Leukocoria—yellow-white pupillary reflex (most common presentation)
 2. Strabismus
 3. Hyphema may be present
- Diagnostic Tests/Findings
 1. Fundoscopic examination under anesthesia or sedation—findings may include:
 - a. Creamy-pink mass
 - b. White avascular tumor mass
 2. CT of the orbits to evaluate extent of tumor and to determine optic nerve or bony structure involvement
 3. MRI to assess for optic nerve invasion
- Management/Treatment: Determined by stage and extent of disease
 1. Surgery
 - a. Resection
 - b. Enucleation
 2. Radiation therapy
 3. Photocoagulation (laser therapy)
 4. Cryotherapy
 5. Chemotherapy—advanced or recurrent disease
 6. Patient/family education regarding side effects of therapy
 7. Anticipatory guidance on developmental issues such as sleep, toileting, discipline, and child-care; referral to genetics for family planning issues
 8. Long term follow-up for recurrence or delayed effects of chemotherapy and/or radiation

Lymphomas

- Definition: Malignant disorders characterized by proliferation of cells, usually restricted to lymphoid cells but may be found in bone marrow; includes Hodgkin's (HL) and non-Hodgkin's (NHL) lymphomas
- Etiology/Incidence
 1. Etiology unknown—possible etiologic factors
 - a. Genetic predisposition
 - b. Environmental exposures
 - c. Epstein-Barr virus
 - d. Immunologic disorders
 2. Third most common childhood cancer—10%
 - a. HL—1:100,000 children

- (1) Peak age—bimodal age incidence curve
 - (a) 15 to 35 years
 - (b) > 50 years
 - (2) Survival—stages I to II, most are cured; stage III, 75% to 90%; stage IV, 60% to 80%
- b. NHL—15:100,000 children between 5 to 10 years
- (1) Peak age—5 to 15 years
 - (2) Survival—stages I to II, 90%; stages III to IV approximately 70%

- Signs and Symptoms
 1. Hodgkin's
 - a. Painless swelling of lymph nodes
 - b. Fatigue (B symptom)
 - c. Decreased appetite and weight loss (> 10% weight loss B symptom)
 - d. Fever (B symptom)
 - e. Night sweats (B symptom)
 2. Non-Hodgkin's
 - a. Asymptomatic if not disseminated
 - b. Difficulty swallowing or breathing
 - c. Swelling in neck, face, upper extremities
 - d. Abdominal pain
- Differential Diagnosis
 1. Other malignancy
 2. Lymphadenopathy associated with infection
- Physical Findings
 1. Hodgkin's
 - a. Affected nodes—often fixed, firm, nontender, discrete; cervical and supraclavicular areas most common
 - b. Splenomegaly
 2. Non-Hodgkin's
 - a. Similar to Hodgkin's
 - b. May vary depending on degree of involvement
- Diagnostic Tests/Findings
 1. Hodgkin's
 - a. Chest radiograph—to explore possibility of mediastinal involvement and examine airway patency
 - b. PET scan to evaluate disease involvement
 - c. Biopsy—tumor giant cells (Reed-Sternberg cells)
 - d. CBC with red cell indices
 - (1) Hemoglobin—decreased
 - (2) Normocytic and normochromic or microcytic and hypochromic
 - e. Serum copper—increased
 - f. Serum ferritin—increased
 - g. Sedimentation rate (ESR)—increased

2. Non-Hodgkin's—tissue diagnosis necessary before treatment is started, findings vary based on specific histologic type
 - a. Isolated peripheral nodes—excisional or fine-needle biopsy
 - b. Mediastinal mass—thoracotomy or mediastinoscopy, parasternal fine-needle biopsy, or thoracentesis (if associated pleural effusion)
 - c. Abdominal mass—open biopsy usually necessary
- Management/Treatment: Plan developed based on stage of involvement
 1. Multiagent chemotherapy
 2. Radiation
 3. Patient/family education regarding side effects of therapy
 4. Primary care considerations—immunizations held while on therapy and resume 6 to 12 months after therapy completed (institutional specific policy); sibling immunizations not affected; fever on therapy requires CBC/blood cultures and hospitalization for neutropenia with fever or if patient has central line
 5. Long term follow-up for delayed effects of chemotherapy and/or radiation

Wilms Tumor (Nephroblastoma)

- Definition: A primary malignant renal tumor
- Etiology/Incidence
 1. Etiology unknown; predisposing factors
 - a. Genetic factors
 - (1) Associated with aniridia, Beckwith-Wiedeman syndrome
 - (2) Familial predisposition
 - b. Environmental factors—chronic chemical exposure (hydrocarbons, lead)
 2. 6% of all cancer in children; 9:1 million Caucasian children/year
 3. Equal frequency in males and females
 4. 78% diagnosed between 1 and 5 years of age; peak incidence 3 to 4 years of age
 5. Survival rate—70% to 90%
- Signs and Symptoms
 1. May be asymptomatic
 2. Abdominal mass—usually nonpainful
 3. Occasionally abdominal pain
 4. Malaise, fever, loss of appetite
 5. Vomiting
 6. Blood in urine
- Differential Diagnosis
 1. Hydronephrosis

2. Polycystic kidney disease
3. Neuroblastoma
4. Rhabdomyosarcoma
5. Lymphoma

- Physical Findings
 1. Abdominal mass—usually asymptomatic
 2. Hypertension
 3. Associated congenital anomalies—aniridia, hemihypertrophy, genitourinary anomalies
- Diagnostic Tests/Findings
 1. CBC and urine analysis for anemia, hematuria
 2. CT of abdomen to evaluate:
 - a. Presence and function of opposite kidney
 - b. Evidence of bilateral involvement
 - c. Evidence of involvement of blood vessels of tumor
 - d. Lymph node involvement
 - e. Liver involvement
 3. Abdominal ultrasound—may indicate that the tumor is intrarenal
 4. Chest radiograph, CT of chest to evaluate for metastasis to lung
- Management/Treatment: Plan developed based on stage of involvement
 1. Surgical excision
 - a. If unilateral—complete nephrectomy
 - b. If bilateral—nephrectomy of more involved site, excision biopsy/partial nephrectomy of smaller lesion in remaining kidney; or no surgery with radiation
 2. Multiagent chemotherapy
 3. Radiation therapy
 4. Patient/family education regarding side effects of therapy
 5. Primary care considerations—immunizations held while on therapy and resume 6 to 12 months after therapy completed (institutional specific policy); sibling immunizations not affected; fever on therapy requires CBC/blood cultures and hospitalization for neutropenia with fever or if patient has central line
 6. Long-term follow-up for disease recurrence and delayed effects of chemotherapy and/or radiation

Osteosarcoma

- Definition: A solid tumor of the bone in which malignant spindle cell stroma produce osteoid; most common form of bone cancer in children
- Etiology/Incidence
 1. Etiology unknown
 2. Associated factors

- a. Genetic factors and family predisposition
 - (1) Increased risk with hereditary retinoblastoma
 - (2) Increased risk with chromosome 13 abnormalities
 - b. Environmental factors—increased risk associated with previously irradiated bone
 - c. Increased risk with taller children (may appear during growth spurt)
3. Incidence—7 teenagers per one million
 - a. Peak incidence during adolescent growth spurt between 15 to 19 years of age
 - b. Male incidence slightly higher than females
- Signs and Symptoms
 1. Local pain
 2. Local swelling
 3. Mass at end of long bone
 4. Decreased range of motion
 - Differential Diagnosis
 1. Growing pains
 2. Trauma
 3. Osteomyelitis
 4. Tendonitis
 5. Septic arthritis
 6. Leukemia (due to bone pain)
 - Physical Findings
 1. Pain over involved site
 2. Palpable mass
 3. Swelling
 4. Decreased range of motion of affected extremity
 - Diagnostic Tests/Findings
 1. Radiograph of affected bone
 2. Computed tomography (CT) or magnetic resonance imaging (MRI) of affected bone
 3. Biopsy of area
 - Management/Treatment
 1. Multiagent chemotherapy
 2. Surgery—amputation/limb sparing surgery
 3. Patient/family education regarding side effects of therapy, anticipatory guidance on coping
 4. Primary care considerations—immunizations held while on therapy and resume 6 to 12 months after therapy completed (institutional specific policy); sibling immunizations not affected; fever on therapy requires CBC/blood cultures and hospitalization for neutropenia with fever or if patient has central line
 5. Long term follow-up for delayed effects of chemotherapy

■ IMMUNE DEFICIENCIES

- Definition: A group of immune defects that can be classified according to:
 1. Deficiency of immunoglobulin
 2. Deficiency of cell-mediated immunity
 3. Deficiency of both antibodies and cell-mediated immunity

Common Variable Immunodeficiency (CVID, “Acquired” Hypogammaglobulinemia)

- Definition: An immune disorder resulting from deficiency of all immunoglobulin (Ig)
- Etiology/Incidence: Genetic defect
- Signs and Symptoms (specific to manner of presentation): Recurrent or severe bacterial infections of respiratory tract (sinusitis, otitis, pneumonia) or skin (cellulitis, abscesses)
- Differential Diagnosis: Other immune deficiencies
- Physical Findings: Specific to manner of presentation
 1. Sinusitis—fever ($T \geq 101.5^{\circ}\text{F}$), periorbital edema, tenderness on percussion over sinuses
 2. Otitis—hyperemic, opaque, bulging tympanic membrane with poor mobility
 3. Pneumonia—retractions, flaring nares, diminished breath sounds, fine and crackling rales on affected side
 4. Cellulitis or abscess—erythema, swelling, purulent drainage
- Diagnostic Tests/Findings: Serum immunoglobulin
 1. IgG—decreased
 2. IgA—decreased
 3. IgM—decreased
- Management/Treatment: Intravenous immunoglobulin G (IgG) replacement (IVIg)

X-linked Agammaglobulinemia (Bruton’s Tyrosine Kinase or BTK)

- Definition: Absent or inadequate gammaglobulins caused by B-cell defect
- Etiology/Incidence
 1. Inherited, X-linked disorder of B lymphocytes
 2. Affected male infants usually asymptomatic for 3 to 6 months due to passive transmission of maternal antibodies, then show frequent infections and failure to thrive

3. Incidence rates unknown
- Signs and Symptoms: (specific to manner of presentation)—recurrent or severe bacterial infections of the respiratory tract (sinusitis, otitis, pneumonia) or skin (cellulitis, abscesses)
 - Differential Diagnosis: Other immune deficiencies
 - Physical Findings: Specific to manner of presentation
 1. Sinusitis—fever ($T \geq 101.5^\circ\text{F}$), periorbital edema, tenderness on percussion over sinuses
 2. Otitis—hyperemic, opaque, bulging tympanic membrane with poor mobility
 3. Pneumonia—retractions, flaring nares, diminished breath sounds, fine and crackling rales on affected side
 4. Cellulitis or abscess—erythema, swelling, purulent drainage
 - Diagnostic Tests/Findings
 1. Serum immunoglobulins
 - a. IgG—decreased
 - b. IgA—decreased
 - c. IgM—decreased
 2. Blood T and B lymphocytes—B cells decreased and T cells normal
 - Management/Treatment
 1. Intravenous immunoglobulin G (IgG) replacement (IVIG)
 2. Prophylactic antibiotics
 3. Antibiotic therapy for recurrent infections

Thymic Hypoplasia (DiGeorge Syndrome)

- Definition: A syndrome that includes T-cell dysfunction secondary to hypoplasia of the thymus
- Etiology/Incidence
 1. Dysmorphogenesis occurs during embryogenesis resulting in hypoplasia of the thymus and parathyroid glands; may also have cardiac defects, abnormal facies, cleft palate, and hypocalcemia
 2. Occurs in males and females equally
 3. Presentation usually results from cardiac failure or, after 24 to 48 hours of age, from hypocalcemia
 4. Sometimes diagnosed during cardiac surgery when no thymus is found in mediastinum
 5. Increased susceptibility to viral, fungal, and bacterial infections

- Signs and Symptoms: Specific to manner of presentation
- Differential Diagnosis: Other immune deficiencies
- Physical Findings
 1. Characteristic features—hypertelorism; shortened lip frenulum; lowset, notched pinnae; and nasal cleft
 2. Other physical findings—specific to manner of presentation
- Diagnostic Tests/Findings
 1. Routine chest radiograph may reveal absent thymus
 2. Blood T and B lymphocytes
 - a. Lymphopenia
 - b. Decreased T cells
 - c. B cells normal
- Management/Treatment
 1. Prophylactic antibiotics
 2. Antibiotic therapy for recurrent infections
 3. Thymus transplant if donor available
 4. Bone marrow transplant

Wiskott-Aldrich Syndrome (WAS)

- Definition: X-linked disorder characterized by thrombocytopenia, eczema, and progressive deterioration of thymus-dependent cellular immunity
- Etiology/Incidence
 1. X-linked recessive inheritance; carrier detection is possible
 2. Specific defect unknown
 3. Incidence—approximately 4 per one million male births
- Signs and Symptoms
 1. Neonatal—bloody diarrhea, prolonged bleeding from circumcision
 2. Later—eczema and frequent infections
- Differential Diagnosis: Usually made during neonatal period due to thrombocytopenia
 1. Other causes of chronic diarrhea
 2. Other immune deficiencies
- Physical Findings: Specific to manner of presentation
- Diagnostic Tests/Findings
 1. Serum immunoglobulins
 - a. IgG—normal
 - b. IgA—increased
 - c. IgM—decreased

2. Blood T and B lymphocytes
 - a. B cells—normal
 - b. T cells—decreased
 3. CBC—thrombocytopenia, small platelets
- Management/Treatment
 1. Platelet transfusions for bleeding
 2. IV immunoglobulins
 3. Prophylactic antibiotics and antibiotic therapy for recurrent infections
 4. Splenectomy—may increase platelet count
 - a. Conjugate and polysaccharide pneumococcal vaccines as recommended by the American Academy of Pediatrics; should be given at least 2 weeks or longer prior to surgery, if possible, to increase the likelihood of eliciting a protective antibody response
 - b. Prophylactic penicillin 250 mg bid for at least 1 to 2 years post-splenectomy
 - c. Increased risk for bacterial infections postsplenectomy—for fever ($T \geq 101.5^{\circ}\text{F}$) immediate referral for blood culture and empiric parenteral antibiotics against encapsulated organisms
 5. Bone marrow transplant if HLA-matched donor is available

■ QUESTIONS

Select the best answer

1. Baseline management of all neonates with ABO incompatibility includes:
 - a. Phototherapy
 - b. Serial monitoring of bilirubin and hemoglobin levels
 - c. Exchange transfusion
 - d. Simple transfusion of packed red blood cells
2. Which of the following is not associated with Rh incompatibility?
 - a. Mother Rh negative, baby Rh positive
 - b. Mother Rh positive, baby Rh negative
 - c. More severe in subsequent sensitized pregnancies
 - d. Hemolysis may occur up to 6 weeks or more
3. Clinical jaundice of the distal extremities would be noted at a bilirubin level of:
 - a. < 5 mg/dL
 - b. 5 mg/dL
 - c. 10 mg/dL
 - d. ≥ 15 mg/dL
4. β -chain synthesis is absent in:
 - a. β -thalassemia minor
 - b. β -thalassemia intermedia
 - c. β -thalassemia major
 - d. α -thalassemia trait
5. Which of the following are most often associated with hemoglobin C?
 - a. Growth retardation
 - b. Hepatosplenomegaly
 - c. Usually asymptomatic
 - d. Frontal bossing
6. Diagnostic findings consistent with β -thalassemia are:
 - a. Hemoglobin—normal
 - b. Reticulocytes—normal
 - c. $\text{Hgb A}_2 < 3.6$
 - d. Hypochromia, microcytosis
7. Asplenic children are at increased risk for which of the following?
 - a. Bacterial infections
 - b. Fungal infections
 - c. Viral infections
 - d. Parasites
8. Which of the following is not considered preventive management for iron deficiency anemia?
 - a. Iron fortified cereal from 6 to 12 months of age
 - b. Iron fortified formula until 6 months of age
 - c. No cow's milk until 1 year of age
 - d. If breastfeeding, supplemental iron drops or iron fortified cereal by 4 to 5 months of age
9. The expected clinical severity of hemoglobin sickle C disease (Hgb SC) is:
 - a. Asymptomatic
 - b. Marked to moderate
 - c. Mild to moderate
 - d. Severe
10. The expected hemoglobin range for sickle cell anemia is:
 - a. 6.5–9.5 g/dL
 - b. 13.5–16.5 g/dL
 - c. 8.5–12.5 g/dL
 - d. 9.5–13.5 g/dL
11. Prophylactic penicillin should be initiated in children with sickle cell anemia by:
 - a. 3 years of age
 - b. 12 months of age
 - c. 2 to 3 months of age
 - d. 9 months of age

12. Hemolysis does not contribute to which of the problems associated with sickle cell disease?
 - a. Chronic anemia
 - b. Splenic sequestration
 - c. Aplastic crisis
 - d. Delayed growth
13. The following blood lead level is not considered lead poisoning:
 - a. $< 10 \mu\text{g/dL}$
 - b. $10\text{--}14 \mu\text{g/dL}$
 - c. $\geq 15 \mu\text{g/dL}$
 - d. $> 25 \mu\text{g/dL}$
14. Which of the following is not a precipitating factor for hemolysis in G-6-PD deficiency?
 - a. Drugs
 - b. Exposure to extreme temperatures
 - c. Ingestion of fava beans
 - d. Infection
15. What percent of factor VIII/IX is associated with severe hemophilia A and B?
 - a. > 1
 - b. 1–5
 - c. 5–25
 - d. 30–50
16. What type of hemorrhage would be expected with severe factor VIII deficiency?
 - a. Severe hemorrhage following moderate to severe trauma
 - b. Gross bleeding following mild to moderate trauma
 - c. Gynecologic hemorrhage
 - d. Spontaneous hemarthrosis
17. Which of the following is the most common type of congenital bleeding disorder?
 - a. Hemophilia A
 - b. Hemophilia B
 - c. von Willebrand Disease
 - d. Idiopathic thrombocytopenia purpura
18. Which of the following medications should be avoided in a child with ITP?
 - a. Decongestants
 - b. Aspirin
 - c. Acetaminophen
 - d. Sulfa drugs
19. The following test is required to diagnose leukemia:
 - a. CBC with differential
 - b. Bone marrow aspiration/biopsy
 - c. Chest radiograph
 - d. Biopsy of an enlarged lymph node
20. Which of the following is not included as part of the initial therapy for ALL?
 - a. Chemotherapy
 - b. Radiation therapy
 - c. Bone marrow transplant
 - d. Intrathecal chemotherapy
21. Which malignancy is associated with genitourinary anomalies?
 - a. Acute lymphocytic leukemia
 - b. Chronic myelogenous leukemia
 - c. Osteosarcoma
 - d. Wilms tumor
22. Which of the following statements are true about immunizations during treatment of childhood cancer?
 - a. Children continue to receive immunizations as usual
 - b. Immunizations are not given during active chemotherapy
 - c. Only live vaccines are held during active chemotherapy
 - d. No family member should be immunized while the child is receiving chemotherapy
23. The peak incidence of osteosarcoma is:
 - a. 4–7 years of age
 - b. 8–11 years of age
 - c. 12–14 years of age
 - d. 15–19 years of age
24. The following type of infection is not associated with hypogammaglobulinemia:
 - a. Sinusitis
 - b. Pneumonia
 - c. Urinary tract infection
 - d. Cellulitis
25. The following diagnostic finding is consistent with X-linked agammaglobulinemia:
 - a. IgG—normal
 - b. B cells—decreased
 - c. T cells—decreased
 - d. IgA—normal
26. The following is not a characteristic feature of DiGeorge syndrome:
 - a. Hypertelorism
 - b. Cleft palate
 - c. Cardiac defects
 - d. Frontal bossing

27. The following diagnostic finding is consistent with Wiskott-Aldrich syndrome:
- IgG—normal
 - IgA—decreased
 - IgM—increased
 - B cells—decreased
28. Management of a patient with a splenectomy does not include:
- Pneumococcal vaccines at least 2 weeks prior to surgery
 - Prophylactic penicillin
 - Blood culture and parenteral antibiotics for febrile illnesses
 - Treating fever with antipyretics only and observing for resolution

■ ANSWERS

- | | |
|--------------|--------------|
| 1. b | 15. a |
| 2. b | 16. d |
| 3. d | 17. c |
| 4. c | 18. b |
| 5. c | 19. b |
| 6. d | 20. c |
| 7. a | 21. d |
| 8. b | 22. b |
| 9. c | 23. d |
| 10. a | 24. c |
| 11. c | 25. b |
| 12. b | 26. d |
| 13. a | 27. a |
| 14. b | 28. d |

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Endocrine Disorders

Malinda Duke

■ THYROID DISORDERS

Hypothyroidism

- Definition
 1. Congenital vs. Acquired (based on timing of disorder)
 - a. Congenital can cause severe mental retardation unless treated early; newborn screening can identify hypothyroid infants by 3 weeks of age, allowing early treatment to avoid the effects of severe neurological devastation
 - b. Acquired may have onset within first year of life, usually onset occurs in childhood or adolescence
 2. Primary, secondary, tertiary (based on site of disorder)
 - a. Primary disease or disorder of thyroid gland (thyroid gland failure)
 - b. Secondary disease or disorder of the pituitary gland that compromises thyroid gland function
 - c. Tertiary disease or disorder of the hypothalamus compromises thyroid gland function
- Etiology/Incidence (most common acquired endocrinopathy)
 1. Congenital hypothyroidism
 - a. Absence (athyreosis), underdevelopment (dysgenesis), of ectopic gland most common
 - b. Inherent dysfunction in transport or assimilation of iodine, or in synthesis or metabolism of thyroid hormone (e.g., thyroid enzyme defects, familial dysmorphonogenesis)
 - c. Maternal disease adversely affecting fetal thyroid development and function (pre-natal exposure to iodine-containing or goitrogenic drugs and agents, e.g., thiouracil, methimazole, iodines; maternal exposure to radioactive iodine; placental crossing of maternal antibodies to fetal thyroid gland)
 - d. Iodine deficiency causing endemic goiter and cretinism
 - e. Hypothalamic or pituitary disorder (e.g., pituitary agenesis, anencephaly)
 - f. Affects 1 infant in every 4000 live births, female to male ratio 2:1
 - g. Higher incidence in Hispanic and Native American infants
 - h. Higher incidence in areas with endemic iodine deficiency
 - i. Higher incidence in women, affecting 2% of women and 0.2% men
 2. Acquired hypothyroidism
 - a. Chronic lymphocytic thyroiditis (Hashimoto's, autoimmune) most common cause beyond perinatal period; often with positive family history; two types: goitrous (more common) and atrophic
 - b. Late manifestation of congenital absence, underdevelopment (dysgenesis), or atrophy of thyroid gland

- c. Late manifestation of congenital defects in synthesizing or metabolizing thyroid hormone
 - d. Ablation of thyroid through medical procedures (e.g., surgery, irradiation, radioactive iodine)
 - e. Exposure to iodine-containing drugs and agents; drug-induced (e.g., antithyroid drugs, excessive iodide, lithium, cobalt); exposure to naturally occurring goitrogens in foods, water pollutants
 - f. Disease of hypothalamus or pituitary (e.g., pituitary tumors, trauma), rare; child will show other signs of hypothalamic or pituitary disease
 - g. Endemic goiter from nutritional iodide deficiency—most common thyroid disease worldwide
3. Hypothyroidism, depending on cause, may be permanent or transient disorder
 4. Severity, compensated or uncompensated hypothyroidism reflects the ability to maintain normal T_4
- Signs and Symptoms
 1. Affects multiple systems; many nonspecific, insidious signs and symptoms; severity depends on age of onset and degree of thyroid deficiency; symptoms vary for infants vs. children
 2. May be familial history of thyroid and pituitary diseases; may have maternal prenatal history of thyroid disease or ingestion of antithyroid medications or foods
 3. May be associated with other autoimmune disease or syndromes (e.g., Down, Turner's)
 4. Neonates/infants
 - a. Infants may have no obvious symptoms during first month of life
 - b. History of lethargy, poor feeding, prolonged elevated bilirubin (>10 mg/dL >3 days of age)
 - c. May be postmature; increased birth weight (>4000 g)
 5. Older infants, children, adolescents
 - a. History of poor growth, intolerance to cold, poor appetite, constipation
 - b. Mental and physical sluggishness, developmental delay
 - Differential Diagnosis
 1. Differentiate primary hypothyroidism due to intrinsic thyroid gland defects from secondary thyroid deficiency caused by pituitary or hypothalamic disorders
 2. Congenital thyroxine-binding globulin (TBG) deficiency is a normal variant of thyroid physiology; TBG deficiency is defined as low T_4 with normal TSH and Free T_4 ; both the TBG is low, T_3 uptake is elevated
3. "Euthyroid sick syndrome" seen in small or sick newborns or in children with acute or chronic severe illnesses, surgery, trauma, or malnutrition; commonly have low T_3 ; normal, low, or free T_4 ; and normal TSH
- Physical Findings
 1. Affects multiple systems; severity of findings depends on age of onset and degree of thyroid deficiency; no findings are entirely sensitive or specific
 2. Neonates/infants
 - a. Prolonged jaundice; poor feeding
 - b. Growth deceleration
 - c. Hypothermia, skin mottling
 - d. Large fontanelles, especially posterior; wide sutures, hirsute forehead, coarse facial features, dull expression, facial edema, nasal discharge, macroglossia
 - e. Normal, slightly enlarged or goitrous thyroid gland; if thyroid ectopic, may see mass at base of tongue or in midline of neck
 - f. Hoarse cry
 - g. Axillary, prominent supraclavicular fat pads
 - h. Respiratory distress in term infant
 - i. Bradycardia (<100 beats/min)
 - j. Distended or protuberant abdomen, umbilical hernia, constipation
 - k. Lumbar lordosis, hypotonia
 3. Infants, children, and adolescents
 - a. Low energy level, increased weight for height, myxedema with hypothyroidism of long duration
 - b. Linear growth retardation or growth deceleration; delayed bone maturation, dentition, tooth eruption
 - c. Decreased concentration, memory impaired, developmental delay, poor motor coordination, dull appearance
 - d. Delayed puberty, occasionally precocious puberty or pseudoprecocity; menstrual disorders
 - e. Skin cool, dry, pale, gray, mottled, thickened, increased pigmentation, carotenemia
 - f. Hair dry, brittle; lateral thinning of eyebrows
 - g. Possible enlarged thyroid gland (goiter); may feel cobblestonelike
 - h. Galactorrhea, constipation
 - i. Myopathy, muscular hypertrophy, poor muscle tone; prolonged relaxation phase of deep tendon reflexes (DTR)

- j. Delayed bone age
 - k. If advanced hypothyroidism and myxedema, may be “chubby” and have periorbital edema
- Diagnostic Tests/Findings
 1. Newborn screening for congenital hypothyroidism is routine in all 50 states
 - a. Abnormal newborn screen—repeat thyroid screen for confirmatory diagnosis with thyroid stimulating hormone (TSH) and thyroxine (T_4)
 - b. Elevated serum TSH and low T_4 diagnostic of transient or permanent primary hypothyroidism
 - c. Positive TSH receptor-blocking antibodies (TRBAbs)—diagnostic for transient congenital hypothyroidism (maternal antibodies)
 - d. Additional thyroid function tests—serum thyroxine-binding globulin (TBG); triiodothyronine resin uptake (T_3 RU)
 2. Acquired hypothyroidism secondary to pituitary or hypothalamic disorders
 - a. Low free (unbound) thyroxine (free T_4), usually normal TSH; and low thyroxine-binding globulin (TBG)
 - b. Abnormal pituitary function tests
 3. Euthyroid sick syndrome (nonthyroidal illness syndrome) secondary to coexisting acute or chronic illness; low T_4 level with normal TBG; low T_3 ; normal TSH; free T_4 and reverse T_3 levels high, normal, or elevated
 4. Autoimmune (Hashimoto’s) thyroiditis (with goiter present)—elevated serum thyroid peroxidase or thyroglobulin antibody titers; autoantibodies are present in 95% of cases; begin treatment therapy when TSH rises above normal and T_4 /free T_4 is low or normal
 5. Repeat thyroid function tests if clinical suspicion of hypothyroidism, history of thyroid disease in pregnancy, positive history of thyroid dyshormonogenesis
 6. May have abnormal thyroid scan, ultrasound, imaging, or bone age results
 - Management/Treatment
 1. Consultation or referral to pediatric endocrinologist for suspected or confirmed hypothyroidism
 2. For congenital hypothyroidism, rapid and adequate thyroid hormone replacement critical to avoid irreparable neurologic impairment and mental retardation; treat within first month of life for best prognosis for optimal intellectual development
 3. Drug of choice daily oral levothyroxine (L-thyroxine); after treatment initiated for hypothyroidism in infancy, monitor T_4 and TSH levels monthly during first 3 months of age, every 2 to 3 months for the remainder of the first year, every 3 to 4 months during second and third year, and every 4 to 12 months thereafter until growth complete; more frequent checks if compliance issues, changes in physical symptoms (especially poor growth), or abnormal values; monitor additionally as needed whenever dose adjusted
 4. Recommended dosages of levothyroxine (T_4) vary by age; dosage/kg/day decreases over time with age
 5. In older children and adolescents, increase thyroid dose gradually to full replacement dose to avoid undesirable side effects and clinical symptoms of thyrotoxicosis (e.g., headaches, abrupt personality changes)
 6. Once older children and adolescents in euthyroid state, monitor adequacy of levothyroxine therapy with regular, periodic T_4 and TSH levels
 7. Give levothyroxine an hour before meals to prevent reduced absorption, especially for infants; iron may interfere with absorption
 8. Educate parents and child about disease, treatment regimen, and routine monitoring; with thyroid replacement, child will be livelier, less docile, may lose water weight, hair of normal texture will replace dry hair, may have transient stomach or headaches
 9. Genetic counseling may be indicated if familial etiology
 10. Trial off of medication may be indicated at age 3, if possibility child had transient congenital hypothyroidism

Hyperthyroidism

- Definition: Excessive production and secretion of thyroid hormone (TH) by thyroid gland (thyrotoxicosis) resulting in increased basal metabolism, goiter, autonomic nervous system disorders, and problems with creatinine metabolism
- Etiology/Incidence
 1. Caused by excess production of thyroid hormone (e.g., Graves’ or autoimmune hyperthyroidism, pituitary tumor) or excess release of thyroid hormone (e.g., subacute thyroiditis, Hashimoto’s toxic thyroiditis, iodine-induced hyperthyroidism), or hot nodules
 2. Most common cause is autoimmune response (Graves’)—body produces thyroid stimulating

immunoglobulins (TSI) which stimulate TSH receptors in thyroid gland, causing overproduction of TH and thyroid hypertrophy; has genetic predisposition

3. If mother is thyrotoxic prenatally or has history of Graves', infants may have transient congenital hyperthyroidism (neonatal Graves') since thyroid-stimulating immunoglobulins (TSI) cross placenta to fetus; rare
 4. Graves' disease is rare, 0.8 per 100,000 children per year and six times more common in girls, usually in second decade and commonly associated in children with other autoimmune disorders
- Signs and Symptoms
 1. May have family history of thyroid disorder (e.g., Graves') or maternal history of antithyroid drug ingestion for treatment of Graves' during pregnancy
 2. Neonates/infants
 - a. Very rare in neonate; signs and symptoms usually present shortly after birth or may present days or weeks later; not common, but can have severe consequences if untreated; diagnosis rarely made in newborn period
 - b. Prematurity, low birthweight, poor weight gain, poor feeding
 - c. Restlessness, irritability, tachycardia
 - d. Can be neonatal Graves' disease, due to transplacental passage of stimulatory maternal antibodies, or autosomal dominant hyperthyroidism requiring antithyroid treatment prior to thyroidectomy
 3. Child/adolescent
 - a. Weight loss, although increased appetite; may have accelerated growth and advanced bone age with long-term illness
 - b. Nervousness, irritability, decreased attention span, behavior problems, decline in school performance, emotional lability, restlessness, fatigue, weakness, heat intolerance, increased perspiration
 - c. Sleeplessness or sleep restlessness, insomnia, nightmares
 - d. Visual disturbances, e.g., increased lacrimation, diplopia, photophobia, blurring
 - e. Palpitations
 - f. Frequent urination and loose stooling, may have enuresis
 - g. Amenorrhea
 - Differential Diagnosis
 1. Neonates—systemic illness, sepsis, and narcotic withdrawal
 2. Children and adolescents

- a. Nodular thyroid disease
- b. Thyroid cancer
- c. Euthyroid goiter
- d. Chronic disease, e.g., pituitary disease, severe anemia, leukemia
- e. Thyroiditis
- f. Accidental or deliberate excessive thyroid hormone or iodine ingestion
- g. Chorea
- h. Psychiatric illness, e.g., anxiety disorder, anorexia

- Physical Findings

1. Neonates and infants
 - a. May be small for gestational age
 - b. Lid retraction, proptosis, periorbital edema
 - c. Face may be flushed
 - d. Enlarged thyroid (goiter)
 - e. Tachycardia without underlying heart disease
 - f. Increased gastrointestinal motility
 - g. Severely affected neonates may have jaundice, microcephaly, frontal bossing, craniosynostosis, ophthalmopathy, exophthalmia, thrombocytopenia, cardiac problems, hepatosplenomegaly, other signs of severe illness
2. Children and adolescents
 - a. Increased energy (with periods of lethargy), decreased sleep, decreased school performance and concentration, personality changes: irritability and emotional lability
 - b. Eye findings, e.g., proptosis, exophthalmos, upper lid lag with downward gaze, lid retraction, stare appearance, periorbital and conjunctival edema
 - c. Variable-size enlarged, tender or nontender, spongy or firm thyroid with palpable border; may have thyroid bruit or thrill
 - d. Tachycardia, systolic hypertension, increased pulse pressure, palpitations
 - e. Proximal muscle weakness, diminished fine motor control, tremor, short DTR relaxation phase
 - f. Advanced skeletal maturation radiographically
 - g. Warm, moist, smooth, diaphoretic skin; face may be flushed; heat intolerance; increased bowel movements

- Diagnostic Tests/Findings

1. If signs or symptoms of thyrotoxicosis or enlarged thyroid, do confirmatory laboratory thyroid function tests

2. Thyroiditis indicated by elevated T_4 , free T_4 , T_3 resin uptake (T_3 RU) with TSH suppression and low serum cholesterol
 3. Circulating thyroid stimulating immunoglobulin (TSI) and other thyroid antibody tests, including thyrotropin receptor antibody (TRAb) titers, are elevated and in most cases thyroid peroxidase (TPO) antibodies are also positive
 4. Although rare in childhood, a child developing acute onset of hyperthermia, severe tachycardia, and restlessness needs evaluation for thyroid crisis or storm
 5. Graves' Disease—elevated T_4 and low TSH; advanced bone age
 6. Radioactive iodine uptake scan shows increased uptake if excess TH production; if increased release of TH only, will have decreased radioactive iodine uptake
 7. High plasma T_4 does not necessarily indicate hyperthyroidism as there is a physiological transient peak of plasma T_4 on day two of life; diagnosis of true hyperthyroidism requires low or undetectable TSH with high T_4 and/or T_3
- Management/Treatment
 1. Consultation or referral to pediatric endocrinologist for suspected or confirmed hyperthyroidism
 2. Treatment dictated by identified etiology for hyperthyroidism and degree of thyrotoxicity; attempt medical management as primary initial therapy
 3. Prompt diagnosis and treatment especially important in neonates as condition may be life-threatening
 4. Treatment goal is prompt return to euthyroidism with use of:
 - a. Antithyroid drugs to inhibit thyroxine (T_4) synthesis and conversion of T_3 to T_4 , e.g., propylthiouracil, methimazole
 - b. Beta-adrenergic receptor blockers to control nervousness and cardiovascular symptoms, e.g., propranolol, atenolol (NOT in asthmatics)
 - c. Side effects of antithyroid medications include rashes, urticaria, arthralgias, and decreased white blood cell count; all children on treatment require a CBC for fever or sore throat
 - d. Ablative therapy with radio-iodine permanently suppresses thyroid function; hypothyroidism induced, but no side effects of medication
 - e. Thyroid surgery no longer considered primary form of treatment except in pregnancy under certain circumstances

5. Restricted physical activity if hyperthyroidism severe or in preparation for surgery
6. Educate parent and child about disease, duration of treatment, side effects of medications or complications if surgery required, adherence to treatment regimen, and, if Graves' disease, need for lifelong monitoring; advise of complications, e.g., thyrotoxicosis
7. Genetic counseling may be indicated if familial etiology

Thyroiditis

- Definition: Inflammation of thyroid gland caused by autoimmune response to the thyroid gland (chronic lymphocytic thyroiditis [Hashimoto's]); infectious agents (acute suppurative and subacute nonsuppurative); or from exposure to radiation or trauma; occasionally idiopathic
- Etiology/Incidence
 1. Acute suppurative thyroiditis with bacterial etiology—e.g., group A streptococci, pneumococci, *S. aureus*, and anaerobes; rare
 2. Subacute nonsuppurative thyroiditis caused by viruses—e.g., mumps, influenza, echovirus, coxsackie, Epstein-Barr, adenovirus; rare in U.S.
 3. Chronic lymphocytic thyroiditis (chronic autoimmune or Hashimoto thyroiditis)—noninfectious autoimmune inflammatory disease of thyroid most common cause of goiter and hypothyroidism in childhood; highest incidence in children 8 to 15 years; more common in females than males (4:1); increasing incidence may be associated with rising incidence of type 1 diabetes
- Signs and Symptoms
 1. With infectious thyroiditis, may have recent history of or concurrent upper respiratory illness
 2. With chronic autoimmune thyroiditis, may have family history of autoimmune thyroid disease
 3. Onset in acute thyroiditis rapid; insidious onset in subacute and chronic lymphocytic thyroiditis
 4. Fever, malaise; may feel quite ill with acute suppurative or subacute thyroiditis, particularly with former
 5. With acute and subacute thyroiditis, pain and tenderness of thyroid with radiation to other areas of neck, ear, chest; with acute suppurative thyroiditis, severe pain with neck extension; no tenderness with chronic lymphocytic thyroiditis

6. Complaints of unilateral or bilateral swelling of the thyroid, complaints of fullness in anterior neck; sensation of tracheal compression
 7. May have sore throat, hoarseness, dysphagia
 8. May have nervousness, irritability, other symptoms of mild hyperthyroidism
- Differential Diagnosis
 1. Infectious toxic thyroiditis must be distinguished from chronic lymphocytic autoimmune thyroiditis; simple goiters due to inborn errors of T_4 synthesis; and chronic autoimmune thyroiditis (Hashimoto's) associated with adolescence in girls; and thyroglossal duct abscesses
 2. Goiters induced by drugs—(e.g., from iodine, lithium, para-amino salicylic acid, thionamides, white turnip plants); by transplacental exposure to TH by breastfeeding of newborns when mothers on antithyroid therapy
 3. Cancerous or cystic thyroid nodules
 - a. Benign causes include multinodular goiter, cysts, follicular adenomas, or thyroiditis
 - b. Malignant causes include papillary, follicular, Hurthle cell, medullary or anaplastic carcinomas or metastatic lesions
 - Physical Findings
 1. Findings variable depending on etiology—i.e., infectious vs. autoimmune
 2. May be toxic-appearing if infectious etiology, but not necessarily thyrotoxic
 3. In infectious thyroiditis—thyroid gland unilaterally or bilaterally enlarged, tender, firm
 4. In chronic lymphocytic thyroiditis, usually symmetric, nontender, firm, freely movable, diffusely enlarged “pebbly” goiter
 - Diagnostic Tests/Findings
 1. Laboratory findings variable depending on etiology
 2. In acute suppurative thyroiditis—usually no consistently associated endocrine disturbances, moderate to marked shift to left in differential, elevated sedimentation rate
 3. Subacute nonsuppurative thyroiditis—radioiodine uptake usually reduced; serum TH levels normal or elevated
 4. In chronic lymphocytic thyroiditis—usually normal T_4 , free T_4 , T_3 RU, but may be elevated or depressed; thyroid autoantibodies usually present, abnormal thyroid scan, surgical or needle biopsy diagnostic but rarely indicated

- Management/Treatment
 1. Physician consultation or referral to pediatric endocrinologist for suspected or confirmed thyroiditis
 2. Specific antibiotic therapy required for acute suppurative thyroiditis and may be needed for subacute nonsuppurative thyroiditis since latter is difficult to distinguish from former; acetylsalicylic acid or other anti-inflammatory drugs
 3. Treatment for autoimmune chronic lymphocytic thyroiditis controversial; levothyroxine may be used to decrease goiter but efficacy in preventing progression of hypothyroidism in long term not supported
 4. Adolescents with autoimmune chronic lymphocytic thyroiditis need lifelong monitoring since development of hypothyroidism is possible, subacute thyroiditis self-limiting
 5. All children with type 1 diabetes should be annually screened for thyroid dysfunction; increased risk of thyroid microsomal antibody
 6. Genetic counseling may be indicated for familial etiology

▣ PITUITARY DISORDER—DIABETES INSIPIDUS (DI)

- Definition: Excretion of large amounts of dilute urine due to insufficient function of antidiuretic hormone (ADH, arginine vasopressin, disorder in ADH receptors in kidney, primary renal disease, or, rarely in children, to primary polydipsia; compromised ability to concentrate urine)
- Etiology/Incidence
 1. Central (hypothalamic, neurogenic, vasopressin-sensitive) DI caused by hypofunction of hypothalamus/posterior pituitary or increased vasopressin metabolism resulting in ADH deficiency
 - a. Genetic, familial autosomal dominant trait, rare
 - b. Congenital, e.g., anatomic defects in brain
 - c. Acquired secondary to accidental or surgical trauma, infection, cerebral anoxia, neoplasm, or infectious disease; trauma following removal of hypothalamic area tumors major cause
 - d. Secondary to autoimmune or infiltrative disease, e.g., histiocytosis, lymphocytic hypophysitis
 - e. Idiopathic
 - f. Others: brain death, increased vasopressin metabolism, drugs, e.g., ethanol

2. Nephrogenic (vasopressin-resistant) DI caused by reduced renal responsiveness to ADH (not CNS mediated)
 - a. Familial—X-linked inherited disorder of ADH receptor sites in kidney in males primarily; females rarely affected, less common etiology but more severe
 - b. Acquired
 - (1) Renal failure, e.g., obstructive uropathy, polycystic kidney disease
 - (2) Electrolyte disorders (hypercalcemia, hypokalemia)
 - (3) Nephrotoxic drugs, e.g., lithium, demeclocycline, amphotericin, methicillin, rifampin
 - (4) Other illness, e.g., sickle cell
- Signs and Symptoms
 1. Symptoms vary depending on etiology, age, anterior pituitary function, preservation of normal thirst, diet
 2. Central (neurogenic) DI
 - a. May have family history of congenital ADH deficiency
 - b. Generally rapid onset; disease may be masked as failure to thrive
 - c. History of poor weight gain, deficient growth if long duration
 - d. Unexplained fever, irritability
 - e. Intense thirst, polydipsia, desire for cold drinks, preference for cold water; irritable when fluid withheld; unable to sleep through night without water intake
 - f. Vomiting, constipation; unexplained fever in infants
 - g. Tendency to avoid diets high in protein and salt
 - h. Polyuria, nocturia, enuresis in previously toilet-trained child; clear urine; unable to concentrate urine after fluid restriction
 - i. May have symptoms of intracranial tumor (headaches, strabismus, double vision, vomiting, precocious puberty)
 - j. May follow intracranial surgical procedures or trauma; CNS disease; rarely idiopathic
 3. Nephrogenic DI
 - a. Genetic or acquired causes—may have family history of congenital nephrogenic DI or maternal history of polyhydramnios; infants may do well on breastfeeding until weaning (breastmilk has low renal solute load), infection, or introduction of solids, then may fail to thrive; often present with fever, vomiting, dehydration
 - b. Female carriers of trait have varying severity of disease
 - c. Poor weight gain, deficient growth if long duration; may be malnourished
 - d. Increased thirst, polydipsia, history of large water intake, poor food intake because of preference for water over milk or solids
 - e. Dehydration, absence of tears, perspiration, if dehydration severe, may have seizures
 - f. Irritability; may have poor attention span, poor school performance
 - g. Vomiting, polyuria, nocturia
 - Differential Diagnosis
 1. Distinguish DI caused by suppression of vasopressin secretion (congenital vs. acquired central DI) from DI caused by reduced renal responsiveness to arginine vasopressin (congenital vs. acquired nephrogenic DI)
 2. Psychogenic polydipsia (compulsive water drinking) and other causes of polyuria (e.g., drug-induced polydipsia [e.g., thioridazine, tricyclics]; hypokalemia; hypercalcemia [including hypervitaminosis D]; primary and secondary renal disease; diabetes mellitus)
 - Physical Findings
 1. Central (neurogenic) DI
 - a. Variable levels of dehydration; if disease unrecognized, infants may have high fever, vomiting, seizures, circulatory collapse
 - b. Poor weight gain, deficient growth if long duration, may be malnourished
 - c. Irritability, may have poor attention span
 - d. May have symptoms of brain tumor (e.g., strabismus, nystagmus)
 2. Nephrogenic DI
 - a. Variable levels of dehydration; dry skin, no tears, no perspiration; if severe, infants may have high fever, convulsions, circulatory collapse
 - b. Failure to thrive, malnourished; if long duration, may have growth retardation, delayed sexual maturation, CNS damage
 - c. Fever, irritability
 - d. Large or distended bladder except immediately after voiding; nonobstructive hydronephrosis, hydroureter
 - Diagnostic Tests/Findings
 1. History of pathological polydipsia and polyuria ($> 2 \text{ L/m}^2/\text{day}$) in children
 2. Urine specific gravity (< 1.005 ; osmolality $< 280 \text{ mOsm/kg}$)
 3. Inability to concentrate urine after fluid restriction
 4. Hyperosmolality of plasma

5. Plasma arginine-vasopressin (AVP) measured—low level indicates complete central diabetes insipidus; nephrogenic DI shows inappropriately high AVP without rise in urine osmolality
 6. Positive water deprivation test—to establish diagnosis of DI and differentiate central vs. nephrogenic DI
 - a. Serum osmolality > 300 mOsm/kg and urine osmolality < 300 mOsm/kg diagnostic of DI
 - b. ADH administration (vasopressin) differentiates central vs. nephrogenic DI
 - (1) Central DI—decreased urine volume and increased urine osmolality
 - (2) Nephrogenic DI—no further changes in urine volume or osmolality
 7. With MRI, absence or diminished “bright spot” in posterior pituitary; diminished blood flow to posterior pituitary; classic finding for a mass of the optic chiasm—bitemporal hemianopsia
 8. Low serum ADH concentration in central DI; high serum ADH level in nephrogenic DI
- Management/Treatment
 1. Immediate physician consultation or referral to pediatric endocrinologist for suspected or confirmed DI
 2. Careful adequate rehydration to avoid seizures, intellectual impairment due to CNS damage from hyponatremia and dehydration, particularly in infants who may not be able to make known their need for water or to obtain water necessary to quench thirst; early recognition and management needed to avoid sequelae of stunted growth and cognitive impairment
 3. Treat treatable underlying causes of nephrogenic DI, e.g., hypokalemia, drug-induced obstructive uropathy
 4. Treatment of choice for diabetes insipidus—intranasal desmopressin acetate (DDAVP); in infants, sometimes given extra free water rather than DDAVP for normal hydration and prevention of unwanted hyponatremia; with nephrogenic DI, may need diuretics, e.g., thiazides, prostaglandin synthesis inhibitors, e.g., indomethacin
 5. Breastfeeding preferable; adequate calories for growth; with nephrogenic DI, water allowed as demanded, sodium intake restricted; teach regarding avoidance of severe dehydration
 6. Genetic counseling may be indicated for familial etiology
 7. Treat other identified causes, e.g., germinomas, histiocytosis, craniopharyngiomas

■ GROWTH DISTURBANCES

Short Stature

- Definition
 1. Variation from average pattern of growth; associated with normal variants of growth and pathologic conditions
 2. Growth adequacy determined by consideration of both growth rate and absolute height; severe form of short stature defined as height > 3 standard deviations below the mean
 3. CDC growth chart should be used to evaluate growth of children, see www.cdc.gov/growthcharts
- Etiology/Incidence
 1. Multiple etiologies—genetic, constitutional, physiologic, environmental, psychosocial
 - a. Normal growth variations (most common)
 - (1) Familial or genetic normal variant of average growth pattern which is familial, racial, or genetic; child “constitutionally small” and remains small as adult
 - (2) Constitutional delay of growth with delayed growth pattern resulting in delayed physical maturity but normal final adult height
 - b. Pathologic causes of short stature
 - (1) Nutritional—hypocaloric, gastrointestinal disorders
 - (2) Endocrine—growth hormone (GH) deficiency, occurs in 1:4000 children
 - (a) Hereditary—rare gene deletion
 - (b) Idiopathic—about 66% cases caused by suspected deficiency or impairment in hypothalamic secretion of human growth hormone releasing (hGH) hormone
 - (c) Acquired—secondary to pituitary/hypothalamic disease, CNS infection, trauma, intracranial tumors
 - (3) Other endocrine disturbances—hypothyroidism, hypopituitarism, excess cortisol, precocious puberty
 - (4) Chromosomal defects (Turner’s, Noonan, Down syndrome)
 - (5) Intrauterine growth retardation (IUGR)—sporadic or associated with syndromes, e.g., de Lange syndrome, Russell-Silver syndrome; infections; placental abnormalities, maternal abnormalities, e.g., drug use, malnutrition

- (6) Bone development disorders—achondroplasia, skeletal disorders
 - (7) Metabolic—storage diseases, inborn errors of metabolism
 - (8) Systemic and chronic diseases and congenital defects—e.g., liver, hematologic, respiratory, cardiovascular, renal, CNS disease, or tumors
 - (9) Associated with chronic systemic diseases, birth defects, mental retardation, cancers
 - (10) Psychosocial factors, e.g., deprivation dwarfism
 - (11) Chronic drug intake, e.g., glucocorticoids
- Signs and Symptoms
 1. Normal growth variations
 - a. Familial short stature—usually small at birth ($\leq 3\%$) but consistent with family pattern
 - b. Constitutional growth delay—usually normal size at birth with declining height and weight to $< 5\%$ between 1 and 3 years of age; delayed pubescence; family history of similar growth pattern in parents or other family members
 2. Pathologic growth variations
 - a. History of poor nutritional intake, malabsorption syndromes
 - b. Symptoms of GH deficiency—failure to grow, headaches, delayed dental development, visual field defects, polyuria, polydipsia, delayed sexual maturation, CNS abnormalities, history of trauma, infection, radiation to CNS
 - c. Signs and symptoms of other endocrine disorders—fever, lethargy, irritability, developmental delay, dull appearance, FTT, increased weight for height, polyuria, polydipsia, constipation, delayed sexual maturation, CNS symptoms, CNS surgery
 - d. Intrauterine growth retardation (IUGR) and low birthweight (LBW); normal BW or normal growth pattern with subsequent onset of decelerated or delayed growth; history of premature aging
 - e. Symptoms of other systemic or chronic illness—FTT, congenital defects including intrinsic diseases of bone
 - f. Dysmorphism at birth, chromosomal abnormalities, syndromes, congenital skeletal defects or anomalies, e.g., abnormal upper to lower body ratio, abnormal or disproportionate features
 - g. Signs and symptoms of neglect, emotional maltreatment; abnormalities in psychosocial development; parents may be overwhelmed or disorganized and not intentionally neglectful or abusive
 - h. Chronic drug intake, e.g., glucocorticoids, high doses of estrogens, androgens
- Differential Diagnosis: Distinguish normal variants of familial short stature and constitutional growth delay from pathologic causes
 - Physical Findings
 1. Familial or constitutional short stature—height, weight, occipital-frontal circumference (OFC) growth curve patterns generally consistent, symmetric
 - a. Familial—growth chart showing $BW \leq 3\%$ but consistent with family pattern; follows growth curve; normal physical examination; radiographic bone age consistent with chronological age
 - b. Constitutional delay—growth chart showing normal size at birth with declining height and weight throughout 1 to 3 years to $< 5\%$; normal physical examination; bone maturation 2 to 3 years behind chronological age
 2. Pathologic short stature
 - a. GH deficiency—BW may be normal, birth length 50% that of normal child; height and weight growth deficits; infantile fat distribution; youthful facial features; midfacial hypoplasia; visual field defects; small hands and feet; newborn may have microphallus (stretched penile length of < 2.5 cm vs. normal mean length of 4 cm); may have CNS findings
 - b. Primordial short stature
 - (1) IUGR—BW and birth length below normal for gestational age, OFC normal or < 3 rd percentile; subsequent growth parallel to or < 3 rd percentile
 - (2) Primordial dwarfism with premature aging—child appears older than age
 - (3) Short stature with and without dysmorphism—height, weight < 3 rd percentile, may have normal physical examination other than small size or may have various abnormal physical findings, e.g., microcephaly
 - c. Short stature associated with chromosomal abnormalities—may have dysmorphism or stigmata of specific congenital or familial disorders, e.g., Turner's syndrome (webbed neck, small jaw, prominent ears, epicanthal folds, low posterior hairline, broad chest, cardiac defects) or Down syndrome

- d. Short stature associated with bone or cartilage development disorders, e.g., skeletal dysplasia, short extremities with normal size head and trunk, frontal bossing, abnormal upper to lower body ratios, abnormal or disproportionate features, rickets, leg bowing
 - e. Short stature associated with symptoms of endogenous cortisol excess, e.g., Cushing's disease with moon facies, hirsutism, "buffalo hump," striae, hypertension, fatigue, voice deepening, obesity, amenorrhea
 - f. Chronic drug intake, e.g., glucocorticoid excess with hypertension, plethora, moon facies, purple striae, interscapular fat pad, truncal obesity, muscle wasting
 - g. Abnormalities in psychosocial development; neglectful parents
- Diagnostic Tests/Findings
 1. Abnormalities in previous sequential, consistent recordings of height, weight, and OFC plotted on age-standardized growth charts
 2. In familial short stature and constitutional delay, height may be < 3rd percentile but growth rate normal; careful family history of familial growth patterns may elucidate familial vs. constitutional delay as etiology of short stature
 3. In growth failure, slower than normal growth rate results in flattened growth curve or decrease in growth parameter percentiles
 4. May have abnormal complete and segmental growth measurements and upper to lower body ratio measurements
 5. Laboratory tests to confirm diagnosis based on clinical findings and to rule out systemic disease or hormonal deficiency; findings depend on etiology
 - a. Abnormal CBC—chronic anemia, infection, leukemia
 - b. Elevated sedimentation rate—collagen vascular disease, cancer, chronic infection
 - c. Abnormal biochemical profiles—adrenal insufficiency, renal disease
 - d. Abnormal stool examination—inflammatory bowel disease, severe parasitism
 - e. Abnormal thyroid function studies—hypothyroidism
 - f. Low serum human growth hormone (hGH), insulin growth factor (IGF-1), IGF-binding protein—growth hormone deficiency (GHD)
 - g. Abnormal urinalysis—renal disease
 6. Delayed maturity on radiographic bone age indicates constitutional delay (generally 2 to 3 years behind chronological age), GH

- deficiency, hypothyroidism, severe systemic illness; normal bone age found in familial short stature
- 7. Nutritional evaluation may show inadequate calories
- 8. Abnormal home/social evaluation may suggest psychosocial etiology
- 9. Abnormalities on skull radiograph, CT, MRI of cranium if intracranial lesion
- 10. Karyotype analysis in short girls with pubertal delay may indicate Turner's syndrome

- Management/Treatment

1. Physician consultation or referral to appropriate pediatric specialists for children with other than familial or constitutional growth delay
2. If familial or constitutional growth delay, only periodic monitoring of growth pattern needed; reassurance
3. If marked psychosocial problems arise in boys because of pubertal delay, short-term testosterone may initiate sexual development
4. Optimize treatment for other endocrine or systemic or chronic illnesses to minimize compromised growth; adequate calories for growth
5. GH therapy is FDA approved for children with known GH deficiency, SGA, or IUGR without "catch-up", CRF, Turner's syndrome, and Noonan syndrome

Excessive Growth

- Definition: Variation from average pattern of growth in linear height with height > 2 SD above the mean; excess height for age
- Etiology/Incidence
 1. Normal variation in growth—constitutional tall structure (familial, genetic) most common, familial tendency to mature early
 2. Pathologic variations in growth
 - a. Endocrine disorders
 - (1) Infant of diabetic mother (IDM)
 - (2) GH excess—usually due to pituitary adenoma; pituitary gigantism
 - (3) Precocious puberty—androgen or estrogen excess prior to puberty from CNS disorder, adrenal or gonadal disorder, e.g., excess of androgens, estrogens, or both, or idiopathic cause leads to accelerated linear growth, early bone and sexual maturation, and adult short stature; more common in females than males; males have more pathologic etiologies, e.g., CNS disease; females have more idiopathic precocious puberty

- b. Genetic causes
 - (1) Marfan syndrome—autosomal dominant, connective tissue disorder
 - (2) Chromosomal abnormalities (e.g., Klinefelter's syndrome in males with ≥ 2 X chromosomes with usual normal adult stature), XYY, XXYY
 - (3) Fragile X syndrome
 - c. Other
 - (1) Idiopathic or exogenous obesity—early puberty with accelerated growth; usually adult height not beyond expected genetic potential
 - (2) Homocystinuria—inherited inborn error of metabolism
 - (3) Cerebral gigantism (Sotos syndrome)—possible hypothalamic dysfunction; adult stature normal to excessive
 - (4) Beckwith-Wiedemann syndrome
 - (5) McCune-Albright syndrome
- Signs and Symptoms
 1. Concern about tall stature or excessive growth primarily with adolescent girls/parents
 2. Symptoms accompanying tall stature variable depending on underlying etiology
 3. Familial or constitutional tall stature—length normal at birth, tall stature evident by 3 to 4 years; growth rate slows after 4 or 5 years with curve, then parallel to normal curve
 4. IDM—history of maternal diabetes, large for gestational age (LGA)
 5. Beckwith-Wiedemann—LGA, rapid growth in childhood; concern about height; symptoms of hypoglycemia
 6. GH excess—symptoms vary depending on age when excess GH secretion occurs; concern about height, other symptoms of GH excess (e.g., headache, excessive perspiration, visual impairment, coarsening of facial features, enlargement of nose, ears, jaw, increases in hand and feet, galactorrhea, menstrual irregularity, polyuria, polydipsia, joint pain)
 7. Precocious puberty—concern about height, increase in growth rate, and early development of pubic hair common presenting signs
 - a. True precocity of familial or idiopathic origin—causes early secondary sex characteristics with testicular enlargement and spermatogenesis in boys, menarche and mature ova in girls
 - b. Incomplete or pseudoprecocity—adrenal or gonadal tumor or dysfunction causes early secondary sex characteristics but no testicular enlargement, no ovulation
 - c. CNS disorders/tumors—more common in males, may have seizures
 8. Marfan syndrome—concern about height, vision problems
 9. Klinefelter's syndrome—concern about height, school, and behavior problems, lowered verbal IQ, vision problems, delayed adolescent development, testes before puberty normal or small
 10. Obesity—normal height, weight at birth
 11. Homocystinuria—concern about height, mental retardation, vision problems, CNS symptoms, back pain
 12. Cerebral gigantism (Sotos)—concern about height; normal height at birth, rapid growth first year of life ($> 97\%$ height at 1 year) to 3 to 4 years, feeding problems, developmental delay
 - Differential Diagnosis: Normal variants of constitutional tall structure need to be distinguished from pathologic causes; distinguish pseudoprecocity from true sexual precocity; cause of testicular enlargement may be testicular tumor; ovarian tumor may cause early menarche
 - Physical Findings
 1. Constitutional tall stature—variant of normal; 2 to 4 SD above average height for age; normal body proportions; normal physical examination and appropriate pubertal development and timing
 2. Endocrine disorders
 - a. IDM—LGA at birth; $> 90\%$ for height and weight
 - b. GH excess—tall; soft tissue growth; prominent mandible; supraorbital ridge; large nose; space between teeth; hypertension; heart failure; large hands and feet, thickened bones, overgrowth of joints of extremities; visceral enlargement; osteoporosis, kyphosis, may have signs of CNS symptoms
 - c. Precocious puberty—tall stature; in males, secondary sexual development before age 9 years; in females, breast development before 8.0 years, sexual pubic hair before 9.0 years, or menses before 9.5 years
 3. Genetic disorders
 - a. Marfan syndrome—tall stature, dolicocephaly, abnormal body proportions, thin extremities, increased arm and leg length, lowered upper/lower segment ratio, increased arm span, arachnodactyly, myopia and other visual abnormalities, external ear abnormalities, pectus excavatum, heart murmur, scoliosis or kyphosis, laxity and hyperextension of joints, hypotonicity

- b. Klinefelter's syndrome—tall stature, underweight for height and age, mental retardation, long legs, low upper/lower body segment ratio, gynecomastia, normal penile size and pubic hair but small, firm testes with decreased sensitivity to pressure, cryptorchidism, hypospadias
- 4. Other causes of tall stature
 - a. Obesity—accelerated height and weight, generally normal examination otherwise
 - b. Homocystinuria—tall stature, myopia and other ocular problems, CNS symptoms, possible convulsions, mental retardation, osteoporosis, vertebral collapse
 - c. Cerebral gigantism (Sotos syndrome)—large BW and height, dysmorphic features, abnormal body proportions with increased arm span, mental retardation, macrocephaly, dolichocephaly, prominent forehead, hypertelorism with other ocular abnormalities, high-arched palate, pointed chin, CNS findings, poor motor coordination
 - d. Beckwith-Wiedemann syndrome—large BW and height, oomphalocele, umbilical hernia, accelerated growth in childhood, macroglossia, high-arched palate, midface hypoplasia, hemihypertrophy
- Diagnostic Tests/Findings
 1. Previous sequential, consistent recordings of height, weight, and OFC plotted on age-standardized growth charts show height > 2 SD above mean for age
 2. Careful family history of tall growth patterns may elucidate familial etiology of tall stature; growth rate normal, growth curve parallels normal curve in familial tall stature
 3. Laboratory tests to confirm diagnosis based on clinical findings and to rule out endocrine disease
 - a. GH excess—low ACTH, FSH, LH; high or normal GH; abnormal GTT
 - b. Cerebral gigantism—normal GH secretion
 - c. Klinefelter's—high pituitary gonadotropin, LH, FSH; azoospermia
 - d. True precocious puberty—elevated basal LH, FSH concentrations; pubertal LH response to GnRH
 - e. Beckwith-Wiedemann—low blood glucose (hyperinsulinemia)
 4. Radiographic bone age not advanced in constitutional tall stature; advanced in cerebral gigantism, obesity, precocious puberty
 5. Abnormal echocardiogram with Marfan; may have abdominal mass on ultrasound with precocious puberty

6. Abnormalities on skull radiograph, CT, or MRI of cranium with intracranial lesion
7. Karyotype analysis may indicate chromosomal abnormalities, syndromes

- Management/Treatment

1. Consultation or referral to appropriate pediatric specialists for children with other than constitutional tall stature
2. Pharmacologic management controversial; if marked concern about calculated predicted adult height, endocrinologist may accelerate epiphyseal closure with gonadal steroids; if precocious puberty, may slow growth with GnRH agonist
3. Homocystinuria—restrict dietary methionine, supplement dietary cystine
4. GH excess from CNS tumor or adrenal or gonadal tumor—surgery as indicated
5. Management of endocrine disease associated with tall stature
6. Beckwith-Wiedemann—treat excess insulin production

▣ ADRENAL GLAND DISORDERS

Adrenocortical Insufficiency

- Definition: Inadequate production and secretion of adrenal hormones caused by failure of adrenals to secrete glucocorticoids, mineralocorticoids, and adrenal androgen (primary adrenal insufficiency, Addison's) or deficient secretion of ACTH from pituitary (secondary adrenal insufficiency)
- Etiology/Incidence
 1. Primary adrenal insufficiency
 - a. Congenital adrenal hyperplasia (CAH)—hereditary
 - b. Chronic adrenal insufficiency (hypoadrenocorticism, Addison's disease) with destruction of adrenals from infection or hereditary autoimmune disease or adrenal calcification
 - c. Congenital absence or underdevelopment of adrenals; newborn adrenal hemorrhage (with complicated or traumatic delivery)
 - d. Malignancy (adrenal tumor)
 2. Secondary adrenal insufficiency
 - a. Hypopituitarism—deficient secretion of ≥ 1 pituitary hormones due to congenital brain malformations, head trauma, histiocytosis, infection, tumors, radiation
 - b. Cessation of glucocorticoid therapy after prolonged large-dose administration of glucocorticoids (e.g., asthma, nephrosis, leukemia)

- c. Other—postsurgical, infants of steroid-treated mothers, respiratory distress syndrome, anencephaly, pituitary defects
- Signs and Symptoms
 1. Acute adrenal insufficiency (adrenal crisis)
 - a. Known or unknown diagnosis of Addison's—crisis may be first presentation of illness
 - b. Recent or concurrent infection or febrile illness; surgery; exposure to excessive heat in susceptible individuals
 - c. History of abrupt withdrawal of large-dose steroid therapy
 - d. Hypotension, shock, muscle weakness, weight loss, confusion, fever, fatigue, nausea, vomiting, diarrhea, dehydration, abdominal pain, anorexia, salt craving despite anorexia, symptomatic hypoglycemia
 2. Chronic adrenal insufficiency—weakness, fatigue, anorexia, hypotension, hyperpigmentation, poor weight gain, weight loss, lethargy, exercise intolerance, headache, nausea, vomiting which may be forceful or projectile, diarrhea, dehydration, salt craving, increased pigmentation, hypotension
- Differential Diagnosis
 1. Differentiate acute adrenal insufficiency from severe acute infections, diabetic coma, CNS disturbances, acute poisoning; in newborn, distinguish from respiratory distress, intracranial hemorrhage, sepsis
 2. Differentiate chronic adrenal insufficiency from anorexia nervosa, muscular disorders (e.g., myasthenia gravis), nephritis, chronic infection (e.g., TB); consider chronic form of disease if patient has recurrent spontaneous hypoglycemia
- Physical Findings
 1. Acute adrenal insufficiency (adrenal crisis)—moribund, fever followed by hypothermia, cachexia, acutely dehydrated, vomiting, hypotension, confusion, coma, weakness, abdominal pain, nausea, increased skin pigmentation
 2. Chronic adrenal insufficiency (Addison's)—lethargy, dehydration, poor weight gain, weight loss, vomiting, hypotension, small heart size on radiograph; bronze skin pigmentation, especially of areola, mucous membranes, hand creases, axilla and groin, extensor surfaces of joints, and surgical scars
- Diagnostic Tests/Findings
 1. Laboratory studies—decreased serum sodium and bicarbonate, PaCO₂, blood pH, and blood volume; hyperkalemia and increased BUN; urinary sodium and sodium-to-potassium ratio elevated relative to degree of hyponatremia; eosinophilia; moderate neutropenia
 2. Confirmatory tests to assess functional capacity of adrenal cortex
 - a. Lack of response in plasma cortisol and aldosterone to ACTH stimulation testing
 - b. Increased baseline serum ACTH in primary adrenal failure
 - c. Decreased urinary free cortisol and 17-hydroxycorticosteroid excretion
 - d. Metyrapone test (in hospital)—nonelevation of morning 11-deoxycortisol level after midnight dose of metyrapone
 - e. Corticotropin-releasing hormone (CRH) test abnormal
- Management/Treatment
 1. Immediate treatment of life-threatening acute adrenal crisis
 2. Referral to endocrinologist for suspected or confirmed adrenal insufficiency
 3. Pharmacologic—after initial stabilization, chronic corticosteroid replacement with glucocorticoids (e.g., hydrocortisone), mineralocorticoid (e.g., fluorocortisone) and/or sodium chloride (table salt); increased dosages may be needed during severe illness, trauma, stress, or surgery
 4. Monitor for steroid excess, particularly for decreasing height and weight; monitor for insufficient glucocorticoid treatment (headache, weight loss, nausea, hypotension); monitor blood glucose, ACTH, sodium, and potassium
 5. Avoid abrupt withdrawal of corticosteroids to avoid adrenal crisis
 6. Education regarding risk of acute episodes; medical alert identification tag; consider emergency hydrocortisone injection kit for use in accident or severe stress

Adrenocortical Hyperfunction

- Definition: Excessive production and secretion by the adrenal gland of cortisol, adrenocortical androgens, estrogen, and/or aldosterone
- Etiology/Incidence
 1. Hypercortisolism (Cushing's)—excess cortisol (ACTH) secretion by adrenals
 - a. Adrenal tumors (Cushing's syndrome)—relatively rare but occurs in all ages

- b. Pituitary adenomas (Cushing's disease)
 - c. Chronic exposure to glucocorticoids to treat inflammation and for immunosuppression (iatrogenic hypercortisolism)
 - d. Ectopic ACTH-secreting tumors—non-pituitary tumors stimulate the adrenal cortex, causing excess ACTH secretion; rare; usually seen in children < 12 years
2. Adrenogenital syndrome—virilizing adrenal tumor causes elevated adrenal androgen secretion
 3. Feminizing adrenal tumors—causes elevated adrenal estrogen secretion
 4. Hyperaldosteronism
 - a. Secondary—physiologic attempts to maintain homeostasis with serum electrolytes and fluid volume due to renal compromise or physiologic response to severe illness
 - b. Primary—due to adrenal tumor or hyperplasia
- Signs and Symptoms
 1. Ubiquitous effects of adrenal hormones lead to multiple and varied signs and symptoms
 2. Hypercortisolism (Cushing's)—slowed growth and development, obesity, emotional lability (depression and euphoria), delayed pubertal onset, easy bruising, increased appetite, back pain
 3. Adrenogenital syndrome—increase in linear growth rate and muscle development, acne, premature pubarche, development of secondary sex characteristics in boys, enlarged and erectile clitoris in females, menstrual irregularities in older girls
 4. Feminizing adrenal tumors—rapidly increasing height, development of secondary sex characteristics in girls with possible breakthrough vaginal bleeding; gynecomastia in males
 5. Hyperaldosteronism—in infants, FTT, vomiting, weakness; may have history of recent diarrhea, increased sweating, heat exposure; history of renal or liver disease (e.g., cirrhosis, nephritis, renal ischemia); in primary hyperaldosteronism, muscle weakness, unusual periodic paralysis; paresthesias, tetany; polyuria; polydipsia
 - Differential Diagnosis
 1. Hypercortisolism should be distinguished from exogenous obesity; virilizing adrenal tumors from virilizing gonadal tumors; and feminizing adrenal tumors from premature thelarche and idiopathic sexual precocity
2. Hyperaldosteronism due to physiologic response to maintain homeostasis in severe illness vs. pathology (e.g., renal disease, adrenal tumor) should be determined
- Physical Findings
 1. Ubiquitous effects of adrenal hormones lead to multiple and varied clinical findings
 2. Hyperadrenocorticism—poor growth rate may precede obesity or other symptoms; relatively short stature; obesity; purple striae; truncal obesity; “buffalo type” adiposity of face, neck, and trunk; fat pad in interscapular area; plethoric or “moon” facies; delayed onset of secondary sex characteristics; muscle weakness; may have virilism; may have hemihypertrophy; delayed skeletal maturation; osteoporosis, especially of spine
 3. Adrenogenital syndrome—increase in linear growth rate; hirsutism; acne; deepening voice; increased muscle mass; masculinization of prepubertal children (boys with pubic, axillary, and sometimes facial hair with adult-size penis, frequent erections, prepubertal or slightly enlarged testes) (girls with pubic and axillary hair with enlarged and erectile clitoris); advanced bone age
 4. Feminizing adrenal tumor—increase in linear growth rate; gynecomastia in males; prepubertal testes, pubic hair; breast development in females, may have pubic hair; advanced bone age
 5. Primary hyperaldosteronism—in infants, FTT; weakness; dehydration; tetany; hypertensive or normotensive; nocturnal enuresis; muscle weakness, unusual periodic paralysis
 - Diagnostic Tests/Findings
 1. Various confirmatory tests are used to determine specific etiology of adrenal hyperfunction
 2. Plasma cortisol concentrations elevated; may be loss of normal diurnal variation in cortisol secretion
 3. Low serum chloride and potassium levels; may have elevated sodium, pH, CO₂
 4. Serum ACTH slightly elevated with adrenal hyperplasia (Cushing's disease), decreased with adrenal tumor, and very elevated with ACTH-producing pituitary or ectopic (extrapituitary) tumors
 5. Low eosinophil counts; leukocyte count with polymorphonuclear leukocytosis with lymphopenia, possibly elevated RBC
 6. Elevated urinary free cortisol and urinary 17-hydroxycorticosteroid excretion; abnor-

- mal urinary 17-ketosteroid excretion; may have glycosuria
- 7. Abnormal patterns of dexamethasone suppression and abnormal CRH stimulation tests
- 8. Adrenogenital syndrome, androgen secretion not suppressed by dexamethasone administration; advanced bone age
- 9. Feminizing adrenal tumor—elevated adrenal steroids in urine; elevated urinary and plasma estrogen levels; advanced bone age
- 10. Hyperaldosteronism, hypokalemia, high aldosterone; may have low or elevated plasma renin level; may have increased chloride, potassium, and prostaglandin excretion in urine
- 11. CT and MRI to assess for adrenal tumors; pituitary imaging to assess for pituitary tumor

- Management/Treatment

1. Referral to pediatric endocrinologist for management of suspected or confirmed adrenocortical hyperfunction
2. Surgery indicated for adrenal tumors, pituitary adenomas, ectopic ACTH-producing tumors
3. ACTH pre and postoperatively to stimulate nontumorous contralateral adrenal cortex
4. Discontinue excessive glucocorticoid therapy if adrenocortical hyperfunction

■ MISCELLANEOUS

Hypoglycemia

- Definition: Symptoms provoked by abnormally low blood glucose levels occurring when child with diabetes receives excessive insulin, fails to eat, or exercises too strenuously; in children without diabetes, blood glucose level must be < 60 mg/dL and < 45 mg/dL in newborns (95% CI)
- Etiology/Incidence
 1. Transitional hypoglycemia
 - a. Occurs during the first 12 hours of life as neonate transitions from interuterine to extrauterine homeostasis
 - b. Refers to only AGA infants
 - c. Treatment is early and frequent feeding on demand (when indicated by feeding cues)
 2. Transient neonatal hypoglycemia
 - a. Most common in preterm infants; these infants have low glycogen and fat reserves and delayed maturity of enzymes needed for gluconeogenesis and fatty acid oxidation
 - b. LGA infants of diabetic mothers (IDM) with hyperplasia of beta cells of pancreas due to chronic intrauterine exposure to elevated maternal blood sugar

- c. Increased glucose use from physiologic stressors secondary to asphyxia, anoxia, respiratory illness, heart disease, infection, cold injury, or starvation
3. Hypoglycemia of childhood
 - a. Hyperinsulinism—caused by abnormalities of beta cells (e.g., adenoma, islet cell hyperplasia); most common cause of persistent, recurrent hypoglycemia in first year; peak onset in first year and after age 3 years
 - b. Functional fasting hypoglycemia (“ketotic hypoglycemia”)—occurs between 1 and 5 years of age; peaks at 2 years; more common in SGA infants, males, and children with BW < 2500 g; associated with fasting in past 24 hours; episodes may occur periodically and with less frequency; rare after 5 years
 - c. Associated with CNS disorders
 - d. Metabolic disorders and endocrine insufficiency, e.g., deficiency; galactosemia, hypopituitarism, congenital hypothyroidism, adrenal insufficiency; usually apparent in first 2 years of life
 - e. Severe malnutrition states, e.g., chronic diarrhea, liver disease
 - f. Other—drug ingestion, drug toxicity (alcohol, aspirin, oral hypoglycemic agents); associated with accidental ingestion or deliberate administration

- Signs and Symptoms

1. Neonatal hypoglycemia findings variable depending on degree of hypoglycemia; irritable, jittery, refusal to feed; tend to be small or large for gestational age
2. Childhood hypoglycemia
 - a. Findings variable depending on degree and etiology of hypoglycemia; may have mood changes, nervousness, weakness, hunger, epigastric pain, vomiting
 - b. May have history of diabetes with recent history of excessive insulin, failure to eat, strenuous exercise; complaints of hunger; intentional or accidental ingestion of oral hypoglycemic agents, alcohol, salicylate
 - c. May have known personal or familial history of inherited metabolic disorder or inherited hormonal deficiency; may have symptoms of metabolic disorder or hormonal deficiency
 - d. Functional fasting (“ketotic hypoglycemia”)—history of vomiting, anorexia, failure to eat previous 24 hours, often have URI; may have early morning seizures

- Differential Diagnosis: Distinguish among various possible etiologies of hypoglycemia (e.g., functional ketotic vs. inherited metabolic and endocrine disease) because of underlying implications for management
- Physical Findings
 1. Neonatal hypoglycemia findings variable
 - a. Cachexic or macrosomic infant
 - b. Irritability, lethargy, weak cry
 - c. Hypothermia, cyanosis, diaphoresis, pallor
 - d. Uncoordinated eye movements, eye-rolling
 - e. Apnea, irregular breathing, tachycardia
 - f. Twitching, jitteriness, convulsions, semi-consciousness, or coma
 2. Childhood hypoglycemia
 - a. Signs as above
 - b. May have diminished growth, difficulty talking, signs of other systemic illness, abdominal or pelvic masses, unsteady gait, concurrent illness
- Diagnostic Tests/Findings
 1. Transient neonatal hypoglycemia, routine assessment of all newborns after birth; especially if high risk for hypoglycemia, e.g., infant of diabetic mother, IUGR, birth asphyxia, cardiac/CNS disease, sepsis (published statistical definition of hypoglycemia is a level of less than 40 mg/dL)
 2. Low blood glucose levels during hypoglycemic episode; consistent and repeated levels below 40 mg/dL with associated signs and symptoms need further confirmatory tests
 3. In hyperinsulinemia, serum insulin levels may be inappropriately elevated when compared with glucose level obtained at same time
- Management/Treatment
 1. Consultation with or referral to endocrinology provider for delineation of etiology and management
 2. Treat hypoglycemic episodes promptly and adequately to prevent CNS injury; especially important for younger children
 3. Hypoglycemic reactions in children with diabetes—see diabetes mellitus in this chapter
 4. Surgery for pancreatic adenoma, partial pancreatectomy if insulin secretion suppression unsuccessful
 5. Children with functional (fasting, ketotic) hypoglycemia—treat with liberal carbohydrate diet with bedtime snacks, moderate restriction on ketogenic foods; avoid prolonged fasting,

especially if child ill; parents may need to check urinary ketones

Hyperglycemia

- Definition: Common hereditary metabolic and endocrine disorder characterized by insulin deficiency resulting in abnormal metabolism of carbohydrate, protein, and fat (type 1, formerly termed insulin-dependent diabetes mellitus [IDDM]), or may have specific carbohydrate intolerance causing insulin resistance at tissue level (type 2 diabetes, formerly termed noninsulin dependent diabetes mellitus [NIDDM])
- Etiology/Incidence: Hyperglycemia results from mechanisms that cause failure of pancreatic beta cell function or other processes not directly involving beta cells; frequency of diabetes mellitus (DM) is rising, increases with age, average age of onset decreasing
 1. Type 1—destruction of beta islet cells of pancreas; uncommon in infancy and toddlerhood, increases until adolescence, then drops sharply; peaks occur between 5 and 7 years and at time of puberty; boys and girls equally affected; more frequently diagnosed in winter, especially with adolescents; environmental factors may increase or decrease expression of diabetes in susceptible individuals; multifactorial, associated with genetic predisposition and environmental factors including viruses (e.g., mumps, coxsackie, congenital rubella), environmental toxins, nutrition, and physical and emotional stress
 - a. Type 1A—immune-mediated (previously termed juvenile diabetes or insulin-dependent diabetes); most common type of diabetes in those < 40 years
 - b. Non-type 1—nonimmune-mediated insulin-deficient diabetes; frequently found in overweight teens and in black and Hispanic children who develop diabetes
 2. Type 2—non-insulin-dependent, non-immune mediated; associated with being overweight; not being prone to ketosis and insensitivity to insulin; insulin resistance associated with childhood obesity, onset most often > 10 years of age; more likely in children and adolescents of predominantly Native American, Hispanic, African-American origins; rapidly increasing form of diabetes; includes maturity-onset diabetes of youth (MODY) characterized by slow onset; treatment with diet and exercise and oral hypoglycemics after initial treatment with

- insulin, less severe symptoms, and autosomal dominant inheritance pattern
3. Other—associated with administration of high doses of steroids (e.g., in renal disease, rheumatoid arthritis, asthma, leukemia) or antileukemic drugs; excessive endogenous cortisol production; CNS tumors, pheochromocytoma; cystic fibrosis
- Signs and Symptoms
 1. May present with very mild symptoms or in diabetic ketoacidosis
 2. Symptoms of type 1 diabetes mellitus
 - a. Polyuria, polydipsia, polyphagia (primary complaints)
 - b. Weight loss or failure to gain weight; variable decrease in linear growth
 - c. Behavioral changes, headache, emotional lability, fatigue, recent “flulike” illness
 - d. Abdominal pain, nausea, vomiting, constipation, nocturia, enuresis
 - e. History of recent illness, stress, missed insulin if known diabetic
 3. Symptoms of type 2 diabetes mellitus
 - a. Many of the above symptoms but generally less severe than in type 1 diabetes mellitus
 - b. Mild to moderate polyuria and polydipsia
 - c. Weight loss
 - d. History of high caloric intake and sedentary lifestyle
 - e. Positive family history of type 2 diabetes mellitus
 - Differential Diagnosis
 1. Diabetes insipidus
 2. Nondiabetes causes of polyuria, e.g., psychogenic polydipsia, CNS injury, tumors
 3. Other causes of fatigue, weight loss, behavioral change, e.g., Hashimoto’s hypothyroidism, systemic illness
 - Physical Findings
 1. Type 1 diabetes mellitus
 - a. Dehydration, irritability
 - b. Weight loss
 - c. Visual disturbances
 - d. Fatigue, muscular weakness, declining physical performance
 - e. Long-term complications—joint contractures, diabetic retinopathy, compromised renal function
 2. Type 1 diabetes with diabetic ketoacidosis
 - a. Marked dehydration, irritability, lethargy, drowsiness, stupor, coma
 - b. Tachycardia, cardiac arrhythmias, Kussmaul breathing (long, deep, labored breathing)
 - c. Dry mucous membranes, cherry-red lips, hypotension, rapid and thready pulse, hyperventilation, low temperature, “fruity” acetone breath
 - d. Vomiting, abdominal spasm, tenderness
 - 3. Type 2 maturity-onset diabetes of youth (MODY)
 - a. Marked obesity
 - b. Acanthosis nigricans
 - c. Vaginal candidiasis in females
 - Diagnostic Tests/Findings
 1. Glycosuria; urinary ketones
 2. Elevated blood glucose—random blood glucose level $>$ or equal to 200 mg/dL or fasting blood glucose levels $>$ or equal to 126 mg/dL sufficient to make diagnosis of type 1 diabetes
 3. Elevated plasma glucose—repeated findings of fasting plasma glucose level $>$ 126 mg/dL or plasma glucose level $>$ 200 mg/dL when taken either randomly (with symptoms of diabetes) or two hours after oral glucose load
 4. Elevated glycosylated hemoglobin (HbA_{1c})
 5. In mild diabetic ketoacidosis (DKA) venous blood pH 7.2 to 7.3; moderate DKA venous blood pH 7.10 to 7.19; severe DKA venous blood pH $<$ 7.10
 6. In type 2 diabetes in children—criteria overweight BMI $>$ 85th percentile for age and sex; weight for height $>$ 85th percentile, PLUS any 2 of the following—positive family history of type 2 diabetes in 1st or 2nd degree relative, ethnic race of high likelihood of diagnosis, positive signs/symptoms of insulin resistance (e.g., acanthosis nigricans, PCOS), or maternal history of diabetes or GDM; possible positive antibody tests—HLA, islet cell; insulin autoantibody in type 2 diabetes, negative islet cell antibodies
 - Management/Treatment
 1. Immediate hospitalization if severe diabetic ketoacidosis
 2. Consultation with physician or referral to pediatric endocrinology team for diagnostic evaluation, initial care and management, and/or for confirmation/management of associated diseases, e.g., hypothyroidism; referral as needed for treatment by ophthalmologist and for emotional/behavioral disorders
 3. After initial stabilization, for children with type 2 diabetes, slow gradual weight loss, exercise regimen, education stressing lifestyle changes

- to achieve weight loss, oral hypoglycemic agents following insulin if insulin initially needed to stabilize blood glucose levels
4. After initial stabilization, children with type 1 diabetes generally treated with combination of short- and long-acting insulin; types, onset, peak action, and duration of insulin varies (see **Table 14-1**)
 5. Critical goal of treatment to maintain blood glucose levels at near normal to prevent acute complications (e.g., hypoglycemia and ketoacidosis), intermediate complications (e.g., lipoatrophy and lipohypertrophy, limited joint mobility, growth failure, pubertal delay), and chronic complications (e.g., retinopathy, nephropathy, and neuropathy)
 6. Educate child and parents regarding disease; self-monitoring of blood glucose minimally 4 times daily; insulin types, timing, dosages, adjustments; optimal nutrition; need for consistency in meal and snack time and in moderate, regular exercise; need for increased insulin during illness or stress; prompt treatment of infection; signs and symptoms of hypoglycemia and ketoacidosis; urine testing; complications of poor blood glucose management; availability of glucose and glucagon for acute hypoglycemic episodes (0.5 mg for children < 5 yrs, 1 mg sc or IM for older children)
 7. Encourage parents to obtain user-friendly resources for helping them to better understand the disease and its management e.g., *Understanding Diabetes* (11th ed.) by H. P. Chase (2006) in English and Spanish and coloring books about diabetes available from The Guild of the Children's Diabetes Foundation 1 (800) 695-2873
 8. Supportive care of patient and family through stages of grieving following diagnosis; emphasize normalcy, view of child as “child with diabetes” vs. “diabetic child”
 9. Recognize phases of diabetes—development of clinical symptomatology; clinical remission or “honeymoon” period of variable duration due to improved beta cell function after initial therapy started; relapse with progressive increase in insulin requirement
 10. Ideal fasting blood glucose level varies with age of child (*Standards of Medical Care in Diabetes*, 2009)
 - a. Toddler and preschool (age 0–6) 100 to 180 mg/dL
 - b. School age (age 6–12) 90 to 180 mg/dL
 - c. Adolescent and young adult (age 13–19) 70 to 130 mg/dL
 11. Treatment of insulin reactions (*Standards of Medical Care in Diabetes*, 2009) http://care.diabetesjournals.org/content/32/Supplement_1/S13.full
 - a. Mild reaction (conscious, hunger at unusual time, paleness, shakiness, or irritability)—do blood sugar test if possible; if blood sugar < 70 mg/dL, give 15 to 20 g fast-acting carbohydrate (example: 4 oz of juice or sugared soda, 15 Skittles, 5 Life Savers, or 1 small tube of cake gel; wait 10 to 15 minutes for sugar to absorb, then recheck BG; if not above 80 mg/dL, retreat; if BG above 80 mg/dL and next

■ **Table 14-1** Bioavailability Characteristics of Insulins

	Human Insulin Type	Onset	Peak Action	Duration
Rapid-Acting Analogs	Humalog (insulin lispro) Novolog (insulin aspart) Apidra (insulin glulisine)	15–30 minutes 10–20 minutes 15–30 minutes	0.5–2.5 hours 1–3 hours 0.5–2.5 hours	5 hours or less 3–5 hours 5 hours or less
Short-Acting Insulin	Regular	0.5–1 hour	2–5 hours	5–8 hours
Intermediate-Acting Insulin	NPH	1–2 hours	2–12 hours	14–24 hours
Long-Acting Insulin Analogs	Lantus (insulin glargine) Levemir (insulin detemir)	1.5 hours 1.5 hours	very small, early steady state after 2nd injection	20–24 hours dose-dependent (bid dosing)
Intermediate-/Rapid-Acting Insulin Mix	Humalog Mix (75%NPH/25%lispro) Novolog Mix (70%insulin aspart protamine suspension/ 30% aspart)	15–30 minutes 10–20 minutes	0.5–2.5 hours 2.4 hours	up to 24 hr up to 24 hr

- meal greater than 1 hour away, give solid food snack (crackers, half sandwich, fresh fruit, etc.) to maintain blood sugar; if symptomatic, but BG not below 70 mg/dL, may give small snack
- b. Moderate reaction (very confused, very pale, very shaky)—give Insta-Glucose, Reactose, Monojel, or any source of simple sugar, like sugared soda or juice (15–20 g); do blood sugar test as soon as possible, repeat blood test after 10 to 15 minutes to make sure blood sugar is > 80 mg/dL; if blood sugar is not > 80 mg/dL, repeat initial treatment and wait another 10 minutes; once blood sugar is > 80 mg/dL, give solid food (protein source)
 - c. Severe reaction (loss of consciousness, seizure, convulsion)—give glucagon and do blood sugar level as soon as possible; if person does not improve after 10 to 20 minutes, call 911 or get extra help; a second dose of glucagon can also be given or intravenous 10% dextrose (1–2 mL/kg)
 - d. Avoidance of recurrent blood glucose concentrations below 72 mg/dL may prevent the development of hypoglycemia unawareness
12. Glycosylated hemoglobin (HbA_{1c}) blood test every 3 months to assess overall blood sugar levels and to help in ascertaining compliance with treatment regimen; gives reliable measurement of long-term glycemic control during preceding 3 months
 - a. In adults, HbA_{1c} values below or around 7% have been shown to reduce microvascular and neuropathic complications
 - b. In children, goals vary by age:
 - (1) Toddlers and preschoolers (age 0–6 yrs) A_{1c} below 8.5%, but above 7.5%; higher levels are allowed in younger children to reduce risk of effects of hypoglycemia on developing brains
 - (2) School age (age 6–12 yrs) A_{1c} goal < 8%
 - (3) Adolescents and young adults (age 13–19 yrs) A_{1c} goal < 7.5%
 - c. Goals should be individualized and based on reasonable benefit-risk assessment
 - d. Goals should be less strict in children with frequent hypoglycemia or hypoglycemic unawareness
 13. Sexually-active adolescents need instruction regarding pregnancy; oral contraceptives are acceptable if blood pressure is normal
 14. Explore with parents additional topics such as use of insulin pumps; successful insulin pump therapy requires parental motivation, ability to

understand pump technology, frequent blood glucose monitoring, and willingness to work closely with the diabetes multidisciplinary team

15. Also discuss use of insulin pens, obtaining medical alert bracelets, communication with extended family and school personnel about child's illness, school diabetes health plans (504 plans—sample plans available www.childrenwithdiabetes.org) and the research directed at diabetes prevention

Disorders of Pubertal Development

- Definition: Abnormal development or delay in initiation of secondary sexual characteristics
- Etiology/Incidence: Abnormalities in pubertal development associated with CNS or gonadal disorder or dysfunction; includes temporary and permanent delays of pubertal onset or true (complete) or pseudoprecocious (incomplete) puberty
 1. Intersex—ambiguous genitalia or inappropriate for gonadal sex due to endocrinopathy
 2. Normal pubertal development—in females, Tanner 2 breast development occurs 8.0 to 10.4 years (white girls); 6.6 to 9.5 years (black girls); and 6.8 to 9.8 years (Hispanic girls); in males, Tanner 2 pubic hair median age 12.0 years (white boys); 11.2 (black boys); and 12.3 years (Hispanic boys)
 3. Precocious puberty—secondary sexual characteristics before 9 years in males; in females, onset of breast development before 8 years, sexual pubic hair before 9.0 years or menses before the age of 9.5 years; however, breast and pubic hair development in females may occur normally for some younger girls, see above
 - a. True (complete) precocious puberty is mediated by pituitary gonadotropin secretion involving all secondary sex characteristics
 - (1) Gonadotropin-releasing hormone-dependent (central)
 - (a) Idiopathic (sporadic or familial)
 - (b) CNS abnormalities
 - (c) Tumors (e.g., LH-secreting adenoma, astrocytoma, craniopharyngiomas)
 - (2) Gonadotropin-releasing hormone-independent (peripheral)
 - (a) Genetic (i.e., CAH in males, McCune-Albright syndrome)
 - (b) Tumors (e.g., adrenal, ovarian, or testicular)
 - (c) Limited (e.g., chronic primary hypothyroidism, ovarian cysts)

- b. Pseudoprecocious (incomplete) puberty involves one type of secondary sexual characteristic (e.g., premature thelarche [breast development] or pubarche [pubic hair development]) mediated by excessive estrogen/androgen stimulation for age from ovaries/testes, adrenal cortex, or exogenous sources
 - 3. Hypogonadism—causes lack of secondary sexual characteristics (sexual infantilism)
 - a. In boys, lack of secondary sexual characteristics after 17 years suggests abnormal testicular maturation; may be due to testicular failure or dysfunction (primary failure due to anorchia, castration, Klinefelter's, mumps, radiation, trauma, tumor, endocrinopathies, etc.) or to pituitary/hypothalamic dysfunction (panhypopituitarism, empty sella syndrome, gonadotropin deficiency, LH, FSH deficiencies, endocrinopathies, etc.)
 - b. In girls, may have lack of onset of secondary sex characteristics and amenorrhea due to primary ovarian failure (due to gonadal dysgenesis, enzyme defects, infection, surgery, radiation, chemotherapy, etc.) or to secondary ovarian failure (hypothalamic disorder or dysfunction, CNS irradiation, eating disorders, excessive exercise, chronic illness, etc.)
- Signs and Symptoms
 1. If intersex, history of confusion of sex assignment at birth
 2. May have normal growth and development through childhood; at puberty, may have abnormal sexual development; females may have virilization, primary amenorrhea; males may have incomplete virilization
 3. May have history of underlying disease, dysfunction, or systemic illness causing delay in/ or premature pubertal onset; history of exposure to radiation, drugs, etc.
 4. May have history of early development of one or all secondary sex characteristics, tall stature, symptoms of endocrine disease (e.g., hypothyroidism, congenital adrenal hyperplasia), intracranial disease (e.g., visual disturbances), abdominal disease (e.g., adrenal or gonadal tumor) or dysfunction (see growth disturbances this chapter)
 5. May have delayed development of secondary sex characteristics and symptoms of metabolic or endocrine disturbances (e.g., constipation); intracranial disease (e.g., failure to grow, seizure); syndrome stigmata (e.g., developmental delays); abdominal disease (abdominal

enlargement) or dysfunction (see growth disturbances and adrenal gland disorders this chapter)

- Differential Diagnosis
 1. Ambiguous genitalia or suspected intersex—distinguish true hermaphroditism (both ovarian and testicular tissue present), female pseudohermaphroditism (female genotype, only ovaries), and male pseudohermaphroditism (genotype male, only testes)
 2. Structural abnormalities of genital tract and associated intracranial, endocrine, abdominal, or pelvic disease
 3. Precocious puberty—distinguish between true (complete) precocious puberty mediated by pituitary gonadotropin secretion involving all secondary sex characteristics vs. pseudoprecocious (incomplete) puberty involving one type of secondary sexual characteristics
 4. Premature sexual development—distinguish normal variants of premature thelarche (isolated premature breast development) and pubarche (early pubic hair development) in girls from pathologic causes; distinguish normal variants of premature adrenarche in boys (adrenal maturation with pubic hair and body odor) from pathologic causes
- Physical Findings
 1. May have normal or abnormal physical examination depending on underlying etiology of pubertal disorder (e.g., normal examination in constitutional delay, findings of chronic or systemic illness, or midfacial defects in pathologic pubertal delay, tall stature in Klinefelter's, etc.)
 2. May have normal or abnormal genitalia depending on underlying etiology of pubertal disorder and timing (e.g., genitalia may be ambiguous in congenital adrenal hyperplasia; may have cryptorchidism in sexual infantilism or in pituitary insufficiency; may have abdominal mass or testicular mass in gonadal tumor; may have microphallus in human GH deficiency, may have small testes in testicular failure, may have abnormal pelvic examination, etc.)
 3. With precocious puberty, may have tall stature, premature secondary sex characteristics
 4. With contrasexual pubertal development may have gynecomastia in males due to hypogonadism from Klinefelter's; excessive virilization of prepubertal girls with pubic hair, oily skin, acne, clitoromegaly, hirsutism
 5. With delayed pubertal development for age (constitutional delay of puberty), may have

normal examination except for delayed growth and development (short stature but low normal growth rate) during childhood; delayed bone age; relatively short legs for height or greater upper/lower body ratio; descended normal testes that are prepubertal in size, consistency

6. With delayed or interrupted pubertal development may have findings consistent with endocrine disorder (e.g., anosmia, the lack of ability to smell, and micropenis in gonadotropin abnormalities; hirsutism and dry hair and skin in hypothyroidism, etc.)
- **Diagnostic Tests/Findings**
 1. Tanner staging of breast, genitals, pubic hair development
 2. Laboratory findings variable depending on etiology of pubertal disorder; tests may include plasma or serum LH, FSH; GH, electrolytes, thyroid tests; blood and urinary pH, urine specific gravity, sedimentation rates, BUN, creatinine, etc.
 3. Tests of hormonal function as needed (e.g., GnRH to differentiate hypogonadotropic hypogonadism and constitutionally delayed puberty)
 4. CT, MRI, or ultrasound as needed to rule in or rule out central CNS, adrenal, renal, gonadal, or thyroid disease; bone age
 5. Other laboratory tests as needed—karyotype if ambiguous genitalia or Klinefelter's suspected
 - **Management/Treatment**
 1. Physician consultation or referral to pediatric endocrinologist for suspected pathologic cause of pubertal delay or abnormal development
 2. Treatment dictated by identified etiology for pathologic cause of pubertal delay or abnormality in pubertal development (e.g., hormonal therapy may be immediate, as with congenital adrenal hyperplasia, or at puberty with testosterone for testicular failure)
 3. Management of underlying endocrine disorders and diseases and systemic illness (e.g., hypothyroidism, poorly controlled diabetes, anorexia, inflammatory bowel disease, etc.)

Gynecomastia

- **Definition:** Visible or palpable glandular enlargement of the male breast occurring commonly in healthy adolescent males (pubertal gynecomastia); occasionally indicative of underlying disease (pathologic gynecomastia); seen frequently in newborns (neonatal gynecomastia)

- **Etiology/Incidence**
 1. Neonatal gynecomastia—due to cross-placental transfer of maternal hormones; usually resolves by 2 to 3 weeks
 2. Pubertal gynecomastia—influence of too little androgen and/or too much estrogen on mammary tissue; transient, may occur in 40% to 66% of normal boys during puberty; onset between 10 and 12 years, peak occurrence 13 and 14 years, usually self-limiting; resolves by 16 to 17 years of age
 3. Pathologic gynecomastia—secondary to drug side effects (e.g., cimetidine, digitalis, phenothiazine, treatment with hCG, testosterone, or estrogen), underlying disease or syndromes (e.g., Klinefelter's), injury to the nervous system, chest wall or testes; or, may be idiopathic
- **Signs and Symptoms**—breast development in other than pubertal females
- **Differential Diagnosis**
 1. Transient pubertal gynecomastia from obesity (lipomastia) and pathologic causes; tumor (lipoma, neurofibroma, cancer)
 2. Breast infection
 3. Fat necrosis due to injury
 4. Drugs (estrogens, anabolic steroids, marijuana)
 5. Klinefelter's gonadal dysfunction
- **Physical Findings**
 1. Neonatal; usually bilateral, often asymmetric breast tissue enlargement; resolves within 1 to 2 weeks
 2. Pubertal (physiologic) gynecomastia—breast tissue enlargement glandular, movable, disk-shaped, below areola, nonadherent to skin or underlying tissue; typically breasts unequal in size and < 3 cm in diameter; breasts may be tender, nipples irritated due to rubbing on clothing; if tissue \geq 4 to 5 cm/diameter and breasts dome-shaped, macrogynecomastia present; Tanner stages II to IV pubertal development with testes \geq 3 cm length
 3. Pathologic gynecomastia—malnourishment, lymphadenopathy, delayed sexual maturity with undermasculinization, signs of chronic disease (e.g., goiter, liver or renal disease, endocrinopathies, cancer, colitis, CF, AIDS); breast tissue \geq 3 cm/diameter, asymmetric, hard, fixed, indurated, not directly beneath areola; may have absent, underdeveloped, or asymmetric testes

- Diagnostic Tests/Findings
 1. Endocrinology studies as indicated
 2. Imaging techniques as appropriate—ultrasonography of testes to identify impalpable testicular tumor; CT, MRI of abdomen to identify adrenal tumors
 3. Karyotyping if Klinefelter's suspected
- Management/Treatment
 1. Neonatal—parent education and reassurance about etiology, transience, and normalcy of condition
 2. Pubertal (physiologic) gynecomastia < 4 cm—explanation, reassurance, and observation; regression usually spontaneous, within a few months, rarely beyond 2 years
 3. Physiologic macrogynecomastia (≥ 4 cm mass)—medical or surgical treatment usually required as regression rare, especially if gynecomastia present for > 4 years; pharmacologic therapy (e.g., tamoxifen, danazol, testosterone) sometimes used
 4. Gynecomastia usually very upsetting to adolescent but often not discussed because of embarrassment; reassure about transience and spontaneous regression

Menstrual Disorders: Amenorrhea

- Definition
 1. Primary amenorrhea—failure of onset of menarche in females who are 16 years and have normal pubertal growth and development; 14 years with absence of normal pubertal growth and development; or in girls who have not begun menstruation 2 years after completed sexual maturation
 2. Secondary amenorrhea—absence of menstruation for > 3 cycles or at least 6 months after menstruation established
- Etiology/Incidence
 1. Primary amenorrhea
 - a. Constitutional/familial (common)
 - b. Obstructions of menstrual flow (e.g., fusion or stenosis of labia, imperforate hymen)
 - c. Estrogen deficiency
 - (1) Primary ovarian insufficiency—organic or functional ovarian failure (e.g., anatomic anomalies, pelvic irradiation, enzyme defects, autoimmune disease, infection)
 - (2) Secondary ovarian insufficiency—organic or functional ovarian failure

- from hypothalamic/pituitary disorders (e.g., decreased gonadotropin secretion, effects of chronic diseases such as DM, CF, anorexia; excessive exercise; endocrine disease)
 - d. Androgen excess (e.g., polycystic ovaries, adrenal androgen excess [Cushing's])
 - e. Ovarian tumors
 - 2. Secondary amenorrhea; many causes same as primary amenorrhea
 - a. Pregnancy (most common)
 - b. Hypothalamic, pituitary, and adrenal disorders or tumors; chromosomal abnormalities (e.g., Turner's); endocrinopathies; chronic illness, especially those causing severe weight loss or malnutrition; conditions affecting gonadal function
 - c. Pharmacologic agents (discontinuance of birth control pills, use of tranquilizers)
 - d. Significant emotional stress or strenuous exercise programs, especially with runners, ballet dancers, and gymnasts; major weight loss
 - e. Uterine dysfunction after abortion, infection, C-section
 - f. Hysterectomy

- Signs and Symptoms
 1. Primary amenorrhea—no history of menses in adolescence; may have symptoms of marked psychosocial stress, adrenal dysfunction or gonadal disease, pituitary or hypothalamic disease, chronic illness including eating disorders, chromosomal abnormalities, pregnancy; may have cyclic abdominal pain without menstruation in pseudoamenorrhea
 2. Secondary amenorrhea—sudden or gradual cessation of menses, symptoms vary depending on underlying etiology, also caused by excessive exercise, athletes have high rates of amenorrhea as well as disordered eating
- Differential Diagnosis
 1. Determine if underlying etiology due to chronic illness, CNS disease, anorexia nervosa, inflammatory bowel disease, diabetes, pituitary adenoma, or thyroid dysfunction
 2. Distinguish primary amenorrhea due to constitutional or familial etiology from pregnancy
 3. Distinguish secondary amenorrhea due to pregnancy (most common cause), underlying disease or disorder
 4. Determine amenorrhea vs. “pseudoamenorrhea” (menstruation occurs but obstruction prevents release of menstrual blood)

- Physical Findings
 1. May have normal physical examination or signs of chronic, systemic illness or syndromes (e.g., underweight, CNS tumor or dysfunction, endocrinopathies, autoimmune disease, anorexia, malnourishment, unusually tall or short stature); may show signs of pregnancy
 2. May have lack of development of secondary characteristics or normal sexual development
 3. Pelvic examination may show pregnancy, reproductive system abnormalities (e.g., cervical atresia, imperforate hymen)
- Diagnostic Tests/Findings
 1. Pregnancy test
 2. Careful family history to rule out constitutional/familial delay, then consultation with physician and/or referral to specialists as needed
- Management/Treatment
 1. Constitutional/familial primary amenorrhea—education, reassurance, monitoring
 2. Amenorrhea associated with other etiologies requires further evaluation, physician consultation or referral to appropriate specialist (e.g., reproductive endocrinologist, surgeon, pediatric neurologist, obstetrician, psychologist)
 3. Treatment directed at management or correction of underlying cause of abnormal menstrual processes (e.g., surgery if imperforate hymen, adrenal tumors)
 4. Sensitivity to significant concern of delayed development by child and family very important
 5. Parent and child education regarding amenorrhea and adherence to any treatment regimen
 6. Genetic counseling may be indicated if genetic etiology

Obesity

- Definition based on:
 1. Degree of somatic overweight that causes detrimental health consequences
 2. Practical definition as a statistical magnitude of overweight for a population, with variance related to race, ethnicity, and socioeconomics
 3. World Health Organization—four subgroups for adults
 - a. Overweight: Body Mass Index (BMI) = 25–30
 - b. Grade 1—moderately obese: BMI = 30–35
 - c. Grade 2—severely obese: BMI = 35–40
 - d. Grade 3—morbidly obese: BMI = > 40
 4. In children, BMI normal curves for age (BMI Curves, 2001) www.cdc.gov/growthcharts
 - a. Overweight: above 85th percentile

- b. Obese: above 95th percentile
- c. Majority of obesity in adulthood has origins in childhood

- Etiology/Incidence
 1. Current prevalence ranges 5% to 10% in some African and Asian countries to 75% in urban Samoa
 2. Epidemic of childhood obesity is occurring at earlier age; childrens' BMI greater than 95th percentile
 - a. 10.4% of 2 to 5 year olds
 - b. 15.3% of 6 to 11 year olds
 - c. 15.5% of 12 to 18 year olds (1999–2000 NHANES data)
 3. Obesity has overtaken AIDS and malnutrition as the number one public health problem
 4. Other factors associated with obesity include genetics (leptin deficiency/leptin receptor deficiency), stress and increased cortisol, sleep deprivation, and inactivity (especially television viewing)
- Signs and Symptoms
 1. Obesity is associated with subclinical chronic inflammation, increased insulin resistance, early atherosclerosis, endothelial dysfunction, nonalcoholic fatty liver disease, polycystic ovary syndrome, infertility in older adolescents and adult women (due to excessive androgen production); hypercapnia associated with obstructive sleep apnea (OSA) can lead to delayed puberty; decreased growth hormone secretion (GH)
 2. Obese children and adolescents have hypertension (10%–30%), elevated total cholesterol, triglycerides, and low-density lipoprotein; also decreased levels of high-density lipoprotein
 3. Significant association between excess weight and asthma; also orthopedic complications (slipped capital femoral epiphysis, Legg–Calvé–Perthes disease); and possible nutritional deficits, especially iron
- Differential Diagnosis
 1. Comorbidities include hyperlipidemia, hypertension, hepatic stenosis, polycystic ovary syndrome, insulin resistance, and type 2 diabetes
 2. Metabolic syndrome—definitions vary
 - a. World Health Organization—antecedent is insulin resistance
 - b. National Cholesterol Education Program—elevated blood pressure, high triglyceride level, low HDL cholesterol level, high fasting glucose, and central obesity (3 of 5 must be met)

- Physical Findings
 1. History—birthweight, parents' BMI, gestational diabetes, prematurity, breastfeeding, neonatal complications (especially CNS)
 2. Dietary history, pubertal status, affect/school performance
 3. Review of medications (esp. atypical antipsychotics), pain, snoring, and stress level
 4. Evaluate linear growth, waist circumference, acanthosis nigricans, fundoscopic exam (rule out pseudotumor cerebri), liver enlargement, hirsutism
- Diagnostic Tests/Findings
 1. Labs—liver functions, fasting lipid panel, fasting glucose and insulin level, HbA_{1c}; also specific studies as indicated, e.g., sleep study, x-ray hips/knees, patients with developmental delays, karyotype, and MRI; OGTT or hormonal assessment may be necessary
 2. Severe obesity in toddler may require genetics testing and leptin levels
 3. Measurements: growth velocity, body mass index, body fat measurement
- Management/Treatment
 1. Lifestyle modification—weight maintenance is recommended for most children
 - a. Dietary intervention—elimination of sugar-containing beverages and change to low-glycemic-load diet
 - b. Physical activity intervention—minimum 30 minutes vigorous exercise 5 days per week; decrease sedentary behaviors (TV, hand-held games) and increase physical education and after-school programs
 - c. School intervention—model health promotion ideals and encourage in-school vigorous activity
 - d. Family intervention—usually child is not the only obese member of the family; promote parenting skills that include healthy dietary choices and avoiding food as a reward
 2. Pharmacotherapy
 - a. Few approved for children—potential for side effects
 - b. Drug for reduction of energy intake—sibutramine (< 16 years)
 - c. Drug for reduction of energy absorption—orlistat (< 12 years)
 - d. Drug for improvement of T2D/insulin resistance—metformin (< 10 years)
 3. Surgery
 - a. Bariatric surgery—for extreme cases with strict criteria

- b. Adjustable gastric banding—not approved for use in adolescents
 - c. Roux-en-Y gastric bypass—safer when candidates carefully selected, surgeon has advanced laparoscopic skills; common complications include iron deficiency, folate deficiency, B₁₂ deficiency, and event requiring surgical intervention
4. Prevention is critical in limiting the epidemic

■ QUESTIONS

Select the best answer

1. Secondary hypothyroidism results from:
 - a. Excess release of thyroid hormone beyond the newborn period
 - b. Intrauterine exposure to thyrotoxic drugs
 - c. Disease or disorder of the thyroid gland itself
 - d. Disease or disorder of the hypothalamus or pituitary gland compromising thyroid function
2. Congenital hypothyroidism has a higher incidence in which of the following populations?
 - a. African-Americans
 - b. Hispanic and Native Americans
 - c. Asian-Americans
 - d. Euro-Americans
3. Which of the following is not a sign or symptom of congenital hypothyroidism?
 - a. Hoarse cry
 - b. Frequent stooling
 - c. Coarse features
 - d. Lethargy
4. The most common cause of hyperthyroidism in children and adolescents is:
 - a. Graves' (autoimmune) disease
 - b. Thyroid cancer
 - c. Thyroid nodules
 - d. Pituitary tumor
5. Which of the following is not found in an adolescent with untreated Graves' disease?
 - a. Behavioral problems
 - b. Sleep disturbances
 - c. Tendency to gain weight easily
 - d. Tachycardia
6. In which one of the following children would you most suspect hyperthyroidism?
 - a. A 16-year-old male who complains about restlessness
 - b. A 14-year-old adolescent female who is heat intolerant and has amenorrhea

- c. A male preteen with behavior problems
d. A six-year-old female who complains of tiredness
7. The most common thyroiditis is:
- Subacute thyroiditis caused by a viral infection of the gland
 - Acute suppurative thyroiditis caused by bacterial infection
 - Caused by exposure to radiation or trauma
 - Hashimoto's or chronic autoimmune thyroiditis
8. Nephrogenic, or vasopressin-resistant, diabetes insipidus:
- Is caused by anatomic defects in the brain causing hypofunction of the pituitary or hypothalamus
 - Results from damage to the hypothalamus or pituitary from surgical trauma or infection
 - Is caused by reduced renal responsiveness to antidiuretic hormone (ADH)
 - Has oliguria as a primary presenting symptom
9. An infant with polydipsia, polyuria, irritability, and failure to thrive, should be evaluated for:
- Diabetes insipidus
 - Homocystinuria
 - Growth hormone deficiency
 - Hyperglycemia
10. Which one of the following is not characteristic of constitutional growth delay?
- There is generally no history of a similar growth pattern in other family members
 - The child usually remains constitutionally small as an adult
 - Final adult stature tends to be normal
 - Weight and height at birth are generally in the lower percentiles
11. A newborn or infant with birth length < 50% and microphallus should be suspected of having:
- Growth hormone deficiency
 - Congenital hypothyroidism
 - Primordial short stature
 - Down syndrome
12. An adolescent male who fails to develop secondary sex characteristics at puberty and who has small, underdeveloped testes should be suspected of having:
- Adrenal hyperplasia
 - Klinefelter's syndrome
 - Marfan syndrome
 - Cerebral gigantism (Sotos syndrome)
13. Individuals with chronic adrenal insufficiency often have:
- Frequent otitis media
 - High energy levels
 - Love for physical activity
 - A craving for salt
14. In the newborn period, infants of diabetic mothers (IDMs) are particularly at risk for:
- Small size for gestational age
 - Intrauterine growth retardation (IUGR)
 - Disorders in bone development
 - Hypoglycemia
15. Which statement is true about true (complete) precocity or incomplete (pseudoprecocity)?
- True precocity occurs because of hormonal stimulation from the pituitary or hypothalamus causing gonadal maturation and fertility
 - Pseudoprecocity does not involve development of any secondary sex characteristics
 - Incomplete precocity is caused by adrenal or gonadal tumor or dysfunction and results in increased linear growth but no development of secondary sex characteristics
 - Incomplete or pseudoprecocity leads to testicular enlargement and ovulation
16. An adolescent who has tall stature, increased arm span, arachnodactyly, laxity of joints, pectus excavatum, and an abnormal echocardiogram would be suspected of having:
- Turner's
 - Beckwith-Wiedemann syndrome
 - Marfan
 - Klinefelter's syndrome
17. Which one of the following is not found in children with growth hormone excess?
- Tall stature
 - Prominent mandible and supraorbital ridge
 - High or normal plasma growth hormone
 - Short stature
18. A pathognomonic skin finding in children with chronic adrenal insufficiency (Addison's) is:
- Purple striae
 - Increased pigmentation in the axilla, groin, areola, hand creases, and in surgical scars
 - Dry, thickened skin
 - Increased perspiration

19. Which of the following findings is not characteristic of children and infants with hyperadrenocorticism?
- Advanced skeletal maturation
 - "Moon" facies
 - Delayed onset of secondary sex characteristics
 - "Buffalo type" adiposity of face, neck, and trunk
20. Transient neonatal hypoglycemia is:
- Most common in AGA infants
 - Low in premature SGA infants
 - Most common in LGA infants
 - Least common in LGA infants
21. Regular insulin:
- Has a quicker onset of effect and longer duration than NPH
 - Has a slower onset of effect and shorter duration than NPH
 - Has a quicker onset of effect and shorter duration than NPH
 - Has the longest duration of the insulins available
22. The preferred name now for insulin-dependent diabetes mellitus (IDDM) is:
- Maturity-onset diabetes
 - Type 1 diabetes
 - Type 2 diabetes
 - Insulin resistance syndrome
23. Blood glucose levels of children 5 to 11 years with diabetes should be maintained between:
- 70 to 180 mg/dL
 - 100 to 200 mg/dL
 - 60 to 80 mg/dL
 - Slightly over 200 mg/dL
24. Blood glucose levels of younger children with diabetes are maintained at slightly higher levels than blood glucose levels of older children because:
- Children have a greater need for available glucose in the blood system
 - Younger children tend to be more active
 - Younger children become more irritable than do older children
 - Lowering the risk of hypoglycemia in younger children is particularly important in order to avoid the potential for hypoglycemia with consequent neurological system damage
25. Glucagon should be used to treat:
- Children with mild hypoglycemia
 - Children with moderate hypoglycemia
 - Children with severe hyperglycemia
 - Children with severe hypoglycemia
26. Which finding is not a sign or symptom of diabetes onset in children?
- Alopecia
 - Glycosuria
 - Polydipsia
 - Polyuria
27. Abdominal pain and vomiting are particularly critical to monitor in children with diabetes because these findings may represent the onset of:
- Ketoacidosis
 - Gastrointestinal infection
 - Hyperglycemia
 - Autoimmune response to the pancreas
28. Which of the following statements is not true about type 1 diabetes?
- The "honeymoon" period postdiagnosis is of variable duration
 - Diabetes is a relatively common disease in childhood
 - Children with type 1 diabetes can switch to oral insulin agents once they reach adulthood
 - Three factors influence a child's potential to develop diabetes—genetic predisposition, autoimmune response, and exposure to viral or chemical agents
29. Precocious pubertal development is defined as the development of secondary sexual characteristics in boys before age __ years and menses in girls before the age of __ years.
- 10 years; 10 years
 - 6 years; 8 years
 - 9 years; 9.5 years
 - 6 years; 9 years
30. In boys, lack of secondary sexual characteristics after 17 years suggests:
- Castration
 - Abnormal testicular function
 - True hermaphroditism
 - Pituitary adenoma

31. The peak incidence for adolescent gynecomastia occurs at age:
- 10 years
 - 13 to 14 years
 - 16 years
 - 18 years
32. The most common cause of primary amenorrhea is:
- Obstructions of menstrual flow
 - Primary ovarian insufficiency
 - Secondary ovarian insufficiency
 - Constitutional or familial

■ ANSWERS

- | | |
|--------------|--------------|
| 1. d | 17. d |
| 2. b | 18. b |
| 3. b | 19. a |
| 4. a | 20. c |
| 5. c | 21. c |
| 6. b | 22. b |
| 7. d | 23. a |
| 8. c | 24. d |
| 9. a | 25. d |
| 10. c | 26. a |
| 11. a | 27. a |
| 12. b | 28. c |
| 13. d | 29. c |
| 14. d | 30. b |
| 15. a | 31. b |
| 16. c | 32. d |

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▣ EDUCATIONAL WEB SITES

- www.hormone.org (The Hormone Foundation)
- www.pituitary.org (Pituitary Network Association)
- www.magicfoundation.org (all endocrine disorders)
- www.caresfoundation.org (congenital adrenal hyperplasia)
- www.pens.org (Pediatric Endocrinology Nursing Society)
- www.childrenwithdiabetes.com (pediatric diabetes)
- www.diabetes.org (American Diabetes Association—all diabetes)
- www.diabetes.org/schooltraining (ADA's training curriculum for schools)
- www.jdrf.org (Juvenile Diabetes Research Foundation)
- www.dsdguidelines.org/files/clinical.pdf (Intersex Society of North America "Scripts for talking with parents")
- www.hgfound.org (Human Growth Foundation)

Multisystem and Genetic Disorders

Rita Marie John

■ INTRODUCTION

While the disorders in this chapter are more rare, care of the patient needs to be coordinated, comprehensive, continuous, culturally sensitive, and multidisciplinary, with quality and safety involved. The concept of the medical home model in primary care provides the family with comprehensive care that coordinates community agencies and personnel. While the medical societies do not see that a pediatric nurse practitioner (PNP) can head the medical home, NAPNAP has issued a position statement that clearly states that PNPs can be the head of the medical home. Care concepts revolving around family-centered care, community agencies, as well as providing transitional care are key to the medical home (American Academy of Pediatrics [AAP], 2008; Duderstadt, 2008). It is important to coordinate specialty care. Knowledge about each condition and its management is rapidly expanding, and personnel at such centers are most current in this essential practice foundation.

The PNP, however, remains responsible for assisting the child and family with all primary care issues, some of which will be disorder specific. The PNP also is a central contact for the child and family for issues of care coordination and community-based interventions, advocacy, assistance with stress management, and promotion of positive adaptation and coping. PNPs working with children who have any of these disorders must become familiar with the issues commonly confronting such children and their families, legislation protecting the rights of individuals who have chronic illness or disability, and support organizations and agencies that can be helpful to caregivers and families.

There are several resources available for children with developmental disabilities. There are now 67 University Centers for Excellence in Developmental Disabilities (UCEDD), funded by the Administration on Developmental Disabilities (ADD). In addition, there are 38 Leadership Education in Neurodevelopmental Disabilities (LEND) Programs (funded by the Maternal and Child Health Bureau [MCHB]) and 21 Intellectual and Developmental Disability Research Centers (IDDRC). The latter are primarily funded by the Eunice Kennedy Shriver National Institute for Child Health and Development (NICHD). Information about these programs can be found at <http://www.aucd.org/template/page.cfm?id=1>. The UCEDD have federal partners including the Administration on Developmental Disabilities, MCHB, National Center on Birth Defects and Developmental Disabilities (NCBDDD) at the Centers for Disease Control and Prevention (CDC), and NICHD.

In addition, collaborating partners include Association of Maternal Child Health Programs, Corporation for National and Community Service, External Partners Group, National Leadership Consortium on Developmental Disabilities, and the National Service Inclusion Project (NSIP). All of these organizations have web sites that can be accessed via a search engine. The Alliance of Genetic Support Groups, (202) 966-5557 and <http://www.geneticalliance.org> is a comprehensive resource for conditions with a genetic basis. In addition, the National Association for Rare Disorders can also be a source of information for parents and providers looking to find out more information about rare disorders.

Where possible, information about disorder-specific support organizations is included in the management sections of this chapter. Exceptional Parent magazine,

416 Main Street, Johnstown, PA 15901, 800-EPARENT, (1-800-372-7368, ext. 203), <http://www.eparent.com> provides a wealth of information for parents of children who have various special healthcare needs. The March of Dimes Birth Defects Foundation, 1275 Mamaroneck Avenue, White Plains, NY 10605, (888) 663-4637 www.marchofdimes.com provides information about congenital diseases and disorders, and the National Information Center for Children and Youth with Disabilities (NICHCY), 1825 Connecticut Ave NW, Suite 700, Washington, DC 20009, www.nichcy.org provides information on all disabling conditions.

■ CONGENITAL DISEASES

Acquired Immune Deficiency Syndrome (AIDS)

- Definition: Advanced stage of illness in individuals infected with the Human Immunodeficiency Virus (HIV); CDC case definition of HIV classifies children according to presence or absence of clinical signs and symptoms and according to status of immune function and clinical findings
- Etiology/Incidence
 1. Each year approximately 6000 HIV infected women with HIV-1 give birth; preventive strategies reduce the risk of mother-to-child transmission to the infant to 1% to 2%
 2. If there is no antiretroviral (ARV) therapy given as many as 30% to 40% of infants can be identified by 48 hours of age
 3. Infants with positive virologic tests results at or before 48 hours are considered to have been infected in utero, but infants having a negative virologic tests during the first week and have subsequent positive test are considered to have intrapartum infection
 4. Cord blood testing is not recommended
- Prevention Strategies
 1. Administration ARV prophylaxis during pregnancy and labor and to the infant for 6 weeks after birth
 - a. Regimes consist of 3 ARV during pregnancy and labor
 - b. Intravenous zidovudine (AZT) is given to all pregnant women with HIV-1 infection during labor even if they receive the above combination
 2. Elective cesarean delivery at 38 weeks before the onset of labor and before the rupture of membranes for women with a viral load > 1000 copies per ml or who have unknown viral load even if they have received 3 ARV
 3. Complete avoidance of breastfeeding
- Care of the HIV-1 Exposed Infant
 1. Review history for possible maternal coinfections including tuberculosis, syphilis, toxoplasmosis, hepatitis B or C, cytomegalovirus, or herpes simplex
 2. Evaluate the infant for signs of coinfection
 3. Specific testing is not recommended unless suggestive by history or physical assessment
 4. HIV-1 DNA PCR or RNA assay at 14 to 21 days and if negative, they should be repeated at 1 to 2 months and again at 4 to 6 months to identify or exclude HIV infection as soon as possible; any positive result at any age is repeated to confirm results
 5. No NAAT is needed at 8 weeks if test results at 2 and 4 weeks are negative
 6. A single negative test at 8 weeks identifies a presumptively uninfected infant
 7. PCP prophylaxis is started at 4 to 6 weeks if the results are indeterminate
- Signs and Symptoms: Median onset for infants with perinatal infection is 12 to 18 months; HIV, however, can be latent for years
 1. Prematurity
 2. Low birthweight
 3. Recurrent serious bacterial or viral infections—especially oral thrush
 4. Failure to thrive
 5. Recurrent fevers
 6. Chronic diarrhea
 7. Diminished activity
 8. Developmental delays
- Differential Diagnosis
 1. Infectious disease or other associated conditions without underlying HIV
 2. Other immune deficiencies of infancy
- Physical Findings
 1. Falling ratio of head circumference to height and weight, due to encephalopathic direct effect on brain growth
 2. Lymphadenopathy— ≥ 0.5 cm at more than 2 sites; bilateral at one site
 3. Hepatosplenomegaly
 4. Parotitis, nephropathy, CNS disease
- Diagnostic Tests/Findings
 1. Detection of HIV—using HIV-1 NAAT (HIV-1 DNA PCR or RNA assay)
 2. For nonbreastfeeding infants, definitive exclusion on HIV-1
 - a. At least 2 negative HIV-1 or RNA or DNA virological test results, one obtained ≥ 1 month and another ≥ 4 months (HIV

- RNA may be preferred in infants born to mother infected outside of North America)
- b. Or two negative HIV-1 tests \geq 6 months of age and no other laboratory or clinical evidence of HIV-1 infection or no AIDS defining conditions
- 3. Cases of HIV meeting AIDS criteria are reportable

- Management/Treatment

1. Risk reduction through prevention of maternal infection, assuring clean blood and tissue supplies, prevention of STD and needle sharing
2. Reduction of risk of perinatal HIV—through treatment of pregnant mother and perinatal chemoprophylaxis of mother and infant
3. At birth, history, physical, and risk assessment, continue or start ARV prophylaxis
4. If mother's HIV-1 serostatus is unknown at the time of labor or birth, do a rapid HIV-1 antibody testing on mother or newborn with consent
5. At 14 days, do HIV-1 NAAT, repeat at 4 weeks if negative
6. At six weeks, stop ARV prophylaxis
7. At 8 weeks, do HIV NAAT if not done earlier
8. Treatment with combination ARV treatment for all HIV-1 infected infants younger than 12 months regardless of symptoms, immunologic, or virologic measures (Havens, 2009)
9. At 12 to 18 months, enzyme immunoassay for antibody to HIV-1
10. PCP prophylaxis in infants
11. Monitoring of CD4⁺ lymphocyte count and percentage, CBC, differential, platelet count
12. Antiretroviral therapy in consultation with HIV management specialists
13. Immunize and TB screening according to guidelines
14. Treatment of infections and other associated conditions
15. Early intervention for developmental delay
16. Maximum supportive care for child and family
17. CDC National Prevention Information Network
P.O. Box 6003
Rockville MD 20849-6003
1 (800) 458-5231
<http://www.cdcnpin.org>

Chlamydia Trachomatis (Refer to Gu/Gyn chapter)

- Definition: Chlamydiaceae divides into two genera, Chlamydophila and Chlamydia; Chlamydophila includes *C. pneumonia*, *C. psittaci*, and nonhuman pathogens; Chlamydia includes *C. trachomatis* and nonhuman pathogens; *C. trachomatis* is an obligate intracellular bacteria that has an infectious elementary body and a metabolically active reticulate form
- Etiology/Incidence
 1. Chlamydia causes neonatal conjunctivitis, trachoma, pneumonia in young infants, genital tract infections, and lymphogranuloma venereum
 2. *C. Trachomatis* has more than 18 variants
 3. *C. Trachomatis* is the most frequent cause of neonatal conjunctivitis
 4. 25% to 50% of perinatally infected infants develop conjunctivitis
 5. Transmission occurs from cervical maternal infection to infant during vaginal delivery 50% of the time
 6. Prevalence among sexually active women about 5%; among adolescents may be as high as 20%
 7. Chlamydia trachomatis is the most prevalent sexually transmitted disease in the U.S., with highest rate from 14 to 24 years
 8. Incidence of neonatal pneumonia if mother is infected is approximately 5% to 30%
- Signs and Symptoms: Chlamydia trachomatis in the neonate
 1. Neonatal conjunctivitis—mild to severe
 - a. Typically develop 5 to 14 days after birth and lasts for longer than 2 weeks
 - b. Conjunctival edema
 - c. Conjunctival injection
 - d. Watery to mucopurulent eye discharge
 - e. Pseudomembrane with bloody discharge if prolonged
 - f. Routine prophylactic ophthalmic drops of silver nitrate, erythromycin, and tetracycline is not effective against *C. trachomatis*
 2. Neonatal pneumonia
 - a. 2 to 19 weeks of age
 - b. Preceding signs include rhinorrhea, congestion, and conjunctivitis
 - c. Tachypnea with persistent “staccato” cough with congestion
 - d. Rales and rarely wheezing
 - e. Preterm infants can have apnea

- Differential Diagnosis
 1. A different infectious agent, *Staphylococcus aureus*, *Haemophilus influenzae*, streptococcus, herpes simplex, enteric bacteria
 2. Chemical reaction to silver nitrate
 3. Bronchitis/pneumonia—respiratory syncytial virus, other viral or bacterial infections
- Physical Findings
 1. Neonatal conjunctivitis
 - a. Injected conjunctiva with exudate within first weeks of life
 - b. Pseudomembrane
 - c. Friable conjunctiva
 2. Neonatal pneumonia
 - a. Afebrile
 - b. Tachypnea
 - c. Rales, but rarely wheezes
- Diagnostic Tests/Findings
 1. Chest radiograph shows hyperinflation and infiltrates but usually there is no lobar consolidations or pleural effusion
 2. Peripheral eosinophilia (> 300 cells/mL)
 3. Elevated serum immunoglobulins
 4. Gold standard is culture and is approved by FDA for use at all sites/culture; the specimen must contain epithelial cells since *Chlamydia* are intracellular; highly specific (98% to 100%) and sensitive (Chandran & Boykan, 2009)
 5. Nucleic acid amplification tests (NAAT) tests are highly sensitive and specific ($> 95\%$) and have a rapid turnover; have potential for false positive results
 6. Enzyme immunoassays (EIA) have a specificity and sensitivity of 60% to 70%
 - a. Perinatally acquired vaginal and rectal infection can be present for up to 18 months; after this, sexual abuse needs to be considered
 - b. Lower genital tract infection is generally asymptomatic
- Management/Treatment: Topical therapy is ineffective; infants less than 6 months, oral erythromycin suspension, 40 to 50 mg/kg/day for 10 to 14 days; approximately 20% of infants need second course of treatment; erythromycin is associated with increased risk of pyloric stenosis

Gonococcal Infection (Refer to Gu/Gyn chapter)

- Definition: Infections caused by acquisition of *Neisseria gonorrhoeae* (GC)

- Etiology/Incidence
 1. Maternal transmission to neonate from contact with vaginal secretions during birth
 2. Second most common sexually transmitted infection with 15- to 19- and 20- to 24-year-old women highest rates of gonorrhea in 2007
 3. Screen for chlamydia if positive for GC
- Signs and Symptoms
 1. Infections in newborn infant usually involves the eye
 - a. Ophthalmia; injected
 - b. Swollen conjunctiva with exudate
 2. Other types of infection include scalp abscess, disseminated disease with bacteremia, arthritis, or meningitis
 - a. Fever
 - b. Irritability
 - c. Rapid respiration
 - d. Rapid heart rate
- Differential Diagnosis
 1. Conjunctivitis
 - a. Chemical reaction to silver nitrate
 - b. Infections due to *chlamydia*, *Staphylococcus aureus*, *Haemophilus influenzae*, streptococcus, herpes simplex, or enteric bacteria
 2. Bronchitis/pneumonia—respiratory syncytial virus (RSV), other viral, bacterial causes
 3. Skin, oropharyngeal, arthritic, vaginal, urethral, rectal, systemic infection
 4. Other causative organisms
 5. Allergy
- Physical Findings
 1. Gonococcal ophthalmia neonatorum—conjunctivitis, appearing 2 to 5 days after birth, occasionally up to 25 days
 - a. Thin, clear discharge from eye that becomes thick, mucoid, sometimes bloody
 - b. Edema of lids, conjunctiva
 - c. Untreated, may progress to edema and ulceration of cornea, globe perforation and blindness, and/or may become systemic infection
 2. Other—scalp abscesses, disseminated infection with signs of sepsis, meningitis, or pain, and decrease in motion if septic arthritis
- Diagnostic Tests/Findings
 1. Gram stain at initial evaluation—microscopic identification of gram-negative intracellular diplococci in smears of exudate followed by laboratory-based testing

2. Culture is preferred—swab from orifice or blood, synovial fluid from affected joint or cerebrospinal fluid specimen
 3. Verify initial positive test (NAAT tests are not approved for prepubertal children)
- Management/Treatment: Follow current CDC guidelines
 1. Ophthalmia neonatorum prophylaxis—silver nitrate 1% aqueous; erythromycin ophthalmic ointment 0.5%; tetracycline ophthalmic ointment 1%
 2. Hospitalize infected neonates and evaluate for disseminated disease
 - a. Ceftriaxone 25 to 50 mg/kg IV or IM in a single dose, not to exceed 125 mg if just GC conjunctivitis
 - b. Cefotaxime is recommended for infants with hyperbilirubinemia
 3. Neonatal disseminated infection
 - a. Ceftriaxone 25 to 50 mg/kg/day IV or IM in a single daily dose for 7 days, with a duration of 10 to 14 days, if meningitis is documented OR
 - b. Cefotaxime 25 mg/kg IV or IM every 12 hours for 7 days, with a duration of 10 to 14 days, if meningitis is documented

Congenital Syphilis (Refer to Gu/Gyn chapter)

- Definition: An infection caused by *Treponema pallidum* that can be acquired in adolescents and congenitally in infants; congenital infection occurs by transplacental transmission of *T. pallidum*, a spirochete; facilitates infection with HIV
- Etiology/Incidence
 1. Rates of congenital syphilis in U.S. for past ten years range from 580 cases to 339; during the early 90s, the rates were as high as 4424 (1991); more common in South/rural areas
 2. *Treponema pallidum*—mother with history of syphilis whose treatment is not documented as complete with full follow-up
 3. Transmission is more common when a mother has primary and secondary infection, rather than latent infection
 4. Maternal risk factors are lack of prenatal care and cocaine abuse
 5. Can be contracted at any stage of pregnancy; intrauterine infection can result in fetal death, stillbirth, prematurity, or clinical congenital disease
- Signs and Symptoms
 1. 60% of infants with congenital infection have no symptoms in the first week of life, but will develop symptoms if not immediately treated; initial presentation up to 2 years of age
- 2. Early congenital syphilis—liver, hematologic, mucous membrane, musculoskeletal, CNS, eye, renal, GI
 - a. Low birthweight/prematurity
 - b. Rhinitis (snuffles), mucous patches
 - c. Jaundice with elevated liver enzymes
 - d. Lymphadenopathy with Coombs-negative hemolytic anemia
 - e. Osteochondritis which causes resistance to movement (pseudoparalysis of Parrot)
 - f. Rash similar to secondary syphilis with desquamation of hands/feet
 - g. CNS abnormalities
- 3. Late congenital syphilis
 - a. Symptoms result from chronic inflammation of CNS, bone, teeth
 - b. Bony changes with anterior tibial bowing, prominence of forehead
 - c. Dental changes with Hutchinsonian (peg-shaped) teeth with lower molars with excessive cusps, enamel defects
- Differential Diagnosis
 1. Any of the “TORCH” congenital conditions
 2. Rash stage—pityriasis rosea, scabies, tinea
- Physical Findings
 1. May be asymptomatic at birth
 2. Nonimmune hydrops
 3. Hepatomegaly
 4. Splenomegaly
 5. Lymphadenopathy
 6. Rhinitis (snuffles)
 7. Pseudoparalysis of Parrot
- Diagnostic Tests/Findings
 1. Tests for syphilis
 - a. Direct visualization of spirochete by dark field microscopy; direct fluorescent antibody test for *Treponema pallidum*
 - b. Nontreponemal tests—Venereal Disease Research Laboratory microscopic slide test (VDRL); rapid plasma reagin (RPR)
 - c. Treponemal test—fluorescent treponemal antibody absorbed; microhemagglutination assay for antibody to *Treponema pallidum* (TP-PA or FTA-ABS)
 2. If the mother has a reactive nontreponemal and treponemal test, the infant's cord blood results should be evaluated with a quantitative nontreponemal serological test (RPR or VDRL) since umbilical cord blood could be contaminated with mother's blood and lead to a false positive; a treponemal test is not necessary
 3. Elevated LFT; positive VDRL in CSF

4. Coombs negative hemolytic anemia, thrombocytopenia
 5. Radiographic changes (“moth eaten” metaphysis)
- Management/Treatment
 1. Prenatal screen all pregnant women; treat all infected individuals, especially sexually active women and partners
 2. Treat infants with proven or highly probable disease and an abnormal physical examination that is consistent with congenital syphilis, a serum quantitative nontreponemal serologic titer that is fourfold higher than the mother’s titer, or has a positive dark field or fluorescent antibody test of body fluid with:
 - a. Aqueous crystalline penicillin G 100,000–150,000 units/kg/day, administered as 50,000 units/kg/dose IV every 12 hours during the first 7 days of life and every 8 hours thereafter for a total of 10 days **OR**
 - b. Procaine penicillin G 50,000 units/kg/dose IM in a single daily dose for 10 days
 - c. Follow-up to determine appropriately falling titers of the serologic testing (nontreponemal test) every 2 to 3 months until there is a decrease fourfold in test or the test becomes nonreactive
 3. For children with reactive serologic tests for syphilis identified after one month, the provider should review maternal serology to determine if the child has congenital or acquired syphilis; treatment consists of:
 - a. Aqueous crystalline penicillin G 200,000 to 300,000 units/kg/day IV, administered as 50,000 units/kg every 4 to 6 hours for 10 days
 - b. Follow-up as above for infected neonates

Herpes (Refer to Gu/Gyn chapter)

- Definition: Group of infectious diseases caused by the herpes simplex virus (HSV) type 1 or 2; HSV-1 is associated with infections of EENT and central nervous system and HSV-2 is associated with anogenital infections; establishes viral latency in sensory ganglia following primary infection
- Etiology/Incidence
 1. More than 80 herpesvirus, 8 infect humans—HSV-1, HSV-2, varicella-zoster virus (VZV), human herpesvirus (HHV-6 & HHV-7), Epstein-Barr virus, cytomegalovirus (CMV) and Kaposi’s sarcoma-associated herpesvirus (HHV-8)
 2. Transmitted through exposure to mucous membranes or skin with active lesion through

direct contact of virus with host mucous membrane or abraded skin

3. HSV can be transmitted to neonatal with incidence from 1:3000 to 1:20,000 live births with 1500–2200 cases in U.S. annually
- Signs and Symptoms
 1. Congenital in utero infection
 - a. Often die in utero
 - b. If survive to term, vesicular lesions, chorioretinitis, microphthalmia, microcephaly, with abnormal CNS
 2. Neonatal skin-eye-mouth (SEM) disease
 - a. Usually presents in first two weeks of life
 - b. Cutaneous lesions generally located on scalp, mouth, nose and eye with the baby contracted from the mother’s genitals
 3. Neonatal CNS disease (60% of cases)
 - a. Infant in second to third week of life
 - b. Neurological signs such as seizures, apneic episodes
 - c. Permanent neurological disability in 40% of survivors
 4. Neonatal disseminated disease
 - a. Mortality rate of 50%
 - b. Acutely ill infant during first week of life
 - c. Multisystem involvement—shock, disseminated intravascular coagulation, and multiorgan failure
 - d. Unresponsive to antibiotic therapy
 - Differential Diagnosis
 1. Bacterial sepsis such as group B streptococcus and viral infections—enterovirus, varicella, influenza A/B, parainfluenza, and adenovirus
 2. Langerhans cell histiocytosis and incontinentia pigmenti can present with vesicles
 - Physical Findings
 1. Neonatal skin-eye-mouth (SEM) disease
 - a. Vesicles with erythematous base, clear or cloudy fluid, sometimes appearing pustular, limited to skin, eyes, and mouth
 - b. Transient fever, malaise (low acuity)
 2. Neonatal CNS disease
 - a. Herpetic skin lesions in 60% of cases
 - b. Cranial nerve abnormalities
 - c. Seizures
 3. Neonatal disseminated disease
 - a. Septic picture—seizures, lethargy
 - b. Fever, tachypnea, labored breathing with onset of HSV pneumonitis
 - c. Hepatomegaly
 - Diagnostic Tests/Findings
 1. Virus isolation from skin, mucous membranes, urine, blood, stool, or CSF

2. Detection of fluorescent antibodies (especially when skin lesions are active)
 3. Polymerase chain reaction (PCR)—helpful in CNS infection
 4. Serology rarely useful in acute HSV infection
 5. Increased CSF protein and pleocytosis
 6. Pulmonary radiograph reveals bilateral, patchy infiltrates from 3 to 10 days of neonatal disseminated disease
 7. Abnormal liver function test results
- Management/Treatment
 1. Infant with known or suspected herpes
 - a. Acyclovir 20 mg/kg body weight IV every 8 hours for 21 days for disseminated and CNS disease (60 mg/kg per day)
 - b. Acyclovir 20 mg/kg IV every 8 for 14 days for SEM (60 mg/kg per day)
 2. Infants with ocular involvement
 - a. 1% trifluridine or
 - b. 0.1% iododeoxyuridine, or
 - c. 3% vidarabine for ocular HSV or for neonatal HSV with SEM involvement

Cytomegalovirus (CMV)

- Definition: Congenital infection cytomegalovirus, a member of herpesvirus family can be symptomatic (10%) or asymptomatic (90%); CMV affects 1 out of 750 babies; asymptomatic infections are the most common but some congenitally infected infants who are asymptomatic at birth can have hearing loss or learning disability later in life
- Etiology/Incidence
 1. Most common intrauterine infection, affecting 1% of newborns
 2. Prevalence varies with age, socioeconomic status, ethnicity, and nationality, with increasing prevalence with age, coming from a developing nation, low socioeconomic group, and being African-American
 3. Risk of severe disease is greater if exposed to gestational primary infection however congenital infection can result from reactivation of latent CMV infection
 4. Preterm infants are at greater risk of symptomatic infection
 5. Vertical transmission to infant
 - a. In utero via placenta
 - b. At birth through passage
 - c. Postnatal infection by ingestion of CMV-positive human milk
- Signs and Symptoms
 1. Symptomatic infection at birth
 - a. Intrauterine growth retardation
 - b. Growth retardation
 - c. Jaundice
 - d. Purpura
 - e. Hepatosplenomegaly
 - f. Microcephaly
 - g. Retinitis
 2. Asymptomatic infection at birth
 - a. Sensorineural hearing loss is most common sequela (Enright & Prober, 2004) with 7.2% to 15% after 6 years of life; may be a greater role in sensorineural hearing loss in children than was originally suspected
 - b. Intellectual consequences with greater risk of school failure
- Differential Diagnosis
 1. Congenital toxoplasmosis
 2. Rubella
 3. Herpes
- Physical Findings: Classic congenital CMV manifestations are noted above
- Diagnostic Tests/Findings
 1. Isolation of CMV from infant urine, pharynx or peripheral blood leukocytes within first three weeks of life
 2. Other tests with variable efficacy
 - a. CMV-specific IGM
 - b. PCR testing of neonatal blood and cerebrospinal fluid
 - c. SMV antigenemia
- Management/Treatment
 1. Primary prevention in women of child-bearing age and younger, since both initial infection and reactivation during pregnancy can cause fetal infection
 - a. Of all the congenital infections, only 22% of mothers knew about this infection; education is needed
 2. Congenitally affected infants—refer to developmental disabilities center for interdisciplinary management of complex disabilities
 3. Asymptomatic infants with congenital CMV
 - a. Refer to audiology for periodic sensorineural hearing evaluation
 - b. Screen frequently for growth retardation and emerging developmental delays
 - c. Refer to ophthalmology—emerging chorioretinitis
 - d. Refer to dentistry—defective tooth enamel
 4. Neonatal acquisition—often presents as afebrile pneumonia after 8-week incubation period; test suspect infants
 5. Seek specialist consultation regarding use of ganciclovir

Congenital Rubella Syndrome (CRS) (Refer to Chapter 9)

- Definition: Multisystem involvement in a neonate associated with maternal infection with rubella virus; up to 85% of infants are affected if the infection is associated with a rash during the first 12 weeks, 54% of infants are affected if the infection occurs from 13 to 16 weeks of gestation, and 25% during the end of the second trimester
- Etiology/Incidence
 1. Maternal to fetal transmission of rubella virus
 2. Average of 5 to 6 cases per year of congenital rubella syndrome reported since 1980
- Signs and Symptoms
 1. Early manifestation of Rubella
 - a. Ophthalmologic—cataracts, pigmentary retinopathy, microphthalmos, congenital glaucoma
 - b. Neurologic—behavior disorders, meningoencephalitis, mental retardation
 - c. Cardiac—patent ductus, peripheral pulmonary artery stenosis
 - d. Auditory—deafness
 - e. Growth—failure to thrive, related to heart defects
 - f. To confirm diagnosis must have a symptom from one of each of the two categories:
 - (1) Cataracts, congenital glaucoma, congenital heart disease (most commonly patent ductus arteriosus or peripheral pulmonary artery stenosis), loss of hearing, pigmentary retinopathy
 - (2) Purpura, hepatosplenomegaly, jaundice, microcephaly, developmental delay, meningoencephalitis, radiolucent bone disease
 2. Delayed manifestation of Congenital Rubella Syndrome
 - a. Can be delayed from two to four years
 - b. Diabetes mellitus
 - c. Progressive encephalopathy resembling subacute sclerosing panencephalitis
 - d. Higher than expected incidence of autism
- Differential Diagnosis
 1. Congenital CMV
 2. Syphilis
 3. Toxoplasmosis
- Physical Findings
 1. Growth retardation
 2. Interstitial pneumonitis
 3. Hepatosplenomegaly
 4. Thrombocytopenia
 5. Blueberry muffin lesions or dermal erythropoiesis
 6. Cataracts, retinopathy, glaucoma
 7. Petechial or purpuric rash
 8. Sensorineural defects
 9. Microphthalmia
- Diagnostic Tests/Findings
 1. Reliable evidence of acute rubella infection
 - a. Positive viral culture for rubella
 - b. Detection of rubella virus by polymerase chain reaction
 - c. Presence of rubella-specific IgM antibody
 - d. Demonstration of a significant rise in IgG antibody from paired acute- and convalescent-phase sera
 - e. Serology is most common
- Management/Treatment
 1. Isolate infants with CRS from pregnant/potentially pregnant women for one full year
 2. Two thirds of infants with CRS show no symptoms at birth; if maternal infection known or suspected, obtain cultures to determine if infant is shedding virus
 3. Early intervention for developmental stimulation
 4. Refer to cardiology, ophthalmology, audiology for evaluation
 5. Psychosocial support to child and family
 6. Ensure appropriate school placement and supports
 7. Rubella is a reportable disease

Hepatitis B (Refer to Gastrointestinal Disorders chapter)

- Definition: Hepatitis B is a Hepadnaviridae virus that primarily affects the liver that can cause an acute hepatitis with recovery, an acute hepatitis with liver failure, or chronic hepatitis that can lead to cirrhosis, primary carcinoma of the liver, or liver failure
- Etiology/Incidence
 1. Person-to-person transmission of hepatitis B virus via close personal contact, saliva and other secretions, blood and blood products through wound exudates, or sexual contact with infected individuals; perinatal vertical transmission from infected/carrier mother to infant
 2. Perinatal transmission from hepatitis B surface antigen (HBsAg) positive with long-term HBeAg mother to infant is 70% to 90% if infant does not receive postexposure prophylaxis; the perinatal transmission risk is about 10% if the

mother only is positive for HBsAg; around 90% of infected infants eventually become chronically infected with HBV

3. 24% of chronic infections are from perinatal transmission
- Signs and Symptoms
 1. Long incubation period 60 to 150 days, average of 90 days
 2. Malaise, anorexia, nausea, vomiting
 3. Right upper quadrant, abdominal pain, fever, headache.
 4. Myalgia, arthralgia and arthritis
 5. Skin rash
 6. Dark urine, begins 1 to 2 days before the onset of jaundice
 - Differential Diagnosis
 1. Hepatitis A, hepatitis C, hepatitis D, hepatitis E
 2. Cystic fibrosis, Wilson's disease, metabolic liver disease
 3. Infectious causes such as CMV, toxoplasmosis, enterovirus, Epstein-Barr virus
 - Physical Findings
 1. Neonatal infection usually results in "healthy" chronic carrier state
 2. Later, progression to liver disease with:
 - a. Liver enlargement, tenderness
 - b. Jaundice
 - c. Arthralgia or arthritis
 - d. Rash or urticaria
 - Diagnostic Tests/Findings
 1. Presence of surface antigen HBsAg, HBeAg antigen antibodies to either of these or HBcAg (core antigen)
 2. Aminotransferase elevation—peaks at about one month
 3. Bilirubin elevation
 4. Elevated WBC; elevated serum α -fetoprotein concentration
 - Management/Treatment
 1. Chronic infection with hepatitis B occurs if there is an absence of IgM anti-HBc or the persistence of HBsAg for 6 months
 2. To try to avoid perinatal exposure—HBIG 0.5 mL + vaccination in first 12 hours of life; the second dose must be given at 1 to 2 months with third dose at 6 months; the infant should have testing for HBsAg and anti-HBs at 9 to 18 months to measure success of vaccination with HBIG at birth
 3. Prevention of hepatitis B is by universally vaccinating all children so that they cannot become carriers of hepatitis B

4. Chronic infection with hepatitis B is being treated in adults with liver failure and chronic hepatitis C; there are 2 classes of agent interferons (interferon- α 2b and peginterferon- α 2a) or long-term use of nucleosides/nucleotide analogues (lamivudine, adefovir, entecavir, tenofovir, and telbivudine) being used as monotherapy or in combination

Toxoplasmosis

- Definition: Infection with protozoan *Toxoplasma gondii*; it is transmitted by domestic or feral cats that excrete oocytes in cat litter, soil, and then it can be transmitted by changing litter or eating unwashed fruits or vegetables; it can be acquired by eating raw or inadequately cooked meat containing oocytes
- Etiology/Incidence
 1. Congenital infection occurs when a pregnant woman becomes acutely infected; the risk of transmission is 10% to 25% in the first trimester but goes to 60% to 90% in the last trimester
 2. 85% of women of child-bearing age are susceptible to infection
 3. Estimated 400 to 4000 cases per year in the United States or 1 to 10 per 10,000 live births
 4. Placenta to fetus transmission of *Toxoplasma gondii* in mother with primary infection during pregnancy
- Signs and Symptoms
 1. Classic triad—chorioretinitis, intracranial calcification, and hydrocephalus occurs in less than 10%
 2. Asymptomatic at birth in 70% to 90%
 3. Nonspecific manifestations of symptomatic congenital infection
 - a. Microcephaly
 - b. Seizures
 - c. Maculopapular rash
 - d. Hepatosplenomegaly
 - e. Jaundice
 - f. Thrombocytopenia
 4. Later manifestations of congenital infection
 - a. Occur as late as second or third decade
 - b. Learning and visual disabilities from chorioretinitis, which damages the retina
- Differential Diagnosis
 1. Cytomegalovirus
 2. Syphilis
 3. Rubella
 4. Hemolytic disease of the newborn

- Physical Findings
 1. Severe
 - a. Microcephaly or hydrocephalus with or without intracranial calcification
 - b. Retinal lesions on fundoscopic examination
 - c. Splenomegaly, hepatomegaly
 - d. Lymphadenopathy
 - e. Jaundice
 - f. Growth failure
 2. Subclinical (later signs)—retinal lesions on fundoscopic examination
- Diagnostic Tests/Findings
 1. Prenatal—fetal blood or tissue analysis; ultrasonography for bilateral, symmetrical ventriculomegaly
 2. Most common method is based on antibody detection with a raise in IgM 1 to 2 weeks after infection persisting up to 18 months; IgG appears subsequent to IgM with persistence of IgG for the remainder of the life; sensitivity is 93% to 100% and specificity is 77% to 99% and therefore needs confirmation
 3. Good standard is Sabin Feldman dye test
 4. In utero testing by polymerase chain reaction assay (PCR)—detection of genomic material in amniotic fluid can be used with sensitivity of 94% and specificity of 100%
 5. Visual detection of tachyzoites in newborn CSF, ventricular fluid, blood, bone marrow, brain or placental tissue, or detection 1 to 6 weeks after mouse inoculation
 6. Intracerebral calcifications with or without hydrocephalus
- Management/Treatment
 1. Prevention is best—pregnant women should not eat raw or undercooked meat; they should wash fruits and vegetables prior to eating and should wear gloves while gardening, avoid changing cat litter
 2. Congenital—pyrimethamine, sulfadiazine, and leucovorin is most common treatment for overt toxoplasmosis in newborn; if treated, ocular prognosis is better, but late onset visual problems can still occur
 3. Maternal infection—spiramycin is used in other parts of the world but not approved by FDA; it can be used with special permission of FDA
 - a. Maternal treatment with pyrimethamine and sulfadiazine
 - b. Clindamycin as alternative if sulfa compound allergic
 - c. Leucovorin is taken with pyrimethamine

Fetal Alcohol Syndrome (FAS) and Fetal Alcohol Spectrum Disorders (FASD)

- Definition: An umbrella term for a set of disorders that result from the consumption of alcohol during pregnancy; FASD occurs three times more often than FAS; recent estimate is that as many as 12.2% of pregnant women drink during the pregnancy
- Etiology/Incidence
 1. An estimated 1000 to 6000 will be born with FAS with a worldwide estimate of FASD of 1 in 100
 2. Dose that causes damage is unknown but even at low levels of consumption; cell adhesion molecules are inhibiting effecting neuronal migration
 3. Alcohol raises acetaldehyde levels and causes apoptotic damage from excess glutamate activity due to gamma amino butyric acid (GABA) withdrawal; this causes apoptotic neurodegeneration reaction deleting neurons from developing sites in the CNS
- Signs and Symptoms
 1. History of drinking during pregnancy
 2. Infant alcohol withdrawal symptoms (irritability, hyperactivity, jitteriness) and alcohol odor to amniotic fluid if alcohol abuse during last days of pregnancy
 3. There needs to be a CNS abnormality showing at least one of three types of deficits or abnormalities—structural, neurologic, and functional
 4. Fetal alcohol spectrum disorders include fetal alcohol syndrome, alcohol-related neurodevelopmental disorder (ARND), and alcohol-related birth defects (ARBD); these effects include problems with math, memory or attention difficulties, poor school performance, and poor impulse control
 - a. A child with ARND can have functional or mental problems because of prenatal alcohol exposure, including behavioral or cognitive abnormalities or a combination of both
 - b. Children with ARBD can have problems with the heart, kidneys, bones, and/or hearing
- Differential Diagnosis: Williams syndrome, Dubowitz syndrome, fetal Dilantin syndrome, Down syndrome
- Physical Findings will vary depending on whether the child has FAS or another one of the FASD; if the child has FAS, there may be:
 1. Small head circumference
 2. Widely spaced eyes with narrow lids and epicanthal folds

3. Short, upturned, or beaklike nose with broad nasal bridge
 4. Thin upper lip-vermilion border
 5. Absent or flattened philtrum
 6. Micrognathia
 7. Dental malocclusion
 8. Congenital heart defects
 9. Hip subluxation or dislocation
 10. Seizure disorder
 11. Attention deficit disorder
 12. Ophthalmologic disorders—myopia/strabismus
 13. Hearing impairment
 14. Growth impairment with below average height and weight for age
 15. Cleft palate
 16. Mental health disorders aside from attention problems (considered part of the neurological problems) include conduct disorders, oppositional defiant disorders, anxiety disorders, adjustment disorders, sleep disorders, and depression
- Diagnostic Tests/Findings
 1. IQ usually in 60 to 80 range stable throughout life
 2. History of maternal alcohol abuse during pregnancy
 - Management/Treatment
 1. Prevention is key; ensure pregnant women know risk of alcohol intake on fetal development
 2. No known treatment to reverse primary defects
 3. Needs appropriate educational plan with needed support
 4. Screen for vision, hearing, neurological disorders
 5. Refer to <http://www.cdc.gov/ncbddd/fas/default.htm>
 6. For information on FASD: National Organization on Fetal Alcohol Syndrome <http://www.nofas.org/>

■ GENETIC SYNDROMES

Trisomy 18 (Edwards Syndrome)

- Definition: Autosomal chromosomal disorder with karyotype 47 XY or XX + 18, trisomy of chromosome 18; associated with severe mental retardation and other congenital defects associated with advanced maternal age; less than 5% survive beyond first year of life
- Etiology/Incidence
 1. Nondisjunction during meiotic division resulting in trisomy of chromosome 18
 2. 1 in 5000 live birth
 3. 10% of cases are mosaic
 4. If there is only a partial duplication of chromosome 18, they will have a milder presentation
- Signs and Symptoms
 1. Growth retardation
 2. Faces—low set ears with prominent occiput and micrognathia
 3. Musculoskeletal—rocker bottom feet and overlapping fingers (second over third, and fifth over fourth)
 4. Cardiac—congenital heart defects
 5. Neurological; severe global developmental delays, CNS defects with hypertonia
 6. Feeding problems
- Differential Diagnosis: Trisomy 13; other rare chromosomal aberrations
- Physical Findings
 1. Retardation of growth with severe failure to thrive (FTT)
 2. Microcephaly, low set ears, prominent occiput, micrognathia
 3. Heart murmur
 4. Clenched hands with over-riding fingers and crossed thumb
- Diagnostic Tests/Findings
 1. Karyotype
 - a. Fluorescent in situ hybridization (FISH) analysis
 - b. Results usually available within 48 hours
 2. Maternal serum quadruple screen include α -fetoprotein, human chorionic gonadotropin, unconjugated estriol, and inhibin A; this test has 11% greater detection rate than the triple screen at a 5% screen positive rate
 3. Echocardiogram to detect congenital cardiac defects
 4. Chorionic villi sampling or amniocentesis with subsequent pregnancies
- Management/Treatment
 1. Genetic counseling
 2. Psychosocial support to parents and family
 3. Refer to cardiology
 4. Support nutritional needs; may require gastric feedings
 5. Prophylactic antibiotics to prevent subacute bacterial endocarditis (SBE), if cardiac effects present

6. Enroll in early intervention program for habilitative therapies
7. Assist family with management of special needs child—may require in-home nursing
8. Support Organization for Trisomy 18, 13, and Related Disorders (SOFT)
<http://www.trisomy.org/>

Down Syndrome (DS) (Trisomy 21)

- Definition: Down syndrome or trisomy 21 is the most common inherited genetic syndrome associated with a variable degree of mental impairment and karyotype 47 XY or XX + 21
- Etiology/Incidence
 1. Three different genetic alterations
 - a. In 95%, Down syndrome is result of a random nondisjunction (trisomy 21)
 - b. Less commonly, it occurs as mosaicism where some cells are affected and others are normal
 - c. Balanced translocation, often involves chromosome 21 and 14
 2. 1 in 800 to 1,000 live births
 3. Affects males and females equally
 4. Risk factors include advanced maternal age, previous child with Down syndrome or another chromosomal abnormality; parental balanced translocation; parents with chromosomal problems
- Signs and Symptoms
 1. Mental retardation, mild to severe
 2. Typical phenotypic signs at birth
 - a. Head—midface hypoplasia; small brachycephalic head with epicanthal folds, flat nasal bridge, upward slanting palpebral fissures, Brushfield spots, small mouth
 - b. Ears—small ears
 - c. Neck—excessive skin at the nape of the neck
 - d. Hands and feet—simian crease and short fifth finger with clinodactyly; a wide space, often with a deep fissure between the first and second toes; lymphedema, brachydactyly (shortened digits)
 - e. Neurological—mental impairment is variable, ranging from mild (IQ: 50–70), to moderate (IQ: 35–50), and only occasionally to severe (IQ: 20–35)
 - f. Cardiac—increased risk of congenital heart defects (50%)
 - g. GI—Hirschsprung's disease (< 1%); gastrointestinal atresias (12%)
 - h. Musculoskeletal—hypotonia; acquired hip dislocation (6%)
- Differential Diagnosis: Other genetic or chromosomal syndromes
- Physical Findings
 1. Phenotype as above
 2. Hearing loss (75%); otitis media (50%–70%)
 3. Obstructive sleep apnea (50%–75%)
 4. Signs of congenital heart disease—50%; endocardial cushion defect most common (45%) with ventricular septal defects (35%) second
 5. Signs of hypothyroidism and other endocrine problems—15%
 6. Signs of anomaly of GI tract 5%; Celiac disease 15% (Davidson, 2008)
 7. Ligamentous laxity—100%
 8. Hematological—leukemia (< 1%)
 9. Eye—eye disease (60%), including cataracts (15%), and severe refractive errors (50%)
 10. Obesity—50% by early childhood
 11. Musculoskeletal—ankle pronation and pes planus
 12. Premature aging
- Diagnostic Tests/Findings
 1. Pre or postnatal chromosome analysis reveals 47 XY or XX + 21 karyotype
 2. CBC with differential to identify those with leukemia; 10 to 15 fold increased risk
 3. Symptoms of atlantoaxial instability (neck pain, decreased range of motion of the neck, gait disturbance, bowel or bladder dysfunction, hyperreflexia or paresthesias): radiographic finding of atlantoaxial instability
 4. Ophthalmologic evaluations every two years between 3 to 5 years then yearly after this (50% risk of refractive errors between 3 to 5 years)
- Management/Treatment
 1. For complete guidelines by age for patients with Down syndrome see <http://aappolicy.aappublications.org/cgi/content/full/pediatrics;107/2/442>
 2. Primary prevention via education re: risk factors; secondary prevention via prenatal diagnosis
 3. Monitor for growth and family support every well visit
 4. CBC at one year, every year starting at age 13 to 21, and if clinically indicated
 5. Initial evaluation by cardiology to rule out congenital heart defect even if no murmurs are heard
 6. Screen for celiac disease using tissue transglutaminase and IgA starting at age 2 (Down Syndrome Medical Interest Group)

7. Early intervention by PT, OT, speech therapists; special education; review individualized educational plan
8. Genetic counseling for parents and older siblings
9. Periodic full history and physical with sensory and developmental evaluations
10. Nutritional support
11. Screen for thyroid disease yearly; up to 30% risk
12. Prompt referral for associated conditions
13. Patient advocate and guide family during transition to adult care
14. Referral to appropriate web sites for education and support
 - a. National Down Syndrome Society
<http://www.ndss.org/>
 - b. National Association for Down Syndrome
<http://www.nads.org/>
 - c. National Down Syndrome Congress
<http://www.ndscenter.org/>

Fragile X Syndrome (FXS)

- Definition: Fragile X syndrome is associated with a range of intellectual impairments from learning problems to autism and anxiety. The causes of FXS are decreased or absent levels of fragile X mental retardation protein (FMRP). Decreased levels occur when there are greater than 200 CGG repeats in the coding of the protein or by a point mutation or deletion in the fragile X mental retardation gene. The severity of cognitive impairment depends on the magnitude of the FMRP deficit.
- Etiology/Incidence
 1. Genetic anomaly, labeled FMRI, on X chromosome at Xq27.3, the same position as the fragile site
 2. Males are more severely affected than females since they only have one X chromosome; if a male has a methylated full mutation, they will have mild to moderate intellectual disability; females with the full mutation typically have learning disabilities, but about 15% have intellectual disabilities
 3. If the individual has premutation, they have a normal IQ but do have cognitive and behavioral problems and can have behavior consistent with autism
 4. Affected individuals—1:4000 males; 1:8000 females
 5. Carrier females—1:157
 - a. Carriers can have premature ovarian failure, female infertility, late onset tremor/ataxia, psychiatric disease, and autism
- Signs and Symptoms
 1. Behavior phenotype of FXS
 - a. Poor eye contact with excessive shyness and anxiety with hand flapping and biting
 - b. Tactile defensives
 - c. Attentional deficit with hyperactivity, hyperarousal to sensory stimuli, and ASD
 2. Speech delay; perseverative speech; echolalia
 3. Poor gross motor coordination
 4. Stereotypes, e.g., talking to self, spinning, hand flapping
 5. History of seizures—17% to 50%
- Differential Diagnosis
 1. Autism, Asperger syndrome, or pervasive developmental disorder
 2. Mental retardation with nonspecific etiology
 3. Klinefelter's syndrome, Sotos syndrome
 4. Attention deficit hyperactivity disorder
- Physical Findings
 1. Macrocephaly
 2. Prominent forehead with long thin face and prominent jaw, especially in adolescence
 3. Macroorchidism in adolescent males; may be seen as early as age 5
 4. Protuberant, large ears, long or wide
 5. Soft, smooth skin
 6. Heart murmur or apical midsystolic click
 7. Serous otitis media
 8. Strabismus—40%
 9. Joint laxity (especially fingers), hip subluxation, occasionally clubfoot
- Diagnostic Tests/Findings
 1. DNA analysis from whole blood in approved laboratory to confirm diagnosis
 2. Prenatal testing from chorionic villus or amniocentesis sample
- Management/Treatment
 1. Treatment of the behavioral problems with the appropriate psychotropic medication and counseling

- b. Testing for carriage should be done if:
 - (1) Family history of mental retardation, developmental disabilities, or autism
 - (2) Infertile women with increased FSH at less than 40 years
 - (3) Egg and sperm donors
6. Approximately 20% of males asymptomatic, but can transmit gene resulting in symptomatic offspring
7. Most common inherited cause of mental retardation (MR)

2. Psychosocial support to parents, child and family
3. Genetic counseling—no spontaneous mutations have been found for fragile X syndrome; all family members should undergo genetic testing to identify transmitting males, carrier females, and affected individuals
4. Regular well-child examination with attention to:
 - a. Cardiac auscultation—if click or murmur heard, obtain echocardiogram, consider referral to cardiologist for possible mitral valve prolapse
 - b. Otitoscopic evaluation—serous otitis media
 - c. Ophthalmologic evaluation—strabismus (40%), myopia
 - d. Developmental evaluation—mild to severe delays (usually moderate)
 - e. Anticipatory guidance
5. Enroll in early intervention as soon as delays are recognized; speech/language therapy and sensory/motor integration therapy thought to be most helpful
6. Ensure appropriate educational placement with necessary supports
7. National Fragile X Foundation
<http://www.fragilex.org/html/home.shtml>

Turner's Syndrome (XO Karyotype)

- Definition: Chromosomal anomaly resulting in 45,XO (female) karyotype, with developmental, cardiac, reproductive, genetic, and psychosocial issues. To make the diagnosis, the female must have the characteristic features of TS with the complete and partial absence of the second X sex chromosome with or without mosaicism of the cell line. If the patient does not have the clinical features of TS but does have the 45,X cell populations, they do not have TS.
- Etiology/Incidence
 1. Nondisjunction during meiotic division, usually maternal; more than half have a mosaic chromosomal complement (45,XO/46/XX)
 2. TS is present in one of 2500 live-born females; many affected embryos do not survive to term
- Signs and Symptoms
 1. Female with unexplained growth failure or pubertal delay
 2. Constellation of any of the following findings
 - a. Edema of the hands or feet (particularly in newborn)
 - b. Nuchal folds (webbed neck)
 - c. Left sided cardiac anomalies, especially coarctation of aorta or hypoplastic left heart
 - d. Low hairline and high-arched palate
 - e. Low set ears with small mandible; chronic otitis media
 - f. Short statures with growth velocity less than the 10th percentile for age
 - g. Marked elevates FSH
 - h. Cubitus valgus
 - i. Nail hypoplasia
 - j. Multiple pigmented nevi
 - k. Short fourth metacarpal (Bondy, 2007)
- Lack of development of secondary sexual characteristics
- Differential Diagnosis
 1. Congenital lymphedema without Turner's karyotype
 2. Coarctation of aorta without Turner's karyotype
- Physical Findings
 1. Signs and symptoms listed above plus:
 2. Neonatal—lymphedema (usually resolved by age 2 years)
 3. Widely spaced, often inverted nipples with "shield" shaped chest
 4. Hypertension and aortic murmur
 5. Ear deformities
 6. Strabismus, amblyopia, ptosis
 7. Scoliosis (10%)
 8. Defective dentition
- Diagnostic Tests/Findings
 1. All Turner's syndrome patients
 - a. Cytogenetic testing for karyotype 45,XO
 - b. Renal ultrasound to detect renal anomalies
 - c. Evaluation for hearing loss; as adult risk of sensorineural loss is 60%
 - d. Cardiac echocardiogram or MRI for:
 - (1) Coarctation of aorta (20%)
 - (2) Bicuspid aortic valve (50%)
 - e. Annually from age 4 onward, T₄, TSH due to high rate of autoimmune thyroid disease
 - f. Abdominal and pelvic ultrasound to detect gonadal dysgenesis
 - g. Plasma gonadotropin studies to detect low levels of normal female hormones
- Management/Treatment
 1. Refer to endocrinology
 - a. To increase adult height, growth hormone therapy
 - b. Hormone (estrogen) replacement therapy beginning about 12 to 13 years of age
 - c. Monitor for hypothyroidism
 2. Genetic counseling

3. Psychosocial support
4. Assistance in school for learning disabilities
5. Referral to cardiology for cardiac anomaly diagnosis and treatment
6. Referral to ophthalmology—strabismus and hyperopia (farsightedness) each occur in 25% to 35% of these children (Bondy, 2007)
7. Referral to ENT if recurrent otitis due to abnormal relationship between the eustachian tube and middle ear since the cranial base is different
8. Referral to orthodontist due to narrowed maxilla and wide, micrognathic mandible as well as pediatric dentist
9. Referral to orthopedics, urology if scoliosis, renal anomalies
10. Screen for celiac disease, which runs from 4% to 6% in patients with Turner's syndrome
11. Turner's Syndrome Society of the United States
12. Information on Turner's syndrome
<http://turners.nichd.nih.gov/>

Klinefelter's Syndrome (XXY Karyotype)

- Definition: Klinefelter's syndrome is a genetic mutation with 47,XXY karyotype or a variant among men with an estimated frequency of 1:500 to 1:1000 newborn (Paduch, Fine, Bolyakov, & Kiper, 2008); linked to advanced maternal age; men with mosaicism are less affected and tend to go undiagnosed
- Etiology/Incidence
 1. Maternal meiotic nondisjunction resulting in contribution of two X chromosomes to maternal zygote (ova); when ova is fertilized by sperm containing one Y chromosome, resulting embryo has Klinefelter's karyotype
 2. Most common numerical chromosomal aberration is double X and a Y
 3. Most common cause of hypogonadism and infertility in men
- Signs and Symptoms
 1. Classic description
 - a. Tall eunuchoid body proportion male, especially at adolescence and beyond
 - b. Slow, incomplete pubertal development
 - c. Sparse facial and pubic hair
 - d. Small hard testicle
 - e. Micropenis with sterility
 - f. Behavioral and psychiatric disorders (shy, immature, anxious, aggressive, antisocial)
 2. More common
 - a. Auditory processing delay
 - b. Language dysfunction

- c. Infertility
- d. Low testosterone such as erectile dysfunction and poor libido
- e. Harder testicles with a volume of 10 ml regardless of penile size, body proportions, or level of androgenization

- Differential Diagnosis
 1. Marfan syndrome
 2. Sotos syndrome
 3. Trisomy 8p
- Physical Findings
 1. Tall for age, with disproportionate lower limb length
 2. Gynecomastia
 3. Small, firm testes
 4. Cryptorchidism
 5. Small phallus
 6. Hypospadias
 7. Less pubic and facial hair
- Diagnostic Tests/Findings
 1. Chromosome analysis yields 47,XXY karyotype or mosaic
 2. Low testosterone levels and elevated estradiol levels are cardinal symptoms of Klinefelter's
 3. LH and FSH elevation starts early in puberty
 4. Full hormonal evaluation including FSH, LH, testosterone, estradiol, prolactin, IGF-1, and cortisol (deficiency of adrenal steroids in 47%)
 5. Hyperestrogenism is common
- Management/Treatment
 1. Early intervention for learning disorders
 2. Counseling/therapy for behavioral disorders
 3. Psychosocial support for family
 4. Genetic counseling
 5. Refer to endocrinology for consideration of testosterone therapy at age 11 or 12
 6. Screen for breast cancer (4%)
 7. Reduction mammoplasty for severe gynecomastia
 8. Support and information for families:
American Association for Klinefelter Syndrome
Information and Support
<http://www.aaksis.org/>
Klinefelter's Syndrome Association UK
<http://www.ksa-uk.co.uk/>

Tay-Sachs Disease

- Definition: Inborn error of metabolism, which causes a deficiency of hexosaminidase A. This lysosomal enzyme deficiency results in the accumulation of G_{m2} ganglioside in the lysosomes of

various tissues, which results in neurologic degenerative disease and death, usually by 4 years of age due to recurrent infection.

- Etiology/Incidence
 1. Autosomal recessive single gene disorder; deficiency of hexosaminidase A (hex A) which is necessary for breakdown of ganglioside G_{m2} ; as glycoside accumulates in neurons, axons degenerate and demyelination occurs
 2. Ashkenazi Jewish population (80%), some Pennsylvania Dutch, Louisiana Cajun, French Canadian; carrier rate may be as high 1:52 of Irish-Americans
 3. Autosomal recessive transmission of the HEXA gene, located on chromosome 15
- Signs and Symptoms
 1. Normal development until age 3 to 6 months, then progressive deterioration
 2. Earliest symptom is irritability and increased reaction to sound
 3. Hypotonia with development delay then deterioration
 4. Vision starts to deteriorate by age 6 months, with blindness as early as 1 year; cherry red spot of the macula is due to degeneration of the ganglion cells
 5. Macrocephalic due to accumulation of the cerebral G_{m2}
 6. Frequent upper respiratory infections
 7. Seizure activity as early as 6 months
- Differential Diagnosis
 1. Sandhoff's disease
 2. Krabbe's disease
 3. Niemann-Pick disease
 4. Farber's lipogranulomatosis
 5. Gaucher's disease
 6. Beals syndrome (congenital contractural arachnodactyly)
 7. Homocystinuria
- Diagnostic Tests/Findings
 1. Gene testing for known mutations
 2. Serum enzymatic assay yields deficiency of hexosaminidase A
 3. MRI shows enlarged caudate nucleus, later progression to cerebral atrophy
- Management/Treatment
 1. Genetic counseling
 2. Primary prevention via carrier screening
 3. Secondary prevention via prenatal diagnosis and elective termination of pregnancy
 4. No known treatment for underlying metabolic deficiency

5. Supportive/comfort care for child; assist to obtain home nursing services as disease progresses and care burden increases
6. Psychosocial support for parents and family
7. The National Tay-Sachs and Allied Diseases Association
<http://www.ntsad.org/>

Marfan Syndrome

- Definition: Inherited disorder of connective tissue; affects the skeletal, cardiovascular, and ocular systems
- Etiology/Incidence
 1. Autosomal dominant inheritance of defective fibrillin gene (FBN1 mapped to chromosome 15 [15q21.1]); 15% sporadic mutation
 2. Incidence is 1:5000–10,000
- Signs and Symptoms
 1. Skeletal—tall stature; long, thin extremities, long fingers, narrow facies
 - a. Major criteria—must have four of these:
 - (1) Loose joints
 - (2) Scoliosis of more than 20 degrees (60%)
 - (3) Pectus excavatum requiring surgery or pectus carinatum
 - (4) Arm to height ratio of greater than 1.05 or reduced upper to lower segment
 - (5) Positive wrist and thumb sign
 - (6) Medial displacement of median malleolus causing pes planus
 - b. Minor criteria
 - (1) Hypermobility of joint
 - (2) Pectus excavatum of moderate intensity
 - (3) High arch palate with crowding of teeth
 - (4) Facial appearance with dolichocephaly, downward slanting palpebral fissures
 2. Cardiovascular
 - a. Major criteria—dilation of aorta with or without aortic regurgitation
 - b. Minor criteria—murmur indicative of mitral valve prolapse, dilation of the pulmonary artery without any reason in patient younger than 40 years, dissecting thoracic or aortic aneurysm in patient younger than 50
 3. Ocular
 - a. Major criteria—ectopia lentis (dislocation of the lens)
 - b. Minor criteria—myopia, abnormally flat cornea, hypoplastic iris or hypoplastic ciliary muscle causes decrease in miosis
 4. Skin—minor criteria—striae atrophicae

5. Pulmonary
 - a. Major criteria—none
 - b. Minor criteria—spontaneous pneumothorax with apical blebs
- Diagnostic Tests/Findings
 1. Diagnosis based on having involvement in two or more systems with major criteria and a third system with minor criteria in the absence of family history
 2. Positive family history plus one or more systems in major criteria and involvement of another organ system
 3. Cardiac evaluation (chest radiograph, electrocardiogram, echocardiogram)—mitral valve prolapse common; signs of dilatation of aortic root or dissecting aortic aneurysm
 4. Ocular evaluation—slit-lamp examination
 5. Skeletal evaluation—scoliosis screening; trunk/extremities ratio
 6. Urine screening for amino acids at birth to evaluate for homocystinuria
 - Management/Treatment
 1. Refer to cardiology for periodic echocardiogram to detect dissecting aortic aneurysm, mitral valve prolapse in severe cases, surgical graft repair of the ascending aorta and aortic valve has been successful
 2. Propranolol to reduce effect of ventricular ejection on ascending aorta
 3. Leukodystrophies
 4. Muscular dystrophy
 5. Refer to ophthalmology for treatment of myopia, lens subluxation, cataracts, glaucoma, and retinal detachment
 6. Refer to endocrinology for hormonal treatment to curtail height, valuable psychological effect; prevention of scoliosis and kyphosis; prevention of secondary problems of feet
 7. Psychosocial support for patient and family
 8. Genetic counseling
 9. Ensure mainstream or inclusive school placement with any necessary supports, with attention to physical activity limitations if cardiovascular involvement
 10. National Marfan Foundation (NMF)
<http://www.marfan.org>

Mucopolysaccharidosis I (MPS IH)

- Definition: Rare group of lysosomal storage disorders caused by a deficiency of lysosomal enzyme α 1-iduronidase; it is an autosomal recessive disorder in which the patient is unable to degrade glycosaminoglycan dermatan sulfate and heparan sulfate; the disease was divided into Hurler syndrome (severe), Hurler Scheie syndrome (intermediate), and Scheie syndrome (mild); experts now feel it should be changed to severe MPS I (Hurler syndrome) and attenuated MPS I (Hurler-Scheie and Scheie syndrome)
- Etiology/Incidence
 1. Due to the deficiency of lysosomal enzyme α 1-iduronidase, the patient accumulates glycosaminoglycan within lysosomes and there is multiorgan dysfunction and damage as a result
 2. 1:100,000 incidence but 50% to 80% exhibit severe form
- Signs and Symptoms
 1. Attenuated MPS I can go undiagnosed for years
 2. Severe MPS I may not be diagnosed until after 12 month to 18 months
 3. Progressive cognitive impairment with progressive neurodegenerative disorder in severe MPS I; early development is normal and delay is more obvious by 1 to 2 years of age
 4. Over time, coarse facial features with enlarged tongue, full lips, flat nasal bridge become more obvious
 5. Communicating hydrocephalus develops over months
 6. Onset of language skills is delayed
 7. Behavior tends to be placid rather than hyperactive or aggressive seen in MPS II or III
 8. Recurrent upper respiratory tract infections with otitis
 9. Snoring and coarse breathing occur due to adenoidal and tonsillar enlargement
 10. Recurrent inguinal hernia
 11. Restrictive lung disease with sleep apnea and asthma
 12. Vision loss due to corneal clouding and sensorineural hearing impairments
 13. Progressive skeletal and joint disease leading to dysostosis multiplex, scoliosis, kyphosis, and hip dislocation
 14. Short stature
- Differential Diagnosis: Other progressive neuropathies and inborn errors of metabolism, particularly other mucopolysaccharidoses and thyroid deficiency
- Physical Findings
 1. Skeletal abnormalities, including spinal anomalies/gibbus formation
 2. Macrocephaly, scaphocephaly
 3. Hepatosplenomegaly
 4. Clouded corneas

5. Inguinal/umbilical hernia
 6. Valvular heart disease; coronary artery disease
- Diagnostic Tests/Findings
 1. Prenatal diagnosis with amniocentesis or chorionic villus sampling and enzyme analysis
 2. Postnatal via serum and urine enzyme analysis
 - Management/Treatment
 1. Genetic counseling
 2. Psychosocial support
 3. Anticipatory guidance
 4. Early intervention; appropriate school placement with supports as needed
 5. MRI of brain and spine
 6. Annual audiology tests and ophthalmologic exams
 7. Pulmonary function tests and sleep studies
 8. Cardiology and orthopedics evaluation
 9. Bone marrow transplantation in selected cases, especially if a human leukocyte antigen (HLA) matched sibling donor is available
 10. Refer to audiology and ophthalmology for evaluation and treatment as indicated
 11. National MPS Society, Inc.
<http://www.mpsociety.org/>

Prader-Willi Syndrome

- Definition: Congenital disorder characterized by voracious, uncontrollable appetite and obesity
- Etiology/Incidence
 1. Usually sporadic mutation; when detectable (about 50%) mutation at same location of chromosome 15 as the mutation for Angelman syndrome, although conditions are very dissimilar due to phenomenon of genetic imprinting
 2. 1:10,000 to 1:15,000 incidence
- Signs and Symptoms
 1. Voracious appetite during childhood and beyond, resulting in severe obesity
 2. Mental retardation
 3. Behavior problems
 4. Hypotonia, poor suck and feeding problems in infancy; resolution with time
- Differential Diagnosis: Other neurological and musculoskeletal disorders with early hypotonia (including cerebral palsy) and developmental delay
- Physical Findings
 1. Obesity
 2. Small hands and feet

3. Small genitalia
4. Cryptorchidism
5. Short stature
6. Scoliosis

- Diagnostic Tests/Findings
 1. In 50% of cases, chromosome analysis detects aberration of chromosome 15 section 15q11 to 15q13
 2. Remainder of cases are diagnosed by clinical signs and symptoms
 3. Growth hormone deficiency frequent, but not universal
- Management/Treatment
 1. Behavioral therapy for control of eating and other problem behaviors
 2. Genetic counseling
 3. Early intervention and appropriate school placement with supports
 4. Psychosocial support for child and family
 5. Refer to endocrinology for evaluation and management of growth hormone deficiency
 6. Prader-Willi Syndrome Association
<http://www.pwsausa.org/>

■ MULTISYSTEM DISORDERS

Cerebral Palsy (CP)

- Definition: A group of disorders of motor control and balance due to a static injury to the developing brain; the motor disorders are often accompanied with disorders of sensation, cognition, or communication with or without seizure disorder; while the manifestations can change over time, the brain lesion is static; the damage that occurs to the brain is permanent, but the outcome can be minimized; the classification of the four types of CP is based on the changes in muscle tone, anatomic region of involvement, and severity of the problem—spastic (diplegia, quadriplegia, hemiplegia), dyskinetic (choreoathetoid, dystonic), hypotonic, and mixed
- Etiology/Incidence
 1. Prevalence is 2.5 per 1000 live births
 2. The etiology can be a CNS insult in the prenatal, perinatal, postnatal periods, or as the result of prematurity
 3. Prenatal causes include brain malformation, in utero stroke, congenital cytomegalovirus infection
 4. Perinatal causes include hypoxic ischemic encephalopathy, viral encephalitis, and meningitis
 5. Postnatal causes include accidental head trauma, anoxic insult, and child abuse

6. High association with prematurity and very low birthweight
 7. In most preterm infants, there is usually no single factor identified as the cause, but there may be multiple risk factors
- Signs and Symptoms
 1. The essential findings include delayed motor milestones, abnormal muscle tone, hyper-reflexia, absence of regression or evidence of a different diagnosis
 2. There may be transient abnormalities in tone and reflexes that interfere with motor progress and can be outgrown
 3. Vision impairment (50%)
 4. Hearing impairment (10%)
 5. Seizures (30%)
 6. Cognitive impairment (50%)
 7. High risk of dental disease
 8. Higher risk of sleep disorders (23%)
 - Differential Diagnosis
 1. Neurodegenerative disorders such as Duchenne muscular dystrophy
 2. Metabolic disorders
 3. Transient toe walking
 4. Familial spastic paraplegia
 5. Genetic disorders such as Miller-Dieker or Rett syndrome
 6. Mitochondrial disorders
 7. Spinal cord tumor/syrinx
 8. Brain tumor
 9. Hydrocephalus, dystonias
 - Physical Findings
 1. Abnormal muscle tone (hypo or hypertonia) with developmental delay
 2. Muscle weakness
 3. Loss of selective control
 4. Retained primitive reflexes or pathological reflexes
 5. Hyperactive tendon and heightened stretch reflexes; positive Babinski
 6. Restricted joint range of motion
 7. Hip click or clunk on Barlow maneuver or Ortolani test
 8. Movement related muscle spasms with spasticity
 9. Low weight for height
 10. Neuromuscular scoliosis
 11. Visual and hearing problems
 12. Swallowing and feeding difficulty with failure to thrive
 13. Respiratory problems
 14. Incontinence
- Diagnostic Tests/Findings
 1. Developmental evaluation—delays in gross motor, fine motor, speech, according to type of CP and presence of mental retardation
 2. Brain MRI—90% will show changes including in utero strokes, major and minor brain malformations, and white matter loss including periventricular leukomalacia (low birthweight infants)
 - Management/Treatment
 1. Coordinate interdisciplinary management to promote optimum health and function
 2. Enroll in early intervention services—speech, OT, PT services
 3. Identify and treat associated conditions (e.g., seizures, visual impairment, hearing impairment, gastroesophageal reflux) and make appropriate referrals
 4. Prevent secondary conditions (e.g., failure to thrive, skin breakdown, dental caries)
 5. Functional therapies to build on strengths and promote compensation for physical impairments
 - a. Adaptive seating
 - b. Bracing
 - c. Wheeled mobility
 6. Parent/family support for positive coping and stress relief
 7. Spasticity relief
 - a. Enteral medication—lioresal, diazepam, dantrium sodium, tizanidine hydrochloride
 - b. Botulinum toxin IM to major affected muscles, or nerve block injections
 - c. Intrathecal lioresal
 - d. Selective dorsal root rhizotomy
 8. Support growth and nutrition, gastrostomy if needed, medication for reflux
 9. Ensure appropriate education with supportive services and therapies
 10. Oromotor therapy including chewing, swallowing, and speech therapy
 11. Seizure prevention
 12. Orthopedist referral for corrective casting, muscle release and lengthening, split tendon transfers, osteotomies, and arthrodeses
 13. Transitional care to adult provider when ready
 14. United Cerebral Palsy
<http://www.ucp.org>

Spina Bifida

- Definition: Congenital abnormality of the CNS or the spine resulting from failure of neural tube closure during early embryonic development; risk

factors for neural tube defect include maternal folic acid deficiency, maternal use of valproic acid or carbamazepine, or maternal diabetes

- Etiology/Incidence
 1. Multifactorial inheritance pattern; environmental contribution not well understood, although addition of folic acid to dietary intake reduces occurrence probability by one half
 2. Spina bifida occulta—incidence up to 10% of population
 3. Myelomeningocele—1:1000; decreasing, probably due to folic acid supplementation and prenatal diagnosis with selective termination (not proven)
- Signs and Symptoms
 1. Neurological—can have hydrocephalus, Chiari malformation and hydromyelia, tethered cord
 2. Orthopedics—hip dislocation, knee contractures, spinal deformities such as kyphosis, scoliosis, fractures
 3. Urological—urinary “dribbling,” unable to achieve urinary continence, frequent urinary tract infections, ureteral reflux with renal damage
 4. GI: Chronic constipation, difficulty with bowel continence
 5. Motor developmental delays, especially lower extremity related gross motor delays
 6. Latex allergies
 7. Intelligence in normal range, but with learning disorders, often with attention deficit hyperactivity disorder (ADHD)
- Differential Diagnosis: Syndromes of which spina bifida is associated, e.g., Meckel-Gruber syndrome
- Physical Findings
 1. Spina bifida occulta—usually benign; may have sacral dimple, hairy patch at base of spine, uneven gluteal folds
 2. Arnold-Chiari Type II CNS malformation (nearly 100%)—associated with progressive hydrocephalus, difficulty swallowing, hypoventilation, apnea
 3. Meningocele or myelomeningocele—signs at birth include lesion at some point along thoraco-lumbar-sacral spine, often with a cystlike structure protruding; neural elements may be apparently absent or may be easily visualized within the sac
 4. Widely spaced cranial sutures, bulging fontanel, macrocephaly (with hydrocephalus)
 5. Lack of typical lower extremity function, sometimes with orthopedic deformity (clubfoot, dislocated or subluxed hip, tibial torsion)
 6. Abnormal deep tendon reflexes in lower extremities
 7. Abnormal neonatal reflexes in lower extremities
 8. Decreased or absent anal wink
 9. Atrophied lower extremity/hip muscles
 10. Scoliosis, kyphosis
 11. Obesity in older children and adolescents (> 50%)
 12. Neurogenic bowel and bladder
 13. Latex sensitivity (> 40%)
- Diagnostic Tests/Findings
 1. Prenatal diagnosis possible by maternal serum screening for elevated α -fetoprotein, followed by ultrasound diagnostics for spinal anomaly and head “lemon sign”
 2. Postnatal diagnosis made on clinical basis
 3. Hydrocephalus after birth, increasing head circumference out of proportion to other growth parameters
- Management/Treatment
 1. Infants diagnosed prenatally should be referred to tertiary center with appropriate supports for birth (possible planned C-section) and immediate neonatal intensive care
 2. Refer to multidisciplinary treatment center for specialty management—assistance from orthopedist, urologist, neurosurgeon, developmental pediatrician, orthotist, physical and occupational therapists, nutritionist, advanced practice nurse, and social worker
 3. Enroll infant in early intervention program as soon as medically stable
 4. Monitor for urinary tract infections; expect less common organisms; monitor for shunt malfunction if presence of shunted hydrocephalus; baseline head CT scan; follow-up if increased intracranial pressure suspected; refer to neurosurgeon for evaluation of suspected shunt malfunction or tethered cord
 5. Monitor for development of orthopedic problems, especially scoliosis and unilateral hip subluxation or dislocation; baseline and follow-up radiographic studies
 6. Monitor for skin breakdown
 7. Nutritional and behavioral intervention to prevent obesity
 8. Test for latex sensitivity (skin or RAST); latex precautions
 9. Anticipatory guidance for development, safety
 10. Psychosocial support to family and child
 11. Assistance finding least restrictive school placement and other community supports—restrict from heavy contact sports only; otherwise full inclusion should be encouraged

12. Genetic counseling
13. All women of childbearing age should consume 0.8 mg folic acid daily to help prevent neural tube defects
14. Referral to Spina Bifida Association of America (SBAA)
www.sbaa.org/

Sudden Infant Death Syndrome (SIDS)

- Definition: The death of infants who are less than one year without a physiological cause despite a complete examination of the case including an autopsy, examination of scene of death, and a review of the child's medical history and record
- Etiology/Incidence
 1. Unknown cause
 2. 2230 SIDS deaths occurred in 2005 with higher rates among non-Hispanic Blacks and American Indian/Alaska natives
 3. Peak incidence 2 to 4 months; uncommon before 2 weeks and after 6 months
- Signs and Symptoms: Infant unexpectedly found lifeless after a period of sleep
- Differential Diagnosis
 1. Meningitis
 2. Intracranial hemorrhage
 3. Myocarditis
 4. Accidental trauma
 5. Child abuse
 6. Metabolic disorder like medium-chain acyl-CoA dehydrogenase (MCAD) deficiency
- Physical Findings
 1. Full cardiorespiratory arrest
 2. Unresponsive to resuscitation
- Diagnostic Tests/Findings: Diagnosis of exclusion, with autopsy and investigation, failure to find adequate cause of death
- Management/Treatment
 1. Risk reduction
 - a. Encourage prenatal care
 - b. Supine sleeping position, not side lying position
 - c. Avoid maternal and passive smoking
 - d. Separate sleeping place for infants
 - e. Avoid soft bedding
 - f. Maintain comfortable room temperature
 - g. Avoid heavy blankets, over-bundling
 - h. Consider offering a clean pacifier at nap and bedtime but do not force it
 - i. Avoid commercial devices to reduce risk of SIDS

- j. Avoid positional plagiocephaly by encouraging the mother to turn the baby's head from side to side, encouraging tummy time, and avoid excessive time in car seat carriers
- k. Apnea monitoring for high risk infants
 - (1) Premature with persistent apnea
 - (2) Infant born after two previous siblings with SIDS
 - (3) Post-apparent life threatening event (ALTE) requiring stimulation
 - (4) Infants with central hypoventilation syndrome
2. After infant's death
 - a. Maximum support to family, others
 - b. Provide factual information
 - c. Assist with necessary tasks
 - d. Assist nursing mother with abrupt cessation of breastfeeding
3. National SIDS Resource Center
<http://www.sidscenter.org>

▣ QUESTIONS

Select the best answer

1. Which of the following is a characteristic physical sign of fragile X syndrome in adolescent males?
 - a. Small posteriorly rotated ears
 - b. Macroorchidism
 - c. Hypertonia
 - d. Double hair whirl
2. Which of the following physical stigmata are common in newborns with Down syndrome?
 - a. Microcephaly, large ears and mouth, flattened philtrum
 - b. Hypotonia, large appearing tongue and small mouth, upward slant to eyes
 - c. Fair mottled skin, large hands and feet, broad stocky neck
 - d. Funnel or pigeon-breasted chest, Brushfield spots, extra digits
3. A two-month-old infant, with a history of sacral myelomeningocele repair, has an increase in head circumference from the 75th to the 95th percentile. What is the most appropriate first action?
 - a. Order a stat head CT scan
 - b. Refer to neurosurgery for management
 - c. Recheck it at the next well-child visit
 - d. Recheck and replot the child's head circumference

4. A 2-week-old presents with mucopurulent eye discharge with injection and edema of the conjunctiva. The 15-year-old mother had no prenatal care. What is the most likely organism?
 - a. Staphylococcus aureus
 - b. Gonococcus
 - c. C. Pneumoniae
 - d. C. Trachomatis
5. A newborn presents with a large VSD, rocker bottom feet, overlapping second and third fingers, and fourth and fifth fingers with hypotonia. What is the most likely diagnosis?
 - a. Fragile X syndrome
 - b. Down syndrome
 - c. Edwards syndrome
 - d. Klinefelter's syndrome
6. Which of the following problems is common in a child with Hurler's syndrome?
 - a. Developmental delay from birth
 - b. Sleep obstructive apnea
 - c. Ectopic lentis
 - d. Congenital heart disease
7. Young infants with cerebral palsy often show:
 - a. Voracious appetite and weight gain
 - b. Increased muscle tone in the first weeks of life
 - c. Hypotonia in the first weeks of life
 - d. Unusually severe reactions to their first immunizations
8. A 5-year-old child born at 28 weeks gestation has mild spastic diplegia. Which evaluation is important before the child is placed in regular kindergarten?
 - a. Stanford-Binet IQ test
 - b. Carey Scale of temperament
 - c. CBC with differential
 - d. Chromosome studies
9. A mother with Marfan syndrome comes with her 5-year-old for a checkup. The child has myopia and a positive wrist and thumb sign. She reports that she was told by her last health-care provider not to worry about this child. What is the next best step?
 - a. Raise the issue of a genetic referral
 - b. Reassure her
 - c. Follow the child for further signs of Marfan syndrome
 - d. Refer to ophthalmology
10. Which of the following is appropriate advice for the mother of a newborn?
 - a. The child should sleep on her back
 - b. The child should sleep on her back or side
 - c. The child should be allowed to sleep in a carseat at night
 - d. The child can sleep on her comforter
11. A five-year-old female presents for a school physical with a complaint of hyperactivity, a mild developmental delay, aversion of gaze, hand mannerism, long thin face with a slightly dysmorphic ear. What is the most likely diagnosis?
 - a. Fragile X
 - b. Turner's syndrome
 - c. Fetal alcohol syndrome
 - d. Williams syndrome
12. Congenital HIV infection:
 - a. Is diagnosed through finding maternal antibodies in infant serum
 - b. Does not respond to antiretroviral therapy
 - c. May be latent for years before clinical signs develop
 - d. Does not include lymphadenopathy as a physical finding
13. An infant presents with cataracts, congenital glaucoma, congenital heart disease, hepatosplenomegaly, thrombocytopenia, blueberry muffin rash, and growth retardation. What is the most likely diagnosis?
 - a. Cytomegalovirus
 - b. Toxoplasmosis
 - c. Rubella syndrome
 - d. Syphilis
14. A well-appearing, well-dressed 11-month-old has a long philtrum, midface hypoplasia, microcephaly, mild developmental delay, and myopia. The mother denies drinking alcohol during the pregnancy. What is the next best step?
 - a. Refer the mother-child to child protective services
 - b. Refer the child to early intervention
 - c. Refer the child to cardiology
 - d. Refer the mother to Alcoholics Anonymous
15. An 18-month-old child exposed to HIV in utero had negative HIV DNA at 2 weeks and 8 weeks. What is indicated at the 18-month visit?
 - a. No further testing is needed
 - b. HIV DNA should be repeated
 - c. Enzyme immunoassay for antibody to HIV-1
 - d. CBC with differential and immunoglobulins

16. Which of the following infant diseases is not associated with exposure in the first trimester of pregnancy?
- Cytomegalic inclusion disease (CMV)
 - Chlamydia conjunctivitis
 - Toxoplasmosis
 - Rubella
17. A baby is born to a mother with chronic hepatitis B. What is the best treatment approach?
- Administer interferon within 24 hours
 - Administer hepatitis B vaccine within 24 hours
 - Administer interferon and nucleotide in combination daily
 - Administer HBIG and hepatitis B vaccine within 12 hours
18. A 2-year-old female has lymphedema of the hands and foot, with low posterior hairline, cubitus valgus, and a history of intrauterine growth retardation. Which of the following defects is the most common among the children with this defect?
- Aortic valve stenosis
 - Mitral valve prolapse
 - Dissecting aortic aneurysm
 - Coarctation of the aorta
19. Joshua is the 9-month-old infant son of parents of Louisiana-French descent. The child stopped rolling over. Mother reports he is increasingly irritable. Which of the following physical exam findings is most consistent with Tay-Sachs disease?
- Cardiomyopathy
 - Retinal detachment
 - "Cherry red" spot on retina
 - Hepatomegaly
20. Prader-Willi syndrome is a congenital genetic disorder characterized by:
- Failure to thrive
 - 100% detection rate with chromosome analysis for a 15q deletion
 - Emergence of spasticity during toddler years
 - Voracious appetite and development of obesity
21. A 15-day-old infant with respiratory distress arrives in the ED. The exam reveals mild cyanosis, hepatosplenomegaly, and features consistent with Down syndrome. Which of the following is the most likely diagnosis?
- Complete AV canal (endocardial cushion defect)
 - Patent ductus arteriosus
 - Atrial septal defect
 - Ventricular septal defect
22. A newborn presents with lymphadenopathy, a decrease in the ability to move the left leg, Coombs-negative hemolytic anemia, hepatomegaly, and snuffles. What is the most likely diagnosis?
- Congenital herpes infections
 - Congenital cytomegaloviral infection
 - Congenital syphilis
 - Congenital gonococcal infection
23. Individuals who have spina bifida are at high risk for allergy to:
- Eggs
 - Pollens
 - Latex
 - Dust mite feces
24. What is the preferred treatment of choice for syphilis?
- Erythromycin
 - Penicillin
 - Cefotaxime
 - Zithromax
25. Which of the following is consistent with neonatal disseminated herpes disease?
- Hyperactive newborn with apparent spasticity
 - Multiple papules scattered over the body
 - Fever, grouped vesicles on the skin
 - Purpuric rash in an acral distribution

▣ ANSWERS

- | | |
|--------------|--------------|
| 1. b | 14. b |
| 2. b | 15. c |
| 3. d | 16. b |
| 4. d | 17. d |
| 5. c | 18. d |
| 6. b | 19. c |
| 7. c | 20. d |
| 8. a | 21. a |
| 9. a | 22. c |
| 10. a | 23. c |
| 11. a | 24. b |
| 12. c | 25. c |
| 13. c | |

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Advanced Practice, Role Development, Current Trends, and Health Policy

Janet Selway

■ INTRODUCTION

Advanced practice registered nurses (APRN) must remain informed regarding role development, current issues and trends related to their practice, as well as changes in healthcare policy, as each has an impact on the evolving practice environment. In July 2008, the National Council of State Boards of Nursing (NCSBN) APRN Committee and the Advanced Practice Nursing Consensus Work Group completed a regulatory model for the future regulation of Advanced Practice Nursing. This document delineates the definition of an APRN based on seven characteristics.

■ ADVANCED PRACTICE NURSING

1. Definition of Advanced Practice Registered Nursing (APRN) according to the Consensus Model for APRN Regulation (APRN Consensus, p. 6), “The definition of an APRN is a nurse:
 - a. who has completed an accredited graduate-level education program preparing him/her for one of the four recognized APRN roles;
 - b. who has passed a national certification examination that measures APRN, role and population-focused competencies and who maintains continued competence as evidenced by recertification in the role and population through the national certification program;
 - c. who has acquired advanced clinical knowledge and skills preparing him/her to provide direct care to patients, as well

as a component of indirect care; however, the defining factor for APRNs is that a significant component of the education and practice focuses on direct care of individuals;

- d. whose practice builds on the competencies of registered nurses (RNs) by demonstrating a greater depth and breadth of knowledge, a greater synthesis of data, increased complexity of skills and interventions, and greater role autonomy;
 - e. who is educationally prepared to assume responsibility and accountability for health promotion and/or maintenance as well as the assessment, diagnosis, and management of patient problems, which includes the use and prescription of pharmacologic and nonpharmacologic interventions;
 - f. who has clinical experience of sufficient depth and breadth to reflect the intended license;
 - g. who has obtained a license to practice as an APRN in one of the four APRN roles—certified registered nurse anesthetist (CRNA), certified nurse-midwife (CNM), clinical nurse specialist (CNS), or certified nurse practitioner.”
2. APRNs are educated in one of the four roles and in at least one of six population foci—family/individual across the lifespan, adult-gerontology, pediatrics, neonatal, women’s health/gender-related, or psych/mental health

3. Many nurses with advanced graduate nursing preparation practice in roles and specialties that do not focus on direct care to individuals and, therefore, their practice does not require regulatory recognition beyond the registered nurse license granted by state boards of nursing. These other advanced, graduate nursing roles are not APRN roles (APRN Consensus, 2008).
4. The National Association of Pediatric Nurse Practitioners, (formerly known as the National Association of Pediatric Nurse Associates and Practitioners) endorsed the July 2008 Consensus Model for APRN Regulation. According to the Consensus Model, a nurse practitioner should have education in the role of nurse practitioner and one or more population foci, including pediatrics as a distinct population focus.
 - a. 1992 Position Statement on entry into practice endorsed the master's level for PNP preparation
 - b. 2008 Position Statement on Doctorate of Nursing Practice (DNP) endorsed DNP as future-oriented goal for entry level for PNP practice. Master's-prepared PNPs are valued; not all master's-prepared PNPs require a DNP.
 - c. 2008 NAPNAP joins the NP Roundtable. The NP Roundtable releases its first statement regarding DNP education, certification, and titling. The NP Roundtable is a coalition of seven NP organizations formed to collaborate, unify, and address issues of importance to NPs. The NP Roundtable includes NAPNAP, the American Academy of Nurse Practitioners (AANP), the American College of Nurse Practitioners (ACNP), the Association of Faculties of Pediatric Nurse Practitioners (AFPNP), the National Association of Nurse Practitioners in Women's Health (NPWH), the Gerontological Advanced Practice Nurses Association (GAPNA, formerly known as the National Conference of Gerontological Nurse Practitioners), and the National Organization of Nurse Practitioner Faculties (NONPF).
 - d. Curricular content includes growth and development, pathophysiology, physical, developmental, family and cultural assessments, laboratory skills, and the diagnosis and management of common childhood illnesses and behavioral problems

Role Development

First NP program established in 1964 prepared pediatric nurse practitioners (PNP) through collaborative efforts of Loretta C. Ford, EdD, RN and Henry K. Silver, MD at the University of Colorado

1. PNP role development provided a model for other emerging nurse practitioner (NP) specialties
2. Original support of PNP role as "physician extender" to improve access concerns due to shortage of primary care providers
3. Most early PNP education occurred within certificate and/or continuing education programs; e.g., Colorado program included 4 months didactic study followed by 18 months clinical practicum training
4. Early research focused on quality of care, cost-effectiveness, productivity, clinical decision-making skills, and role satisfaction of the PNP
5. National Association of Pediatric Nurse Practitioners (NAPNAP) organized in 1973 to establish PNP practice guidelines
6. Early resistance to NP role as too much of a "medical model" from mainstream graduate nursing education that focused on "nursing model" of clinical nurse specialist (CNS) role development
7. During 1980 to 1989 more physicians resulted in less need for nurse practitioner (NP)
8. During 1990 to 1998 increased emphasis on primary care resulted in decreased need for specialty care; NP seen as viable, cost effective member of healthcare delivery team
9. The role of the PNP continues to expand, and now involves secondary and tertiary care; PNPs now practice in a variety of settings

Conceptual Models for Advanced Practice Nursing

1. Benner's model of expert practice (1985)
2. Calkin's model of advanced nursing practice (1984)
3. Shuler's model of NP practice (1993; 1998)
 - a. Holistic patient needs
 - b. NP/Patient interaction
 - c. Self-care
 - d. Health prevention
 - e. Health promotion
 - f. Wellness
4. Consensus Model for Future Regulation of APRN Practice (2008)

Advanced Practice Trends and Issues

- Components of Advanced Practice Registered Nursing Role
 1. Coordinator of care
 2. Patient advocate
 3. Accountable for patient outcomes and cost effectiveness
 4. Direct care giver
 5. Educator
 6. Administrator
 7. Researcher
 8. Consultant
 9. Case manager
 10. Change agent

- Standards and Scope of Practice
 1. Standards of practice
 - a. Described by ANA (1998) as authoritative statements by which to measure quality of practice, service, or education
 - b. Establishes minimum levels of acceptable performance
 - c. Provides consumer with means to measure quality of care received
 - d. Both generic and specific specialty standards exist
 - e. Specialty groups have also developed standards, including—National Association of Pediatric Nurse Practitioners (NAPNAP), Association for Women’s Health, Obstetric, and Neonatal Nurses (AWHONN), formerly NAACOG
 - f. PNP relevant standards of practice
 - (1) American Nurses Association (ANA) Maternal-Child Health (MCH) Standards—first published in 1983
 - (2) NAPNAP Standards—first published in 1987, most recently updated 2004
 - (3) AWHONN Standards
 - g. Can be used to provide legal expectations of practice but were not designed to define standards of practice for clinical or legal purposes
 2. Scope of practice
 - a. Based on what is legally allowable in each state under its Nurse Practice Act
 - b. Provides guidelines vs. specific mandates for nursing practice
 - c. Is not mandated
 - d. Varies widely from state to state and over time
 - e. Often based on legal requirements within state and national standards
 - f. NAPNAP first published Scope of Practice for PNPs in 1983 with updated statements published in 1990, 2000, 2004
 3. Nurse practice acts
 - a. Authorizes Boards of Nursing in each state to establish statutory authority for licensure of registered nurse (RN)
 - b. Authority includes use of title, authorization for scope of practice, and disciplinary grounds
 - c. Evolves from statutory law which, after interpretation, becomes regulatory language
 4. Clinical practice guidelines or protocols
 - a. Definition: “Systematically developed statements to assist practitioner and patient about appropriate care for specific clinical outcomes” (IOM, 1990)
 - b. Needs/requirements for guidelines/protocol development
 - (1) “The PNP uses a framework for practice that incorporates both scientific and theoretical bases
 - (2) The scope of healthcare services and standards of practice provided by PNPs are impacted by state Nurse Practice Acts, licensure and regulatory mechanisms, work setting privileges, and/or credentialing and collaborative agreements where necessary NAPNAP, 2004.” (NAPNAP, 2004)
 - (3) Variable requirements depending on individual state nurse practice act and standards of practice
 - (4) Protocol requirements may be met with recognized reference books and published clinical guidelines
 - c. Examples of pediatric-related practice guidelines for preventive care
 - (1) Bright futures (MCHB)
 - (2) Guidelines for Adolescent Preventive Services (AMA)
 - (3) Guide to Clinical Preventive Services
 - d. Examples of pediatric-related practice guidelines for illness management
 - (1) Asthma (NIH, AAP)
 - (2) Hearing screening (NIH, AAP)
 - (3) HIV—Agency for Healthcare Research and Quality (AHRQ), formerly Agency for Health Care Policy and Research (AHCPR)
 - (4) Otitis media with effusion (AHRQ)
 - (5) Pain (AHRQ)
 - (6) Sickle Cell disease (AHRQ)

g. *Chart Overview of Nurse Practitioner Scopes of Practice in the United States*, compiled by the Center for the Health Professions, UCSF (2007)

Regulation of Advanced Nursing Practice

1. Credentialing—regulatory mechanism(s) to insure accountability for competent practice
 - a. Mandates accountability/responsibility for competent practice
 - b. Validation of required education, licensure, and certification
 - c. Necessary to assure public of safe health care provided by qualified individuals
 - d. Necessary to assure compliance with federal and state laws related to nursing practice
 - e. Acknowledges APRN advanced scope of practice
 - f. Should provide appropriate avenues for public or individual practice complaints
 - g. Allows profession to be accountable to public and its members by enforcing professional standards for practice
 - h. Past tension between certification bodies, State Boards of Nursing, and nursing education accrediting organizations regarding role and responsibility for credentialing is being addressed by the development of a mechanism named “LACE” that enhances the communication and transparency among APRN licensure, accreditation, certification, and education bodies
 - i. National task force on quality nurse practitioner education updated in 2006 to 2008 with broad-based representation
 - (1) Cofacilitator—National Organization of Nurse Practitioner Faculties (NONPF)
 - (2) Cofacilitator—American Association of Colleges of Nursing (AACN)
 - (3) American Academy of Nurse Practitioners (AANP)
 - (4) American Nurses Credentialing Center (ANCC)
 - (5) Commission on Collegiate Nursing Education (CCNE)
 - (6) National Association of Nurse Practitioners in Women’s Health
 - (7) Association of Faculties of Pediatric Nurse Practitioners
 - (8) Pediatric Nursing Certification Board
 - (9) The National Certification Corporation for the Obstetric, Gynecologic and Neonatal Nursing Specialties
 - (10) National League for Nursing Accrediting Commission
2. Licensure is the granting of authority to practice
3. Prescriptive authority (Byrne, 2008)
 - a. Physician signature on prescriptions written by NP is NOT required in any state
 - b. Controlled substance prescriptions must include the NP’s name and DEA number
 - c. As of 1998, all states have approved and/or implemented some degree of prescriptive authority
 - d. Required pharmacology education within graduate program and continuing education to maintain authority—specific requirements vary by state
 - e. Scope of prescriptive authority varies by state; full scope includes ability to obtain federal DEA registration number
4. Multistate Nurse Licensure Compact
 - a. Since 1998, 17 states have passed legislation to recognize nursing RN licensure among participating states
 - b. In 2000, Wisconsin became the first state to include advanced practice nurses within legislation on multistate compacts
 - c. NCSBN and NP stakeholders developed a position paper in 2000 to guide multistate recognition of APRN licenses/authority to practice
 - d. Current and detailed information available at the National Council of State Boards of Nursing: http://www.ncsbn.org/regulation/nlc_licensure_aprn.asp
5. Clinical privileges
 - a. Possibility of hospital staff membership opened to nonphysician providers by Joint Commission on Accreditation of Health Care Organizations (JCAHO) in 1983
 - b. Current issue for APRN practice
6. Accreditation is the formal review and approval by a recognized agency of educational degree or certification programs in nursing or nursing-related programs
7. Certification
 - a. Definition—the formal recognition of the knowledge, skills, and experience demonstrated by the achievement of standards identified by the profession
 - b. Purpose—to assure the public that an individual has mastered a body of knowledge and acquired skills in a particular specialty
 - c. May be required for state licensure and reimbursement
 - d. Required for APRN practice in most states
 - e. NP credentialing currently available through National Commission for Certifying Agencies (NCCA) recognized certifying agencies, <http://www.noca.org>
 - (1) Pediatric Nursing Certification Board (PNCB)—formerly NCBPNPN
800 South Frederick Avenue, Suite 104
Gaithersburg, MD 20877-4250

- 1 (888) 641-2767
<http://www.pncb.org/ptistore/control/index>
 (a) PNP-PC (Primary Care)
 (b) PNP-AC (Acute Care)
- (2) American Nurses Credentialing Center (ANCC)
 8515 Georgia Avenue, Suite 400
 Silver Spring, MD 20910-3492
 1 (800) 284-2378
<http://www.nursecredentialing.org/>
 (a) Pediatric Nurse Practitioner—PNP-BC
 (b) Family Nurse Practitioners
 (c) Adult Nurse Practitioner
 (d) Several types of other nurse practitioner specialties
- (3) American Academy of Nurse Practitioners (AANP)
 Certification Administration
 PO Box 12846
 Austin, TX 78711
 (512) 442-4262
<http://www.aanp.org/AANPCMS2>
 (a) Family Nurse Practitioners
 (b) Adult Nurse Practitioners
- (4) National Certification Corporation
 PO Box 11082
 Chicago, IL 60611-0082
<http://www.nccwebsite.org/>
 (a) Neonatal Nurse Practitioner (NNP)
 (b) Womens Health Nurse Practitioner (WHNP)
- (5) Oncology Nursing Certification Corporation
 125 Enterprise Drive
 Pittsburgh, PA 15275
 1 (877) 769-ONCC
<http://www.oncc.org/>
 (a) Advanced Oncology Certified Nurse Practitioner (AOCNP)

8. Education

- By 1989, 85% federally funded NP programs were graduate level
- Although there was a recent trend for blended roles of CNS and NP within graduate education, these roles have been clearly defined by the APRN Consensus Model
- Most current programs require 2 years of full-time or 3 to 4 years of part-time graduate study

• Curriculum Guidelines and Content

- Association of Faculties of Pediatric Nurse Practitioner and Associate Programs (AFPNP/

- AP)—first published terminal competencies in 1981, later updated in 1996
- National Association of Nurse Practitioner Faculties (NONPF)—published curriculum guidelines and standards for NP education (1990) updated in 2008
 - Core graduate nursing content includes—nursing theory and research, organizational/leadership theory, ethical/legal issues, multicultural care, economics, community based care, managed care, and healthcare delivery systems
 - Broad based vs. specialty core content for advanced practice—“population focus” rather than “specialty” is the term now used to describe nurse practitioner tracks; population focus best describes the broad area of practice for which competencies exist to supplement core role preparation; specialty is a more narrow focus of practice that may include added emphasis of education after the initial broad-based education of role and population focus
 - American Association of Colleges of Nursing publishes position statement—practice doctorate should be the DNP degree, and should be the entry level for advanced nursing practice, including but not limited to the four APRN roles—nurse practitioner, nurse midwife, nurse anesthetist, and clinical nurse specialist (2004)

▣ PRACTICE ISSUES

- Collaborative practice
 - Definition—ANA's *Nursing: A Social Policy Statement* (2003) describes collaboration as “true partnership” in which all players have and value power, recognize and accept separate and combined areas of responsibility and activity, and share common goals
 - Purpose—to enhance quality of care and improve patient outcomes through ongoing continuity and coordination of care
 - Interdisciplinary teams—examples of collaborative practice
 - According to the 2008 APRN Consensus Model, Boards of Nursing should license APRNs as independent practitioners with no regulatory requirements for collaboration, direction, or supervision
- Case management
 - Definition—“A collaborative process which assesses, plans, implements, coordinates, monitors, and evaluates options

- and services to meet an individual's health needs through communication and available resources to promote quality cost-effective outcomes." (Case Management Society of America, 2008)
- b. Purpose—to mobilize, monitor, and control resources that patient uses over course of an illness while maintaining a balance between quality and cost
 - c. Components of role
 - (1) Planning care for cost effectiveness and optimal outcomes
 - (2) Procuring and coordinating care
 - (3) Monitoring and evaluating outcomes
 - (4) Performing physical assessments
 - (5) Selecting laboratory and other tests
 - (6) Prescribing
 - (7) Requires that provider have strong communication skills and clinical expertise
 - (8) Provides care along continuum, decreases fragmentation of services, enhances patient and family quality of life, and contains costs
 - d. Key features associated with case management models
 - (1) Standardized appropriate use of resources aimed at identified outcomes within appropriate time frames
 - (2) Promotes collaborative practice among disciplines
 - (3) Promotes coordinated continuity of care over course of illness
 - (4) Promotes job satisfaction for providers
 - (5) Promotes patient and provider
 - e. Populations appropriate for case management
 - (1) Those for whom course of treatment is costly and unpredictable
 - (2) Those who experience frequent or chronic readmissions to hospital
 - (3) Those involved with multiple providers or multiple disciplines
3. Quality improvement (QI)
 - a. Definition—Organized creation of beneficial change ... the attainment of unprecedented levels of performance
 - b. Alternative terms—Total Quality Management (TQM); Continuous Quality Improvement (CQI); differs from Quality Assurance (QA) in being continuous rather than episodic process
 - c. Systematic, organized structures, processes, and expected outcomes focus on defining excellence and assuring accountability for quality of care
 - d. Provides framework for ongoing evaluation of practice through identification of norms, criteria, and standards that measure program effectiveness and minimize liability
 4. Risk management
 - a. Systems and activities designed to recognize and intervene to decrease risk of injury to patients and subsequent claims against healthcare providers; based on assumption that many injuries to patients are preventable
 - b. Evaluates sources of legal liability in practice such as:
 - (1) Patients
 - (2) Procedures
 - (3) Quality of record keeping
 - c. Areas of liability risk
 - (1) Practitioner-client relationship
 - (2) Communication and informed consent
 - (3) Clinical expertise
 - (4) Self-evaluation by professionals of need to stay current
 - (5) Documentation
 - (6) Consultation and referral
 - (7) Policies, procedures, and protocols
 - (8) Supervision of others
 - d. Includes educational activities that decrease risk in identified areas
 5. Malpractice
 - a. Professional misconduct, unreasonable lack of skill; infidelity in professional or fiduciary duties; illegal, immoral conduct resulting in patient harm
 - b. Alleged professional failure to render services with degree of care, diligence, and precaution that another member of same profession in similar circumstances would render to prevent patient injury

- c. Malpractice insurance
- (1) Does not protect APRN from charges of practicing medicine without a license if APRN is practicing outside legal scope of practice for that state
 - (2) National Practitioner Data Bank collects information on adverse actions against healthcare practitioners, including nurses
 - (3) Types of coverage
 - (a) Occurrence coverage—covers malpractice event which occurred during policy period, regardless of date of discovery or when claim filed
 - (b) Claims made coverage—covers only claims filed during policy coverage period, regardless of when event occurred; optional tail coverage contract extends the coverage of a claims-made policy into the future to cover all claims filed after the basic claims-made coverage period
6. Negligence—failure of individual to do what a reasonable person would do that results in injury to another
7. Reimbursement: Whether working independently, sharing a joint practice with a physician, or practicing within a hospital or managed care system, APRNs must be reimbursed appropriately. Standards that determine private pay insurance mechanisms are often modeled after federal policies such as Medicaid and Medicare. However, even when the federal government establishes mandates that encourage direct payment of nonphysician healthcare providers, barriers to reimbursement are often encountered in state level rules and regulations (Hamric, 2000). National survey data indicate that nearly half of all major managed care organizations in the United States refuse to credential nurse practitioners as primary care providers (Hansen-Turton, Ritter, & Torgan, 2008).
- a. Medicaid
- (1) Authorized in 1965 as Title XIX of Social Security Act
 - (2) Federal/state matching program with federal oversight
 - (3) Financed through federal and state taxes, with between 50% and 83% of total Medicaid costs covered by federal government
 - (4) Does not cover all people below federal poverty level, but state Medicaid programs are required by federal government to cover certain categories such as:
 - (a) Recipients of Aid to Families with Dependent Children (AFDC)—states set own eligibility requirements for AFDC
 - (b) People over 65, blind, or totally disabled who are eligible for cash assistance under federal Supplemental Security Income (SSI) program
 - (c) Pregnant women (for pregnancy-related services only) and children under six with family incomes up to 133% of federal poverty level
 - (d) Children born after September 1983 up to age 19 in families whose income is at or below federal poverty level
- (5) States can choose to cover “medically needy”
- (6) Coverage required for certain services
 - (a) Hospital and physician services
 - (b) Laboratory and radiographic services
 - (c) Nursing home and home health-care services
 - (d) Prenatal and preventive services
 - (e) Medically necessary transportation
- (7) States can add services to list and can place certain limitations on federally mandated services
- (8) Although Medicaid recipients cannot be billed for services, states can impose nominal copayments or deductibles for certain services
- b. Medicare
- (1) Federally mandated program established in 1965, provides health insurance for aged and disabled individuals, and people of all ages with end stage renal disease.
 - (2) Eligibility covers hospital service, physician services, and other medical services
 - (3) Income level does not impact eligibility
 - (4) Medicare Part A
 - (a) Those 65 years of age and older who are eligible for Social Security are automatically enrolled, whether or not they are retired—persons are eligible for Social Security when they (or their spouses) have paid into Social Security system through

- employment for 40 quarters or more
- (b) Those who have paid into system for less than 40 quarters can enroll in Medicare Part A by paying monthly premium
- (c) Those who are under age 65 and are totally and permanently disabled may enroll in Medicare Part A after receiving Social Security disability benefits for 24 months
- (d) Those with chronic renal disease requiring dialysis or transplant may also be eligible for Part A without a two-year waiting period
- (e) Services covered include some hospitalization costs; some skilled nursing facility costs, although custodial care is not covered; home health care—100% for skilled care; 80% of approved amount for medical equipment; and hospice care—100% for most services
- (f) Payment for hospitalization is based on projected costs of caring for patient with given problem; each Medicare patient admitted to a hospital is classified according to a diagnosis-related group (DRG); the hospital is then paid a predetermined amount for each patient admitted with the given DRG, if hospital costs are above payment rate, the hospital must absorb loss; if costs are below payment rate, hospital allowed to keep a percentage of excess
- (g) APRN not paid directly for services delivered in a hospital
- (5) Medicare Part B—Supplementary Medical Insurance (SMI)
 - (a) Monthly premium is charged
 - (b) Some low-income people are eligible to have monthly premium paid by Medicaid
 - (c) Financed by general federal revenues and by Part B monthly premiums
 - (d) Covers all medically necessary services—80% of an approved amount after annual deductible; includes physician services, physical, occupational, and speech therapy; medical equipment and diagnostic tests, and some preventative care such as Pap tests, mammograms, hepatitis B, pneumococcal and influenza vaccines can be included in medical expenses
- c. APRN—Medicaid/Medicare coverage
 - (1) Omnibus Budget Reconciliation Act (OBRA) 1989—mandated Medicaid reimbursement for certified pediatric and family nurse practitioners began July 1, 1990; providers required to practice within the scope of state law and do not have to be under supervision or associated with a physician or other provider
 - (a) Level of payment determined by states—reimbursement rates range from 70% to 100% of fee-for-service physician Medicaid rate (Pearson, 2003)
 - (b) Pediatric and family nurse practitioners may bill Medicaid directly after attaining provider number from state Medicaid agency
 - (c) States can elect to pass laws allowing them to extend Medicaid payment to other types of NP not identified in federal statutes
 - (2) Legislation (1997) has expanded direct Medicare reimbursement for APRN in all geographic locations
 - (a) APRN reimbursement at 85% of physician fee schedule when billing independently using APRN billing number; direct physician supervision not required
 - (b) When APRN is employed by physician, the physician practice may receive 100% of customary physician charge, according to “Incident to” rules (Buppert, 1998)
 - (c) APRN must be RN currently licensed to practice in the state in which services are rendered; must meet requirements for NP practice in state in which services are rendered; must be currently certified as a primary care NP; must have successfully completed a formal advanced practice educational program of at least one academic year that includes at least four months of classroom instruction and awards a degree, diploma, or certificate OR, have successfully completed a formal advanced practice educational program and have been

- performing in that expanded role for at least 12 months during the 18-month period immediately preceding February 8, 1978, the effective date for the provision of services of NP as reflected in the conditions for certification for rural health clinics
- (d) NP covered services are limited to services an NP is legally authorized to perform under the state law in which the NP practices and must meet training, educational, and experience requirements prescribed by the Secretary of Health and Human Services
- (3) NP services covered under Part B if service would be considered physician's services if furnished by MD or doctor of osteopathy (DO); if NP is legally authorized to perform services in the state in which they are performed; if services are performed in collaboration with MD/DO (collaboration specified as a process whereby NP works with physician to deliver health care within scope of NP expertise with medical direction and appropriate supervision as provided for in jointly developed guidelines or other mechanisms defined by federal regulations and law of the state in which services are performed); and services are otherwise precluded from coverage because of one of the statutory exclusions
- (4) "Incident to" refers to services provided as an integral, yet incidental, part of the physician's personal, professional services in the course of diagnosis or treatment of injury or illness—these services must occur under direct personal supervision of a physician, and the APRN must be an employee of the physician group; services must occur during the course of treatment where the physician performs an initial service and subsequent services in a manner that reflects the physician's active participation and management of the course of treatment; direct personal supervision does not mean that the physician must be in the same room as the APRN, however, the physician must be present in the office suite and available for assistance and direction while the APRN provides patient care (Buppert, 1998)
- (5) When APRN performs "incident to" service in physician's office, billing must be submitted to Medicare by employing physician, under the physician's name, provider number, and CPT code; payment is made at full physician rate and is paid to physician or physician practice
- (6) When APRN provides service in skilled nursing facility, or nursing facility located in urban area as defined by law, Medicare payment can be obtained; Medicare reimbursement is also available for APRN services in skilled nursing facilities (SNF) in nonrural areas on a reasonable charge basis; this amount may not exceed physician fee schedule amount for service and payment is made to the APRN's employer
- (7) Centers for Medicare and Medicaid Services (CMS)
- (a) Formerly Health Care Financing & Administration (HCFA)
- (b) Oversight of several federal programs including Medicare, Medicaid, State Children's Health Insurance Program (SCHIP), HIPPA, and CLIA
- (c) <http://cms.hhs.gov>
- d. Other third party payers
- (1) Private insurer reimbursement is contract specific per state insurance commission
- (2) Civilian Health and Medical Program of the United States (CHAMPUS)
- (a) Federal health plan for military personnel, including surviving dependents, families, and retirees
- (b) APRN reimbursement for services
- (3) Federal Employees Health Benefit Program (FEHBP)
- (a) One of largest employer sponsored group health insurance programs
- (b) APRN recognized as designated health care provider
- e. Methods of payment for advanced practice nurses
- (1) Fee-for-service model
- (a) Unit of payment by visit or procedure
- (b) Can occur with utilization review, in which case payer has right to authorize or deny payment of

- expensive medical interventions such as hospital admission, extra hospital days, and surgery
- (2) Episodic model
 - (a) One sum is paid for all services delivered during a given illness
 - (b) DRG fee payment
- (3) Capitation model, PPO, and HMO are covered in section on managed care in this chapter

■ PROFESSIONAL ORGANIZATIONS

1. Purpose and benefits
 - a. Establish practice standards
 - b. Collective voice to promote nursing and quality of care
 - c. Monitor and influence policy and legislative initiatives
 - d. Position papers on practice issues
 - e. Disseminate information
 2. Examples
 - a. American Nurses Association (ANA)
 - b. National Association of Pediatric Nurse Practitioners (NAPNAP)
 - c. Gerontological Advanced Practice Nurses Association (GAPNA) (formerly the National Conference of Gerontological Nurse Practitioners)
 - d. National Organization of Nurse Practitioner Faculties (NONPF)
 - e. American Academy of Nurse Practitioners (AANP)
 - f. American College of Nurse Practitioners (ACNP)
 - g. Nurse Practitioner Associates for Continuing Education (NPACE)
 - h. National Association of School Nurses
 - i. Association for Women's Health, Obstetrics, and Neonatal Nurses (AWHONN)
 - j. The National Association of Nurse Practitioners in Women's Health (NPWH) (formerly the National Association of Nurse Practitioners in Reproductive Health [NANPRH])
- Research in Advanced Practice: "Practice-based research is essential to the development of advanced practice nursing for the future." (NONPF, 1995, p. 84)
 1. Major trend is outcome studies
 2. Sources of federal funding
 - a. Agency for Healthcare Research and Quality (AHRQ)
 - (1) Formerly the Agency for Health Care Policy and Research (AHCPR)
 - (2) <http://www.ahrq.gov/>

- b. National Institutes of Health (NIH)
 - (1) Includes the National Institute for Nursing Research (NINR)
 - (2) <http://www.nih.gov/>
 - c. Maternal and Child Health Bureau (MCHB)
 - (1) Functions within Health Resources and Services Administration (HRSA)
 - (2) <http://www.mchb.hrsa.gov/>
3. Sources of research findings
 - a. Conferences
 - b. Scholarly publications
 - c. Distribution of summaries of research studies
 4. Use of research in practice setting
 - a. Develop research-based clinical pathways
 - b. Track clinical outcomes and variances
 - c. Demonstrate quality and cost effectiveness of care
 - d. Give structure to demonstration projects
 - e. Persuade lawmakers of NP value and contributions in today's healthcare system
 - f. Improve quality and patient outcomes
 5. Benefit of research for patients
 - a. Provides thorough understanding of patient situation
 - b. Provides more accurate assessment of situations
 - c. Increases effectiveness of interventions
 - d. Increases provider sensitivity to patient situations
 - e. Assists providers to more accurately determine need for and effectiveness of interventions
 6. Barriers to research utilization
 - a. Time and cost of conducting research studies
 - b. Resistance to change in work setting
 - c. Lack of rewards for using research findings
 - d. Lack of understanding or uncertainty regarding research outcomes
 7. Strategies to overcome barriers to research utilization
 - a. Creation of organizational culture that values and uses research
 - b. Creation of environment where questions are encouraged, critical thinking is appreciated, and nursing care is evaluated
 - c. Support for research through time allocation and financial commitment

■ HEALTH POLICY

- Policy Influences
 1. Healthy People 2000
 - a. Published in 1990 by U.S. Department of Health and Human Services; midcourse review of progress published in 1995;

- subsequent review of progress published in December 1999
- b. Purpose—committed nation to obtain three broad goals
 - (1) Increase span of healthy life for all Americans
 - (2) Reduce health disparities among Americans
 - (3) Achieve access to preventive services for all Americans
 - c. Contained 300 specific objectives based on 22 priority areas leading to socially and economically productive lives
 - d. Objectives focused on equal access, acceptability, availability, continuity, cost, and quality of care
 - e. Objectives organized within broad categories of health promotion, health protection, and preventive services
 - f. Identified priority of systematic collection, analysis, interpretation, dissemination, and use of data to understand national health status and plan effective prevention programs
 - g. Individuals, communities, and organizations were responsible for determining how they would achieve the goals by the year 2000
 - h. Progress (1998–1999) towards meeting Healthy People 2000 goals
 - (1) Fifteen percent of objectives met in areas of nutrition, maternal and child health, heart disease, and mental health
 - (a) There were 17 maternal/infant health objectives; progress made on 8 of the 17, including:
 - (i) Perinatal/infant mortality—although U.S. ranks 25th among industrialized countries for infant mortality
 - (ii) Screening for fetal abnormalities and genetic disorders
 - (b) No progress on four or movement away on five of the target goals for problems including fetal alcohol syndrome and low birthweight
 - (2) Forty-four percent of objectives, including those related to childhood immunizations, breastfeeding, regular dental visits, mammography screening, and consumption of fruits and vegetables per day are proceeding on track
2. Healthy People 2010
 - a. Released in 2000 by U.S. Department of Health and Human Services
 - b. Builds on initiatives set in Healthy People 2000
 - c. Purpose—designed to achieve two broad-based goals
 - (1) Increase quality and years of healthy life
 - (2) Eliminate health disparities
 - d. Contains 467 objectives with twenty-eight focus areas
 - e. Objectives focus on partnering for health improvements, eliminating health disparities, increasing quality and years of healthy living, and harnessing technology for health
 - f. Objectives categorized into the following focus areas:
 - (1) Physical activity and fitness
 - (2) Nutrition
 - (3) Tobacco use
 - (4) Educational/community-based programs
 - (5) Environmental health
 - (6) Food safety
 - (7) Injury/violence prevention
 - (8) Occupational safety and health
 - (9) Oral health
 - (10) Access to quality health services
 - (11) Family planning
 - (12) Maternal, infant, and child health—fetal, infant, child, and adolescent deaths; maternal deaths and illnesses; prenatal care; obstetrical care; risk factors; developmental disabilities and neural tube defects; prenatal substance exposure; breastfeeding, newborn screening, and service systems for children with special healthcare needs
 - (13) Medical products safety
 - (14) Public health infrastructures
 - (15) Health communication
 - (16) Prevention and health promotion
 - (17) Disability and secondary conditions
 - (18) Heart disease and stroke
 - (19) Kidney disease
 - (20) Mental health and mental disorders
 - (21) Respiratory diseases
 - (22) Sexually transmitted diseases
 - (23) Substance abuse
 - g. Ten “leading health indicators” (LHI)—physical activity, overweight and obesity, tobacco use, substance abuse, responsible sexual behavior, mental health, injury and violence, environmental quality, immunizations, access to health care

- h. Written for states and communities to tailor health objectives to their specific needs
<http://www.healthypeople.gov>
- 3. Prevention guidelines
 - a. “Put Prevention Into Practice”—campaign established by U.S. Public Health Service to enhance delivery of preventive care in primary care practice
www.ahrq.gov/Clinic/ppipix.htm
 - b. *Guide to Clinical Preventive Services* (U.S. Preventive Services Task Force, 2008) presents national clinical preventive services guidelines for practice and educational settings
<http://www.ahrq.gov/clinic/pocketgd.htm>
 - (1) Age and gender specific
 - (2) Suggests targeted examinations, immunizations and health counseling that should be part of periodic health visits
- 4. Nursing’s Agenda for Health Care Reform (ANA, 2008)
<http://198.65.150.241/readroom/rnagenda.htm>
 - a. Supports a shift from an emphasis on illness and cure to an orientation towards wellness and care
 - b. Calls for primary care in community settings
 - c. Fosters consumer responsibility for personal health
 - d. Facilitates utilization of the most cost-effective providers and therapeutic options in the most appropriate settings
 - e. A federally-defined standard package of essential healthcare services available to all citizens and residents of the United States, provided and financed through an integration of public and private plans and sources
 - f. Cost-effective phased-in essential services, including coverage of services for pregnant women and children, and a “Health Start” plan to assist vulnerable populations
 - g. Planned change to anticipate health service needs that correlate with changing national demographics
 - h. Case management for those with continuing health needs
 - i. Certain provisions for long-term care
 - j. Insurance reforms to provide access to care and reduce excessive costs
 - k. Access to services assured by no payment at the point of service and elimination of balance billing in both public and private plans.

- l. Establishment of public/private sector review operating under federal guidelines and including payers, providers, and consumers; to determine resource allocation, cost reduction approaches, allowable insurance premiums, and fair and consistent reimbursement levels for providers; this review would progress in a climate sensitive to ethical issues

Utilization of Health Policy

- 1. Shifting trend toward primary care and early preventive measures; supports need for APRN
- 2. Four major factors influencing healthcare delivery services
 - a. Payers—individual healthcare consumers, businesses that pay for health insurance for employees, and government through public programs and entitlement programs such as Medicare and Medicaid
 - b. Insurers—take money from payers, assume risks, and pay providers
 - c. Providers—includes hospitals, physicians, nurses, APRN, physician assistants, pharmacies, home health agencies, and long-term care facilities
 - d. Suppliers—pharmaceutical and medical supply industries
- 3. Legislative strategies and political involvement
 - a. Professional organizations monitor policy issues and keep membership informed—e.g., NAPNAP legislative newsletter
 - b. Local networks of APRN develop practice guidelines and advocate for policies to enhance practice

■ TYPES OF HEALTHCARE DELIVERY SYSTEMS

- 1. Primary health care
 - a. Definition: “Primary care is the provision of integrated, accessible healthcare services by clinicians who are accountable for addressing a large majority of personal healthcare needs, developing a sustained partnership with patients, and practicing in the context of family and community” (Institute of Medicine, Committee on Future of Primary Care, 1994)
 - b. Activities and/or functions define boundaries of primary care, such as curing or alleviating common illnesses and disabilities
 - c. Entry point to a system that includes access to secondary and tertiary care

- d. Attributes include care that is accessible, comprehensive, coordinated, continuous, and accountable
 - e. Strategy for organizing healthcare system as a whole; gives priority and allocates resources to community-based rather than hospital-based care
 - f. Categories of primary care providers (PCP) and nature of care
 - (1) Medical specialties—family medicine, general internal medicine, general pediatrics, obstetrics, and gynecology
 - (2) Other experts have included NP and physician assistants (PA) as primary care providers (PCP)
 - g. Many definitions stress self-responsibility for health
2. Managed care
- a. Definition: “An integrated network that combines financing and delivery of healthcare services to covered individuals.” (Mahn & Spross, 1996)
 - (1) Network connects consumers, sponsors, providers, and third party payers
 - (2) Initial managed care organization was Kaiser Health Plan (California) established in 1930s
 - b. Objectives
 - (1) Manage use and price of health care delivery system
 - (2) Control type, level, and frequency of treatment
 - (3) Restrict level of reimbursement for services
 - c. Type of health insurance plan designed to control costs while assuring quality care
 - d. Obligation to manage is shared among providers, consumers, and payers
 - (1) Providers no longer dictate price of care delivery; must assume more financial risk for population assigned to them for care
 - (2) Consumers have fewer choices of coverage, providers and greater financial responsibility
 - (3) Payers manage healthcare dollars through benefit design, selective contracting, and shifting financial risk to providers
 - e. Types of managed care plans
 - (1) Health Maintenance Organizations (HMO)
 - (a) Most common type
 - (b) By 1994, enrollment at 52 million (20.3% of population)
 - (c) Offer pre-established benefit package—including preventive, inpatient, and outpatient care
 - (d) HMO contracts with providers to provide care to enrollees
 - (e) Providers at financial risk resulting in incentive to provide high quality, cost effective care
 - (f) Enrollees select a primary care provider (PCP) who manages total care by authorizing specialty visits, hospitalization, and other services
 - (g) PCP may be MD, APRN or PA providers; serving as “gate keepers”
 - (2) Preferred Provider Organizations (PPO)
 - (a) Compromised managed care option that is alternative between indemnity and HMO insurance
 - (b) Uses financial incentives to influence consumer and provider behaviors
 - (c) Refers to variety of arrangements between insurers, providers, and third-party payers rather than standard plan
 - (d) Often owned by large insurance companies such as Prudential, Travelers, and Aetna
 - (e) Available primarily to employed commercial population
 - (3) Point of Service Plans (POS)
 - (a) Consumers decide whether to use a provider network, or seek care outside the network
 - (b) If variation of HMO plan, PCP coordinates care for enrollees; if variation of PPO plan, enrollees may choose lower cost options outside of provider network
 - (c) Most rapidly growing type of managed care
 - (4) Integrated delivery systems
 - (a) Vertical integration of services across levels of care into seamless system with improved access for enrollees
 - (b) Capitated payment—financial risk shifts from payor to provider; unit of value is cost per member per month (PMPM); providers receive age and sex adjusted budget to cover services to maintain wellness of specific target population
 - (c) Emphasis on provision of appropriate but not unlimited care with

- financial benefit of keeping population healthy through systematic preventive services
- f. Reimbursement under managed care
 - (1) Providers accept financial risk for care provided to specific population of enrollees
 - (2) Capitated payment
 - (a) Provider receives payment in advance
 - (b) Payment level reflects expected utilization by enrolled population for which provider is responsible
 - (3) Provider must control volume and cost
 - (4) Efficiency usually rewarded through bonus payments for operating within budget and meeting goals for quality and efficiency
 - g. Monitoring, evaluation, and accreditation in managed care
 - (1) Health plan employer data and information set (HEDIS)—provides quality measures and compares with benchmark standards and goals
 - (2) National committee for quality assurance (NCQA)
 - (a) Major accreditation body for managed care organizations
 - (b) Standards in six critical areas are evaluated—quality management, utilization management, credentialing, preventive health services, medical records, member's rights/responsibilities
 - (3) NCQA reported reviews of 30% of the 554 HMOs in the U.S. as of 1995; 31% received full accreditation; 56% received one year or provisional accreditation; 12% were denied accreditation (Mischler & Quinn, 1995)
 - h. Challenges and opportunities of managed care for APRN
 - (1) Need for balance between quality of care and costs inherent in diagnosis/management per client visit
 - (a) Educational programs must incorporate managed care content into curriculum
 - (b) APRN must combine strong clinical and financial skills to determine cost of providing care to target population
 - (c) Success in managed care environment requires systems-thinking skill to complement primary care skills; blended NP/CNS models may provide this necessary

- linkage with added focus on case management, utilization/resource management, quality improvement, and client education/advocacy within systems of care
- (2) APRN strategies for success within evolving managed care environment
 - (a) Determine strategies to increase efficiency without sacrificing quality of client-provider interactions; e.g., group well-child visits
 - (b) Lobby for APN inclusion on provider panels
 - (c) Maintain partnerships with APRN educational programs for collaborative study and documentation of APRN effectiveness

■ HEALTH INSURANCE PORTABILITY AND ACCOUNTABILITY ACT OF 1996 (HIPPA)

1. Health Insurance Reform—addresses preexisting conditions and provides portability of health insurance when an employee changes or loses their job
2. Administrative simplification per Centers for Medicare and Medicaid Services (CMS) (2003)
 - a. Electronic transactions and code set standards
 - b. Privacy requirements
 - c. Security requirements
 - d. National identifier requirements
3. CMS responsible for oversight and compliance with administrative simplification mandates except for privacy provisions which is overseen by the Office of Civil Rights (OCR)
4. Target dates for implementation and compliance with Administrative Simplification
 - a. Electronic transactions and code set standards—October 2003
 - b. Unique identifiers—July 2004
 - c. Privacy—April 2003
 - d. Security—April 2005
5. “Covered entities”
 - a. Health plans
 - b. Healthcare clearinghouses
 - c. Healthcare providers who conduct any electronic transitions of health related information
6. Website information
 - a. Center for Medicare and Medicaid Services, HIPPA information for Health Care Providers’ Offices:
<http://www.cms.hhs.gov/EducationMaterials/Downloads/HIPAA101-1.pdf>

- b. U.S. Department of Health and Human Services, The Office for Civil Rights
web site re: HIPPA:
<http://www.hhs.gov/ocr/privacy/index.html>

■ QUESTIONS

Select the best answer

1. If the two broad-based goals of Healthy People 2010 are met, the nation will:
 - a. Increase quality and years of healthy life span, and eliminate health disparities
 - b. Increase healthy life span, increase access, and improve economics related to health for all Americans
 - c. Increase healthy life span, reduce health disparities, and improve economics related to the health of all Americans
 - d. Reduce health disparities, increase access to healthcare services, and make life more socially acceptable for all Americans
2. Which of the following objectives from Healthy People 2000 has not made progress towards achieving its targeted goal?
 - a. Low birthweight
 - b. Infant mortality
 - c. Fetal mortality
 - d. Screening for genetic disorders
3. Preventive health guidelines include references to:
 - a. Immunizations, health screening, disease prophylaxis, education, and infection control
 - b. Immunizations, counseling, health screening, disease prophylaxis, and education
 - c. Health screening, disease prophylaxis, counseling, and CPR
 - d. Health screening, disease prophylaxis, education, immunizations, and CPR
4. Nursing's Agenda for Health Care Reform:
 - a. Is supportive of equal access, cost effective, high quality care
 - b. Is a mandate to all nurses in the U.S.
 - c. Is a summary of nursing research related to healthcare reform
 - d. Is a report of the status of nursing in the 1990s
5. The nurse practitioner role was initially established to:
 - a. Improve access to care and partially solve physician shortage
 - b. Reduce the nursing shortage and improve access to care
 - c. Improve working conditions of nurses while improving access to care
 - d. Improve nursing's image through expansion of the role
6. Early nursing research focused on:
 - a. The response of policy makers to the nursing shortage
 - b. The effectiveness of the NP as a primary care giver
 - c. An effort to demonstrate quality and cost effectiveness of NP
 - d. The role of the NP as a physician extender
7. Which of the following is not a major factor influencing healthcare delivery services?
 - a. Provider
 - b. Payers
 - c. Insurers
 - d. Agencies
8. All definitions of primary health care include:
 - a. The concept of universal access and accountability
 - b. The concept of universal access and AIDS prevention
 - c. The concept of universal access and a focus on self-responsibility for health
 - d. The concept of universal access and a focus on reimbursement for services rendered
9. Standards of practice are:
 - a. Authoritative statements used to measure quality
 - b. Used to measure outcome but are not authoritative
 - c. Designed for legal purposes
 - d. Not designed for legal purposes and cannot be used to measure quality
10. Quality improvement activities include:
 - a. Patient satisfaction surveys only
 - b. Peer review, patient satisfaction surveys, chart audits
 - c. Defining four practice domains
 - d. Systems to decrease risk of injury to patients

11. Most risk management programs are based on the assumption that:
 - a. Many injuries to patients are preventable
 - b. Most legal liability is a result of poor documentation
 - c. Most injuries to patients are not preventable
 - d. Malpractice insurance is generally unnecessary
12. If an APRN practices beyond his/her scope:
 - a. Malpractice insurance will protect him/her from a charge of practicing medicine without a license
 - b. Malpractice insurance will not protect him/her from a charge of practicing medicine without a license
 - c. He or she is legally accountable to the certifying body
 - d. The collaborating physician is legally accountable to the certifying body
13. Standards of practice may be used to:
 - a. Establish minimal levels of performance
 - b. Establish reimbursement schemes for APRN
 - c. Mandate nursing practice across the nation
 - d. Mandate nursing practice in certain states
14. Scope of practice:
 - a. Is identical across the states
 - b. Is determined by the federal government
 - c. Is mandated by the federal government
 - d. Varies from state to state
15. Medicaid provides health insurance coverage to:
 - a. Certain categories of people whose personal income falls below the federal poverty level
 - b. Anyone whose personal income falls below the federal poverty level
 - c. Newborns, pregnant women, and those over 65 whose personal income falls below the federal poverty level
 - d. Those who are elderly
16. Medicaid reimbursement is available to an APRN:
 - a. Practicing in federally designated areas
 - b. At a rate that is between 70% and 100% of the physician rate
 - c. Only if the APRN is in collaborative practice with a physician
 - d. Practicing in nursing homes only
17. Medicare reimbursement for services:
 - a. Is not dependent on the patient's income level
 - b. Depends on the patient's income level
 - c. Is not available to APRN under any circumstances
 - d. Is only available to APRN who is in collaborative practice with a physician
18. Medicare Part A covers:
 - a. Hospital, skilled nursing facility, and hospice care
 - b. All medically necessary services
 - c. Skilled nursing facility care only
 - d. Hospice care only
19. Medicare Part B covers:
 - a. All medically necessary services
 - b. Inpatient hospital care
 - c. Outpatient physician services only
 - d. Skilled nursing facility and hospice care
20. To receive Medicare reimbursement, APRN must:
 - a. Be nationally certified and maintain prescriptive privileges
 - b. Maintain a current license in the state in which they are practicing
 - c. Practice in a designated medically underserved area
 - d. Practice with a physician
21. The term "incident to" refers to:
 - a. The occasions when an APRN practices independently but occasionally consults with a physician
 - b. The notion that the physician must be present in the office suite and immediately available to provide assistance in order for the APRN to bill for services rendered
 - c. The notion that a physician must examine the patient along with the APRN if Medicare is to be billed for services rendered
 - d. Medicaid only and is not pertinent to Medicare billing
22. "Incident to" billing is specific to:
 - a. Medicare
 - b. Medicaid
 - c. Medicare and Medicaid
 - d. Private insurance companies

23. The Civilian Health and Medical Program of the United States (CHAMPUS):
- Is a federal health plan which covers health care for military personnel and their families and recognizes APRN as reimbursable provider
 - Is a federal health plan which covers health care for military personnel and recognizes APRN as reimbursable provider
 - Is a federal health plan which covers health care for military personnel and their families but does not recognize APRN as reimbursable provider
 - Only covers hospital expenses of military personnel and their families
24. The knowledge base of the APRN is based on:
- Medical content
 - Theoretical content only
 - Scientific content and theory
 - Theory and research
25. The role of the APRN has traditionally focused on:
- The delivery of primary health care to all people
 - The delivery of acute health care to all people
 - Chronic care
 - The medical model
26. The nurse practitioner role began:
- With the establishment of a pediatric nurse practitioner program in an effort to expand the role of the registered nurse in order to meet the needs of the children of the nation
 - As a result of the new entitlement programs, Medicare and Medicaid
 - When it was evident that the medical schools across the U.S. could not prepare enough family practitioners to meet the nation's need
 - As an experimental program at Duke University Medical Center
27. Legal authority for APRN practice is granted by:
- Federal law
 - Regulations from the Department of Health and Human Services
 - State law and regulations
 - The Board of Medicine in most states
28. Direct reimbursement to APRN has resulted in:
- Increased access to cost-effective, quality primary care
 - Increased malpractice claims against APRN
 - Decreased consumer choice of healthcare providers
 - Proliferation of APRN in independent practice
29. Malpractice insurance:
- Protects an APRN from charges of practicing medicine without a license when they are practicing outside the legal scope of practice
 - Does not protect an APRN from charges of practicing medicine without a license when they are practicing outside the legal scope of practice
 - Does not pay for legal defense if the APRN is practicing beyond the legal scope of practice
 - Is important, but should not be purchased if the facility in which the APRN is employed carries good coverage
30. Collaborative practice:
- Limits autonomy and not reasonable in current managed care environment
 - Will enhance quality of care and improve patient outcomes
 - Will limit consumer choice of providers
 - Excludes the concept of interdisciplinary teams
31. Case management:
- Balances quality and cost of patient care
 - Has not been found to be cost effective
 - Decreases the autonomy of the APRN
 - Is rarely used today
32. The major trend in health policy research today is:
- Outcome studies
 - Primary care studies
 - Studies that compare practice strategies of MD and NP
 - Studies that compare patient satisfaction with care delivered by MD versus NP
33. Current prescriptive authority for APRN:
- Varies among the states
 - Is fairly consistent among the states
 - Provides DEA numbers for APRN
 - Allows APRN to move freely from state to state

34. Managed care is a term that describes:
- An established system of healthcare delivery that is mandated by the federal government
 - A network of providers who contract to provide services for a specific group of enrollees
 - A system that does not recognize APRN as a primary provider
 - A network of hospitals and nursing homes that provide care to chronically ill people
35. Certification is:
- A procedure through which the government appraises and grants certification to the APRN
 - Granted by the individual states
 - Governed by each state's Board of Nursing
 - A process in which a nongovernmental agency or group verifies that an APRN has met certain predetermined standards for specialty practice
36. Licensure:
- Is a federal process that is used to standardize healthcare facilities
 - Is granted by a state government agency and grants permission to engage in the practice of a given profession
 - Cannot be used to prohibit anyone from practicing a given profession
 - Is a federal process that is used to standardize educational programs
37. Reimbursement under managed care:
- Requires that the provider accept the financial risk for the care provided to a specific population of enrolled patients
 - Requires that the managed care organization accept the financial risk for the care provided to a specific population of enrolled patients
 - Does not reward efficient care delivery
 - Is not available to APRN
38. An integrated delivery system:
- Is one that delivers high quality care but is often not cost effective
 - Delivers a vertical integration of services with capitated payment
 - Does not include rationing of resources
 - Does not include a capitated payment scheme

39. The APRN Consensus Model is:
- A mandate from the NCSBN that defines advanced practice nursing
 - A proposed regulatory model for advanced practice nursing
 - A proposal for federal legislation for advanced practice nursing
 - Approved only by the American Nurses Association

■ ANSWERS

- | | |
|-------|-------|
| 1. a | 21. b |
| 2. a | 22. a |
| 3. b | 23. a |
| 4. a | 24. c |
| 5. a | 25. a |
| 6. c | 26. a |
| 7. d | 27. c |
| 8. a | 28. a |
| 9. a | 29. b |
| 10. b | 30. b |
| 11. a | 31. a |
| 12. b | 32. a |
| 13. a | 33. a |
| 14. d | 34. b |
| 15. a | 35. d |
| 16. b | 36. b |
| 17. a | 37. a |
| 18. a | 38. b |
| 19. a | 39. b |
| 20. b | |

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Index

A

- abdomen, normal findings, 51
 - abdominal migraine, 204
 - abdominal pain, acute, 202–203
 - abdominal pain, chronic, 203–204
 - ABO incompatibility, 299
 - abstinence (contraceptive method), 288
 - abuse and neglect, 22–23
 - adolescent pregnancy and, 293
 - accreditation, 376
 - acne vulgaris, 168–169
 - “acquired” hypogammaglobulinemia, 314
 - acquired immune deficiency syndrome (AIDS), 350–351
 - acquired syphilis, 283–285
 - acrocyanosis, 53
 - ACTH, 254*t*
 - acute abdominal pain, 202–203
 - acute infectious gastroenteritis, 195–199
 - causative agents, U.S. most common, 197*t*
 - acute myocardial inflammatory disease, 129–130
 - acute nasopharyngitis (common cold), 111
 - acute otitis media (AOM), 97–98
 - perforation of tympanic membrane, 100
 - acute perforations of ear, 100
 - Adam’s forward-bending test, 239
 - adenovirus, 195, 195*t*
 - ADH (antidiuretic hormone). *See* diabetes insipidus
 - ADHD. *See* Attention Deficit Hyperactivity Disorder
 - adolescents
 - growth and development, 16–17
 - health maintenance and promotion, 37–80
 - child health supervision, 37–38
 - example questions and answers, 77–80
 - history, 38–42
 - laboratory tests and values, 54–56
 - normal findings and common variations, 44–53
 - physical examination, 42–44
 - problem-oriented health records, 62
 - schedule of visits, 62–76
 - screening and assessment tests, 57–59
 - special examinations, 59–62
 - history, 42
 - physical examination, 43, 43*t*, 44*t*
 - schedule of visits, 75–76
 - pregnancy, 290–293. *See also* contraception
 - example questions and answers, 293–298
- adrenal gland disorders, 332–335
 - adrenocortical hyperfunction, 333–335
 - adrenocortical insufficiency, 332–333
 - adrenogenital syndrome, 334–335
- advanced practice nursing, 373–391
 - about, 373–377
 - example questions and answers, 387–390
 - Health Insurance Portability and Accountability Act of 1996 (HIPPA), 386–387
 - health policy, 382–384
 - practice issues, 377–382
 - professional organizations, 382
 - types of healthcare delivery systems, 384–386
- Ages and Stages Questionnaires (ASQ), 32
- aggression, 25–26
- AIDS (acquired immune deficiency syndrome), 350–351
- Ainsworth, Mary, 9
- airway. *See* lower respiratory disorders
- albinism, 157–158
- alcohol, 30. *See also* substance abuse
 - adolescent pregnancy and, 292
 - fetal alcohol syndrome (FAS) and fetal alcohol spectrum disorders (FASD), 358–359
- allergic contact dermatitis, 162–163
- allergic hypersensitivity reactions. *See* hypersensitivity conditions, dermatologic
- allergic rhinitis, 103–104
- allergies, history of, 40
- Allis sign, 52
- alpha (α) thalassemia, 301–302
- amaurosis (blindness), 92–93
- amblyopia (lazy eye), 93–94
- amenorrhea (menstrual disorders), 342–343
- amphetamines, 30
- anabolic steroids, 31
- analysis-level questions, 4
- ANCC examination, overview of, 3*t*
- anemia, screening for, 59
- animism, 14
- anorexia nervosa, 27–28
- anovulatory cycles, 16
- ant stings, 178–179
- antibiotic therapy for otitis media, 99*t*
- antidiuretic hormone (ADH). *See* diabetes insipidus
- antiepileptic drugs, 254*t*
- AOM. *See* acute otitis media
- APGAR scores, 59, 60*t*
- aphthous ulcers (canker sores), 108

- apnea, 146–147
 apparent life threatening event (ALTE), 146
 appendicitis, 202–203
 application questions, 4
 APRN. *See* advanced practice nursing
 arthritis, juvenile idiopathic, 242–244
 aryngotracheobronchitis (croup), 135–136
 assessment. *See also* screening and assessment tests
 physical growth and development, 31
 associations, professional, 382
 associative play, 15
 asthma, 142–144
 astigmatism, 92
 astrocytomas (brain), 263
 athlete's feet (tinea pedis), 175–176
 athletics, injuries from, 240–241
 atonic seizures, 252
 atopic dermatitis, 161–162
 atrioventricular block, 124
 atrioventricular canal, 121*t*
 atrioventricular septal defect, 122*t*
 attachment, 9, 13
 Attention Deficit Hyperactivity Disorder (ADHD), 23–24
 authoritarian parenting, 14
 authoritative parenting, 14
 Autism Spectrum Disorder (ASD), 19–21
 autoimmune thrombocytopenia, 309–310
 autosomal chromosomal disorders. *See* genetic syndromes
- B**
- back. *See* musculoskeletal disorders
 bacterial dermatologic conditions, 166–168
 cellulitis, 166–167
 impetigo, 167
 involving pilosebaceous unit, 168–170
 acne vulgaris, 168–169
 folliculitis and furuncles, 169–170
 staphylococcal scalded skin syndrome, 167–168
 bacterial endocarditis, prevention of, 131*t*
 bacterial vaginosis (BV), 287–288
 band cell or stab variations, 55
 Bandura, Albert, 8
 Barlow test, 52
 Barlow's sign, 234
 basophil, 55
 Bayley Infant Neurodevelopmental Screener (BINS), 32
 Bayley Scales of Infant Development, Second Edition (BSID-II), 32
 bedbug bites, 176–177
 bee stings, 178–179
 behavior
 aggression, defiance, and disruptive behaviors, 25–26
 history, 40
 screening and assessment tests, 32
 behaviorism, 8
 beta (β) thalassemia, 301–302
- bilirubin
 neonatal hyperbilirubinemia, 300–301
 normal values, 56
 Binet, Alfred, 7
 binocular vision, 47
 birth issues with adolescents, 290–293
 birthmarks (nevi), 53
 capillary hemangioma (strawberry nevus), 155–156
 port-wine stain, 155
 salmon patch (nevus simplex), 154–155
 bites, cat. *See* cat scratch disease
 bites, insect, 176–178
 tick bites. *See* lyme disease; Rocky Mountain spotted fever
 black widow spider bites, 177–178
 blank slate, child as, 8
 bleeding disorders, 307–310
 hemophilia, 307–308, 308*t*
 idiopathic thrombocytopenia (ITP, autoimmune thrombocytopenia), 309–310
 von Willebrand Disease (VWD), 308–309
 bleeding from nose (epistaxis), 104–105
 blepharitis (granulation of eyelids), 87–88
 blindness (amaurosis), 92–93
 blocked tear duct (dacryostenosis), 46, 86–87
 blood. *See* hematologic disorders
 blood glucose
 hyperglycemia, 336–339
 hypoglycemia, 335–336
 normal values, 56
 blood pressure
 hypertension, 125–126
 norms, 44, 44*t*
 blood urea nitrogen (BUN), 56
 Bloom's taxonomy of cognition, 2–3
 bones. *See* musculoskeletal disorders; osteosarcoma
 Bordetella pertussis. *See* pertussis
 Borrelia burgdorferi. *See* lyme disease
 Bowlby, John, 9
 bowleg (genu varum), 236–237
 BPD. *See* bronchopulmonary dysplasia
 brachial cleft cyst, 45
 bradycardia, 124
 brain stem gliomas, 263
 brain tumors, 263–264
 breastfeeding
 anticipatory guidance and interventions, 67–68
 hospital newborn visit guidance, 64–65
 infant guidance, 70
 newborn guidance, 66–67
 prenatal guidance, 63–64
 breastmilk jaundice, 300
 breasts
 development in females, 16
 normal findings, 50–51
 physical examination, 61
 breath sounds, 49
- bronchiolitis, 137–138
 bronchitis, 137
 bronchopulmonary dysplasia (BPD), 144–145
 Bronfenbrenner, Urie, 10
 brown recluse spider bites, 177–178
 Bruton's tyrosine kinase (BTK), 314–315
 BTK (Bruton's tyrosine kinase), 314–315
 bulbar conjunctiva, 46
 bulimia nervosa, 27–28
 BUN. *See* blood urea nitrogen
 burns, 164–166
 sunburns, 165–166
- C**
- Café au lait Spots, 156
 Campylobacter jejuni, 196*t*, 197, 198
 cancers, 310–314
 example questions and answers, 316–318
 leukemias, 310–311
 lymphomas, 312–313
 neuroblastoma, 311
 osteosarcoma, 313–314
 retinoblastoma, 91, 311–312
 Wilms' tumor (nephroblastoma), 313
 canker sores (aphthous ulcers), 108
 capillary hemangioma (strawberry nevus), 155–156
 caput succedaneum, 45, 83
 carbamazepine, 254*t*
 cardiovascular disorders, 119–134
 acute myocardial inflammatory disease, 129–130
 congenital heart disease/defects, 119–120, 121*t*, 122*t*
 congestive heart failure (CHF), 120–122, 122*t*
 disturbances of rate and rhythm, 123–125
 example questions and answers, 132–133
 hypercholesterolemia, 126–127
 hypertension, 125–126
 Kawasaki disease (mucocutaneous lymph node syndrome), 127–129
 murmurs, 49, 122–123
 rheumatic fever/heart disease, 130–131
 case management, 377–378
 cat scratch disease (CSD), 222
 cataracts, 89–90
 causative agents, U.S. most common, 195–196*t*
 CBC with differential, 54–55
 CD (Crohn's disease). *See* inflammatory bowel disease
 Celiac disease, 201–202
 cell count (CSF test), 56
 cellulitis, 166–167
 central apnea, 146
 central diabetes insipidus (DI), 326–328
 centration, 14
 cephalohematoma, 45, 84
 cerebrospinal fluid (CSF)

- hydrocephalus, 45, 82–83
tests, 56
- certification, about, 376–377
- certification, self-assessment for, 1
- certification test questions, about, 2–5. *See also* example questions and answers
- certification test taking strategies, 1–6
- cervical lymphadenitis, 113
- CF. *See* cystic fibrosis
- chalazion, 87
- Chandler, Michael, 10
- changing American family, growth and development and, 17–18
- chemical injuries to eye, 96
- chemistry tests and findings, 55–56
- CHF. *See* congestive heart failure
- chickenpox (varicella-zoster virus [VZV]), 220–221
- chigger bites, 176–177
- child health maintenance. *See* health maintenance and promotion
- child health supervision, 37–38
- childhood depression, 28–29
- chlamydia, 282–283
- chlamydia trachomatis, 351–352
- cholesteatoma, 101
- cholesterol
hypercholesterolemia, 126–127
hypertension and, 125–126
screening, 56, 59
- chromosomal disorders. *See* genetic syndromes
- chronic abdominal pain, 203–204
- chronic diarrhea, 201
- chronic perforations of ear, 100
- chronic rhinitis, 104
- CHS. *See* Current Health Status
- classic developmental theories and theorists, 7–9
- classical conditioning theory, 8
- cleft lip/palate, 107
- clinical privileges, 376
- clonazepam, 254*t*
- clonic seizures, 252
- Clostridium botulinum*. *See* infant botulism
- Clostridium difficile*, 196*t*, 197, 199
- Clostridium tetani*. *See* tetanus (lockjaw)
- clubfoot (talipes equinovarus congenita), 235
- CMV (cytomegalovirus), 355
- coarctation of the aorta, 121*t*, 122*t*
- cocaine, 30
- cognitive development
adolescents, 17
infants (birth to 2 years), 11–12
school-aged children, 15
screening and assessment tests, 32
toddlers and preschoolers, 14
- cognitive domain, 7
- cognitive levels of questions, 2–4
- cold sore (herpes labialis), 108–109, 170–171
- collaborative practice, 377
- combined hormonal contraceptives, 289
- common cold (acute nasopharyngitis), 111
- common insect bites, 176–177
- common variable immunodeficiency (CVID, “acquired” hypogammaglobulinemia), 314
- communication in child health supervision, 37–38
- complete heart block (CHB), 124
- comprehension questions, 3–4
- conceptual models for advanced practice nursing, 374
- concussion, 260
- condoms (male), 288
- conduct behaviors, 25
- conductive hearing loss, 101–102
- condylomata acuminata (genital warts), 286–287
- confidentiality issues in child health supervision, 37–38
- congenital diseases, 350–359
acquired immune deficiency syndrome (AIDS), 350–351
chlamydia trachomatis, 351–352
congenital rubella syndrome (CRS), 356
congenital syphilis, 353–354
cytomegalovirus (CMV), 355
example questions and answers, 369–371
fetal alcohol syndrome (FAS) and fetal alcohol spectrum disorders (FASD), 358–359
gonococcal infection, 352–353
hepatitis B, 356–357
herpes, 354–355
toxoplasmosis, 357–358
- congenital heart disease/defects, 119–120, 121*t*, 122*t*
- congenital rubella syndrome (CRS), 356
- congestive heart failure (CHF), 120–122, 122*t*
- conjunctivitis, 46, 84–86
of childhood, 85–86
of newborn (ophthalmia neonatorum), 84–85
- constipation, 205–206
- contact dermatitis (allergic), 162–163
- contact irritant dermatitis (diaper dermatitis), 163–164
- contextual developmental theories and theorists, 9–10
- contraception, 288–290
- contusion, head, 260
- cooperative play, 15
- corneal abrasion, 94–95
- CP (cerebral palsy), 366–367
- CRAFFT Tool for Adolescents, 42
- craniosynostosis, 84
- creatinine, 56
- credentialing, 376
- Crohn’s disease (CD). *See* inflammatory bowel disease
- croup (aryngotracheobronchitis), 135–136
- CRS (congenital rubella syndrome), 356
- cryptorchidism (undescended testes), 272–273
- Cryptosporidium parvum*, 196*t*, 197, 199
- CSD (cat scratch disease), 222
- CSF tests, 56
- Current Health Status (CHS), 40
- curriculum in advanced nursing practice, 377
- Cushing’s syndrome (hypercortisolism), 333–335
- cutis marmorata, 53, 153
- CVID (common variable immunodeficiency), 314
- cystic fibrosis (CF), 140–142
- cytomegalovirus (CMV), 355
- D**
- dacryostenosis (blocked tear duct), 46, 86–87
- DAI (diffuse axonal injury), 260
- Darwin, Charles, 7
- DDH (developmental dysplasia of the hip), 234–235
- defiance, 25–26
- dehydration, 53
assessment of, 197*t*
- demographic changes, American family, 17
- dental caries, 107–108
- dental development and care, 11, 16*t*
guidance
infants, 69
toddlers and preschoolers, 73
normal findings, 48
- Denver II test, 32
- depigmentation. *See* pigmentation conditions
- depression, 28–29. *See also* mental health
- dermatitis conditions, 161–164
atopic dermatitis, 161–162
contact dermatitis (allergic contact dermatitis), 162–163
contact irritant dermatitis (diaper dermatitis), 163–164
seborrhea dermatitis, 164
- dermatologic conditions, 153–184
bacterial conditions, 166–170
burn conditions, 164–166
dermatitis conditions, 161–164
example questions and answers, 184–190
fungal conditions, 172–176
hypersensitivity conditions, miscellaneous, 181–184
insect conditions, 176–181
melanocyte cell and pigmentation conditions, 156–159
newborn exanthema, 153–154
papulosquamous conditions, 159–161
vascular lesions, 154–156
viral conditions, 170–172

- dermatologic system
 differential diagnosis, 192*f*
 normal findings, 52–53
 terminology for, 191*t*
- development. *See* human growth and development
- “development in context”, 10
- developmental dysplasia of the hip (DDH), 234–235
- developmental theories and theorists, 7–10
 classic, 7–9
 transactional and contextual, 9–10
- diabetes insipidus (DI), 326–328
- diabetes mellitus (DM), 336–339
- diaper dermatitis, 163–164
- diaphragm (contraceptive method), 288–289
- diarrhea, chronic, 201
- diffuse axonal injury (DAI), 260
- DiGeorge syndrome (thymic hypoplasia), 315
- diphtheria, 214–215
- disciplinary behavior. *See* behavior disorders
- cardiovascular. *See* cardiovascular disorders
- dermatologic. *See* dermatologic conditions
- ears. *See* ears, disorders of
- endocrine. *See* endocrine disorders
- eyes. *See* eyes, disorders of
- gastrointestinal. *See* gastrointestinal disorders
- genetic. *See* genetic syndromes
- genitourinary. *See* genitourinary disorders
- gynecological. *See* gynecological disorders
- hematologic. *See* hematologic disorders
- immunologic. *See* immune deficiencies
- infectious diseases. *See* infectious diseases
- lower respiratory. *See* lower respiratory disorders
- mouth. *See* mouth, disorders of
- multisystem. *See* multisystem disorders
- musculoskeletal. *See* musculoskeletal disorders
- neurological. *See* neurological disorders
- nose. *See* nose, disorders of
- oncologic. *See* cancers
- throat. *See* throat, disorders of
- disorders of pubertal development, 339–341
- disruptive behavior disorders, 25–26
- disturbances of rate and rhythm, 123–125
- DM (diabetes mellitus), 336–339
- DMD (duchenne muscular dystrophy), 245–246
- domains of human growth and development, 7
- domains of practice, 3*t*
- Down syndrome (DS) (trisomy 21), 360–361
- dramatic or pretend play, 15
- drug abuse. *See* substance abuse
- drug eruptions (dermatologic), 181–182
- DS (Down syndrome), 360–361
- duchenne muscular dystrophy (DMD), 245–246
- dysmenorrhea, 277–278
- E**
- early adolescence, 17
- ears. *See also* hearing disorders, 96–103
 acute otitis media (AOM), 97–98
 cholesteatoma, 101
 chronic/acute perforations, 100
 example questions and answers, 114–117
 external otitis media (EOM), 96–97
 hearing loss, 47, 101–103
 mastoiditis, 100–101
 otitis media with effusion (OME), 99
 tympanostomy tubes, 99–100
 normal findings, 47
- eating disorders, 27–28
- EC (emergency contraception), 290
- ecchymoses (“black eye”), 96
- ecological model of development, 10
- education in advanced nursing practice, 377
- Edwards Syndrome (trisomy 18), 359–360
- EI (erythema infectiosum), 219–220
- egocentrism, 14, 17
- elective termination of pregnancy, 291–292
- elimination
 guidance, 72
 history, 40
- emergency contraception (EC), 290
- emotional abuse. *See* abuse and neglect
- encopresis, 205
- endocarditis, bacterial, prevention of, 131*t*
- endocrine disorders, 321–348
 adrenal gland disorders, 332–335
 adrenocortical hyperfunction, 333–335
 adrenocortical insufficiency, 332–333
 disorders of pubertal development, 339–341
 example questions and answers, 344–347
 growth disturbances, 328–330
 excessive growth, 330–332
 short stature, 328–330
 gynecomastia, 341–342
 hyperglycemia, 336–339
 hypoglycemia, 335–336
 menstrual disorders (amenorrhea), 342–343
 obesity, 343–344
 pituitary disorder—diabetes insipidus (DI), 326–328
 thyroid disorders, 321–326
 hyperthyroidism, 323–325
- hypothyroidism, 321–323
 thyroiditis, 325–326
- Entamoeba coli, 196*t*, 197, 199
- Entamoeba histolytica, 196*t*, 197, 199
- enterobiasis vermicularis (pinworms), 199
- enuresis, 271–272
- environmental history, 39–40
- EOM. *See* external otitis media
- eosinophil, 55
- ependymomas (brain), 263
- epiglottitis (supraglottitis), 113–114
- epilepsy, 251–255
- epistaxis, 104–105
- Erikson, Erik, 8
- erythema infectiosum (fifth disease), 219–220
- erythema multiforme major (Stevens-Johnson syndrome), 183
- erythema multiforme minor, 182–183
- erythema toxicum, 53
- erythema toxicum neonatorum, 153–154
- ethologic theories of human growth and development, 8–9
- ethosuximide, 254*t*
- example questions and answers
 advanced practice nursing, 387–390
 cardiovascular disorders, 132–133
 dermatologic conditions, 184–190
 endocrine disorders, 344–347
 gastrointestinal disorders, 208–211
 genitourinary/gynecological disorders and adolescent pregnancy, 293–298
 head, eyes, ears, nose, mouth, throat disorders, 114–117
 health promotion, 77–80
 hematologic/oncologic/immunologic disorders, 316–318
 human growth and development, 32–35
 infectious diseases, 229–231
 lower respiratory disorders, 149–151
 multisystem and genetic disorders, 369–371
 musculoskeletal disorders, 246–249
 neurological disorders, 264–267
- exanthem subitum (roseola), 219
- excessive growth, 330–332
- expressive language, 14
- external otitis media (EOM), 96–97
- eyelids, 46
- eyes, 84–96. *See also* vision disorders of, 84–94
 amblyopia (lazy eye), 93–94
 blepharitis (granulation of eyelids), 87–88
 blindness (amaurosis), 92–93
 cataracts, 89–90
 chalazion, 87
 conjunctivitis of childhood, 85–86
 conjunctivitis of the newborn (ophthalmia neonatorum), 84–85

- dacryostenosis (blocked tear duct), 46, 86–87
- example questions and answers, 114–117
- glaucoma, 90
- hordeolum (stye), 88
- nystagmus, 91
- orbital/periorbital cellulitis, 88–89
- refractive errors, 91–92
- retinoblastoma, 91
- strabismus, 90–91
- injuries to, 94–96
- normal findings, 46–47
- F**
- face, normal findings, 46
- failure-to-thrive (FTT), 18–19
- family functioning, 17–18
- family history (FH), 41
- family structure, variations in, 17
- farsightedness (hyperopia), 91–92
- FAS (fetal alcohol syndrome), 358–359
- FASD (fetal alcohol spectrum disorders), 358–359
- febrile seizures, 255
- feet. *See* musculoskeletal disorders
- female social development, 9
- females
- vs. males. *See* gender differences
 - puberty. *See* puberty
- feminizing adrenal tumors, 334–335
- fetal alcohol spectrum disorders (FASD), 358–359
- fetal alcohol syndrome (FAS), 358–359
- fever blister (herpes labialis), 108–109, 170–171
- FH. *See* family history
- fibroadenoma, 50
- fibrocystic breasts, 50–51
- fifth disease (erythema infectiosum), 219–220
- fine motor development milestones, 12*t*, 14*t*
- fire ant stings, 178–179
- First Step test, 32
- flea bites, 176–177
- flu. *See* influenza
- folliculitis and furuncles, 169–170
- fontanel, 44–45
- foreign body
- aspiration of, 136–137
 - eye injury, 95
 - nose injuries, 104
- Fragile X syndrome (FXS), 361–362
- functional dyspepsia, 204
- functional gastrointestinal disorders, 203–205
- abdominal migraine, 204
 - chronic abdominal pain, 203–204
 - functional dyspepsia, 204
 - irritable bowel syndrome (IBS), 204–205
- fungal dermatologic conditions, 172–176
- tinea capitis (ringworm of the scalp), 172–173
- tinea corporis (ringworm of the body), 173–174
- tinea cruris (jock itch), 174–175
- tinea pedis (athlete's feet), 175–176
- furuncles and folliculitis, 169–170
- FXS (Fragile X syndrome), 361–362
- G**
- G-6-PD deficiency, 306–307
- galactorrhea, 50
- Galeazzi's sign, 234
- gastroesophageal reflux disease (GERD), 193
- gastroesophageal reflux in infancy (GER), 193–194
- gastrointestinal disorders, 193–211
- acute abdominal pain, 202–203
 - acute infectious gastroenteritis, 195–199
 - assessment of dehydration, 197*t*
 - causative agents, U.S. most common, 197*t*
 - chronic diarrhea, 201
 - constipation, 205–206
 - example questions and answers, 208–211
 - functional, 203–205
 - gastroesophageal reflux in infancy (GER), 193–194
 - hepatitis, 206–208, 356–357
 - hernia, 208
 - inflammatory bowel disease (IBD), 199–200
 - malabsorption, 201–202
 - pinworms (enterobiasis vermicularis), 199
 - pyloric stenosis, 194–195
- gender differences. *See also* females; males in moral development, 9
- gender identity, 15
- generalized seizures, 252, 255
- genetic syndromes, 359–366
- about, 349–350
 - Down syndrome (DS) (trisomy 21), 360–361
 - example questions and answers, 369–371
 - Fragile X syndrome (FXS), 361–362
 - Klinefelter's syndrome (XXY karyotype), 363
 - Marfan syndrome, 364–365
 - mucopolysaccharidosis I (MPS IH), 365–366
 - Prader-Willi syndrome, 366
 - Tay-Sachs disease, 363–364
 - trisomy 18 (Edwards Syndrome), 359–360
 - Turner's syndrome (XO karyotype), 362–363
- genital herpes simplex virus (HSV), 285–286
- genitourinary/gynecological disorders, 269–298, 274. *See also* contraception
- cryptorchidism (undescended testes), 272–273
- dysmenorrhea, 277–278
- enuresis, 271–272
- example questions and answers, 293–298
- genitourinary trauma, 279
- glomerulonephritis (GN), 279–280
- hydrocele, 273
- hydronephrosis, 280
- hypospadias, 273–274
- labial adhesions, 275–276
- meatal stenosis, 274–275
- menstrual disorders (amenorrhea), 342–343
- premenstrual syndrome, 278–279
- renal tubular acidosis (RTA), 280–281
- sexually transmitted infections (STIs), 281–288
- acquired syphilis, 283–285
 - bacterial vaginosis (BV), 287–288
 - chlamydia, 282–283
 - genital herpes simplex virus (HSV), 285–286
 - genital warts (condylomata acuminata), 286–287
 - gonorrhoea, 281–282
 - screening, 59
 - trichomoniasis, 287
- STIs. *See* sexually transmitted infections
- testicular torsion, 275
- trauma, 279
- urinary tract infection (UTI), 269–271
- vulvovaginitis, 276–277
- genu valgum (knock-knee), 237
- genu varum (bowleg), 236–237
- GER (gastroesophageal reflux in infancy), 193–194
- GERD (gastroesophageal reflux disease), 193
- german measles (rubella), 217–218
- Gesell, Arnold, 7–8
- gestational age, weight assessment by, 10
- Giardia lamblia, 196*t*, 197, 198–199
- Gilligan, Carol, 9
- glaucoma, 90
- global development screening and assessment, 32
- glomerulonephritis (GN), 279–280
- glucose
- hyperglycemia, 336–339
 - hypoglycemia, 335–336
 - normal values, 56
- glucose-6-phosphate dehydrogenase (G-6-PD) deficiency, 306–307
- GN (glomerulonephritis), 279–280
- gonococcal infection, 352–353
- gonorrhoea, 281–282
- Gower's sign, 52
- graduate education, advanced nursing practice, 377
- grand mal (tonic-clonic) seizures, 252, 255
- granulation of eyelids (blepharitis), 87–88

- great arteries, transposition of, 121*t*
 gross motor development milestones, 12*t*, 14*t*
 growing pains, 238
 growth. *See* human growth and development
 growth disturbances, 328–330
 excessive growth, 330–332
 short stature, 328–330
 gynecological disorders. *See* genitourinary/
 gynecological disorders
 gynecomastia, 50, 341–342
- H**
- hair, pubic, 16–17
 Hall, G. Stanley, 7
 hallucinogens, 31
 hand, foot, and mouth disease, 109
 harlequin color change, 53
 Harlow, Harry, 8
 HAV. *See* hepatitis
 HBV. *See* hepatitis
 HCV. *See* hepatitis
 HDV. *See* hepatitis
- head
 disorders of, 81–84
 caput succedaneum, 45, 83
 cephalohematoma, 45, 84
 craniosynostosis, 84
 example questions and answers, 114–117
 hydrocephalus, 45, 82–83
 macrocephaly, 45, 82
 microcephaly, 45, 81–82
 injuries, 240–241, 260–262
 normal findings, 44–45
 normal growth, 81*t*
 head tilt, 45
 headaches, 255–257
 Health Insurance Portability and
 Accountability Act of 1996 (HIPPA),
 386–387
 health maintenance and promotion, 37–80
 child health supervision, 37–38
 example questions and answers, 77–80
 history, 38–42
 laboratory tests and values, 54–56
 normal findings and common variations,
 44–53
 physical examination, 42–44
 problem-oriented health records, 62
 schedule of visits, 62–76
 screening and assessment tests, 57–59
 special examinations, 59–62
 Health Maintenance Organizations
 (HMOs), 385
 health policy, 382–384
 health supervision, 31–32
 healthcare delivery systems, types of,
 384–386
 Healthy People 2010, 383–384
 Healthy People 2000, 382–383
- hearing. *See also* ears
 development of, 11
 screening, 57
 hearing loss, 47, 101–103, 102
 heart
 disorders. *See* cardiovascular disorders
 normal findings, 48–49
 hematocrit, normal values, 54
 hematologic disorders, 299–316
 ABO incompatibility, 299
 bleeding disorders, 307–310
 hemophilia, 307–308, 308*t*
 idiopathic thrombocytopenia
 (ITP, autoimmune
 thrombocytopenia), 309–310
 von Willebrand Disease (VWD),
 308–309
 example questions and answers, 316–318
 glucose-6-phosphate dehydrogenase
 (G-6-PD) deficiency, 306–307
 hemoglobinopathies, 301–302
 iron deficiency anemia, 302–303
 lead poisoning (plumbism), 305–306
 neonatal hyperbilirubinemia, 300–301
 Rh incompatibility, 299–300
 sickle cell disease, 303–305, 303*t*, 304*t*
 hematology test, 54–55
 hematoma (head), 260
 hemoglobin
 hemoglobinopathies, 301–302
 normal values, 54
 sickle cell disease, 303–305, 303*t*, 304*t*
 hemophilia, 307–308, 308*t*
 hepatitis, 206–208, 356–357
 heredity-environment interactions, 9–10
 hernia, 208
 herpangina, 109
 herpes, 354–355
 genital herpes simplex virus, 285–286
 herpes labialis (cold sore, fever blister),
 108–109, 170–171
 herpes simplex stomatitis, 108–109
 HEV. *See* hepatitis
 hierarchy of needs (Maslow), 9
 HIPPA (Health Insurance Portability and
 Accountability Act of 1996), 386–387
 hips. *See* musculoskeletal disorders
 Hirschsprung's disease, 205
 history, 38–42
 history of present illness (HPI), 39–40
 HIV (human immunodeficiency virus). *See*
 also sexually transmitted infections
 acquired immune deficiency syndrome
 (AIDS), 350–351
 screening, 59
 hives (urticaria), 184
 HL (Hodgkin's lymphoma), 312–313
 Hodgkin's lymphoma (HL), 312–313
 home environment, screening and
 assessment tests, 32
 hordeolum (stye), 88
 hornet stings, 178–179
- hospital newborn visit breastfeeding
 guidance, 64–65
 hostile aggression, 15
 HPI. *See* history of present illness
 HSV. *See* herpes
 human growth and development, 7–36
 by age group
 infants, 10–13
 toddlers and preschoolers, 13–15
 school-aged, 15–16
 adolescents, 16–17
 changing American family and,
 17–18
 developmental theories and theorists,
 7–10
 example questions and answers, 32–35
 growth disturbances, 328–330
 excessive growth, 330–332
 short stature, 328–330
 head, normal, 81*t*
 health supervision and, 31–32
 physical, deviations in, 18–31
 abuse and neglect, 22–23
 aggression, defiance, and disruptive
 behaviors, 25–26
 Attention Deficit Hyperactivity
 Disorder (ADHD), 23–24
 Autism Spectrum Disorder (ASD),
 19–21
 childhood depression, 28–29
 eating disorders, 27–28
 failure-to-thrive (FTT), 18–19
 learning disabilities (LD), 26–27
 obesity, 21–22
 substance abuse, 30–31
 suicidal behavior, 29–30
 humanistic theories of human growth and
 development, 9
 hydrocele, 273
 hydrocephalus, 45, 82–83
 hydronephrosis, 280
 hyperaldosteronism, 334–335
 hyperbilirubinemia, neonatal, 300–301
 hypercholesterolemia, 126–127
 hypercortisolism (Cushing's syndrome),
 333–335
 hyperglycemia, 336–339
 hyperopia (farsightedness), 91–92
 hyperpigmentation. *See* pigmentation
 conditions
 hypersensitivity conditions, dermatologic,
 181–184
 drug eruptions, 181–182
 erythema multiforme major (Stevens-
 Johnson syndrome), 183
 erythema multiforme minor, 182–183
 urticaria (hives), 184
 hypertension, 125–126
 hyperthyroidism, 323–325
 hyphema, 95
 hypoglycemia, 335–336
 hypogonadism, 340–341

- hypopigmentation. *See* pigmentation conditions
- hypoplastic left heart syndrome, 122*t*
- hypospadias, 273–274
- hypothalamic diabetes insipidus (DI), 326–328
- hypothyroidism, 321–323
- I**
- IBD (inflammatory bowel disease), 199–200
- IBS (irritable bowel syndrome), 204–205
- idiopathic scoliosis, 239–240
- idiopathic thrombocytopenia (ITP, autoimmune thrombocytopenia), 309–310
- IM (infectious mononucleosis), 225–226
- imaginary audience, 17
- immune deficiencies, 314–316
 - common variable immunodeficiency (CVID, “acquired” hypogammaglobulinemia), 314
 - example questions and answers, 316–318
 - thymic hypoplasia (DiGeorge syndrome), 315
 - Wiskott-Aldrich syndrome (WAS), 315–316
 - X-linked agammaglobulinemia (Bruton’s tyrosine kinase or BTK), 314–315
- immunization
 - guidance, 74
 - history, 40
- impetigo, 167
- imprinting, theory of, 8
- infant botulism, 226
- infantile spasms (West syndrome), 252
- infants (birth to 2 years)
 - breastfeeding guidance, 66–67
 - growth and development, 10–13
 - health maintenance and promotion, 37–80
 - child health supervision, 37–38
 - example questions and answers, 77–80
 - history, 38–42
 - laboratory tests and values, 54–56
 - normal findings and common variations, 44–53
 - physical examination, 42–44
 - problem-oriented health records, 62
 - schedule of visits, 62–76
 - screening and assessment tests, 57–59
 - special examinations, 59–62
 - maternal-infant bonding, 9
 - newborn examination and screening, 57–58, 59–60
 - parent-infant interaction, 9, 13
 - physical examination, 42, 43*t*, 44*t*
 - reflexes, 52*t*
 - schedule of visits, 65–71
- infectious diseases, 213–231
 - cat scratch disease (CSD), 222
 - diphtheria, 214–215
 - example questions and answers, 229–231
 - fifth disease (erythema infectiosum–ei), 219–220
 - infant botulism, 226
 - infectious mononucleosis (IM), 225–226
 - influenza, 216–217
 - lyme disease, 223–225
 - malaria, 228–229
 - mumps, 221–222
 - pertussis (whooping cough), 215–216
 - poliomyelitis, 226–227
 - Rocky Mountain spotted fever (RMSF), 223
 - roseola (exanthem subitum), 219
 - rubeola (german measles), 217–218
 - rubeola (red measles), 218–219
 - septicemia (sepsis), 213–214
 - tetanus (lockjaw), 227–228
 - varicella-zoster virus (VZV) (chickenpox), 220–221
- infectious gastroenteritis, acute, 195–199
 - causative agents, U.S. most common, 197*t*
- infectious mononucleosis (IM), 225–226
- infestations
 - pediculosis infestation (lice), 180–181
 - pinworms (enterobiasis vermicularis), 199
 - scabies infestation, 179–180
 - tinea capitis (ringworm of the scalp), 172–173
 - tinea corporis (ringworm of the body), 173–174
 - tinea cruris (jock itch), 174–175
 - tinea pedis (athlete’s feet), 175–176
- inflammatory bowel disease (IBD), 199–200
- influenza, 216–217
- informed consent in child health supervision, 37–38
- inguinal hernia, 208
- inhalants/volatile substances, 31
- injuries
 - to eyes, 94–96
 - genitourinary trauma, 279
 - head injuries, 260–262
 - nose injuries, 104
 - prevention of, 69–70. *See also* safety sports injuries, 240–241
- innocent heart murmurs, 49, 123
- insect dermatologic conditions, 176–179, 176–181
 - bites and stings
 - common insect bites, 176–177
 - insect stings, 178–179
 - spider bites, 177–178
 - infestations, 179–181
 - pediculosis (lice), 180–181
 - scabies, 179–180
- instrumental aggression, 15
- instrumental conditioning theory, 8
- insulin-dependent diabetes mellitus (IDDM), 336–339
- insurance, malpractice, 378–379
- integrated delivery systems, 385–386
- interactionist—structuralist stage theory of cognitive development, 8
- internal tibial torsion, 236
- interpersonal relationships, 9
- intersexuality, 339–341
- interval history, 42–43
- intracranial headaches, 256
- intrauterine device (IUD), 289
- intussusception, 202
- invincibility fable, 17
- iris, normal findings, 46–47
- iron deficiency anemia, 302–303
- irritable bowel syndrome (IBS), 204–205
- issues in advanced practice nursing, 375
- itch mites. *See* scabies infestation
- ITP (idiopathic thrombocytopenia), 309–310
- IUD (intrauterine device), 289
- J**
- jaundice, 53
- jock itch (tinea cruris), 174–175
- joints. *See* musculoskeletal disorders
- juvenile idiopathic arthritis, 242–244
- K**
- Kawasaki disease (mucocutaneous lymph node syndrome), 127–129
- Kennel, John, 9
- ketones, normal values, 56
- Klaus, Marshall, 9
- Klinefelter’s syndrome (XXY karyotype), 363
- knees. *See* musculoskeletal disorders
- knock-knee (genu valgum), 237
- knowledge-based questions, 3
- Kohlberg, Lawrence, 9
- L**
- labial adhesions, 275–276
- labial agglutination, 275–276
- labial fusion, 275–276
- laboratory tests and values, health promotion, 54–56
- lactation. *See also* breastfeeding
- lactose malabsorption, 201–202
- language development
 - infants (birth to 2 years), 13
 - milestones, 13*t*
 - screening and assessment tests, 32
 - theories, 9
 - toddlers and preschoolers, 14
- larynx. *See* lower respiratory disorders
- late adolescence, 17
- lazy eye (amblyopia), 93–94
- LCPD (Legg-Calvé-Perthes disease), 238
- LD. *See* learning disabilities
- lead poisoning (plumbism), 305–306
- lead screening, 56, 58–59
- learning disabilities (LD), 26–27
- Legg-Calvé-Perthes disease (LCPD), 238

- legs. *See* musculoskeletal disorders
- Lennox-Gastaut syndrome, 252
- leukemias, 310–311
- lice (pediculosis infestation), 180–181
- licensure, 376
- lips
- cleft lip/palate, 107
 - herpes labialis (cold sore, fever blister), 108–109
 - normal findings, 48
- Locke, John, 8
- lockjaw (tetanus), 227–228
- long-acting progestins, 289
- Lorenz, Konrad, 8
- loss theory, 9
- louse infestation, 180–181
- lower respiratory disorders, 135–151
- apnea, 146–147
 - asthma, 142–144
 - bronchiolitis, 137–138
 - bronchitis, 137
 - bronchopulmonary dysplasia (BPD), 144–145
 - croup (aryngotracheobronchitis), 135–136
 - cystic fibrosis (CF), 140–142
 - example questions and answers, 149–151
 - foreign body aspiration, 136–137
 - pneumonia, 138–140
 - tuberculosis (TB), 147–149
- lungs. *See also* lower respiratory disorders
- normal findings, 49–50
- lyme disease, 223–225
- lymph nodes
- cervical lymphadenitis, 113
 - enlarged, 45–46
 - Kawasaki disease (mucocutaneous lymph node syndrome), 127–129
 - normal findings, 53
 - retropharyngeal abscess, 112
- lymphocytes, normal values, 55
- lymphomas, 312–313
- M**
- macrocephaly, 45, 82
- Mahler, Margaret, 8
- malabsorption, 201–202
- malaria, 228–229
- male condoms, 288
- male social development, 9
- males
- vs. females. *See* gender differences
 - puberty. *See* puberty
- malignant melanoma, 157
- managed care, 385, 386
- mandibular eruption (teeth), 16*t*
- mappractice, 378–379
- Marfan syndrome, 364–365
- marijuana, 30
- Maslow, Abraham, 9
- masses
- breast (adolescent), 50–51
 - neck, 45
- mastoiditis, 100–101
- maternal deprivation experiments, 8
- maternal-infant bonding, 9. *See also* breastfeeding
- maturational-organismic theory, 7–8
- maxillary eruption (teeth), 16*t*
- maximizing study time, 5–6
- MCHC, normal values, 55
- MCV, normal values, 55
- measles
- rubella (german measles), 217–218
 - rubeola (red measles), 218–219
- meatal stenosis, 274–275
- mechanistic-learning theory, 8
- Medicaid, 379–381
- Medicare, 379–381
- medulloblastoma (brain), 263
- melanocyte cell and pigmentation
- conditions, 156–159
 - albinism, 157–158
 - café au lait spots, 156
 - malignant melanoma, 157
 - Mongolian spots, 156–157
 - pityriasis alba, 159
 - vitiligo, 158–159
- meningitis, 262–263
- menstrual disorders (amenorrhea), 342–343
- mental health
- childhood depression, 28–29
 - screening and diagnostic classifications, 32
 - substance abuse, 30–31
 - suicidal behavior, 29–30
- metatarsus adductus/metatarsus varus, 235–236
- microcephaly, 45, 81–82
- middle adolescence, 17
- migraine headaches, 256
- milia, 154
- Mischel, Walter, 8
- mites. *See* scabies infestation
- mixed apnea, 146
- mixed hearing loss, 101–102
- models for advanced practice nursing, 374
- molluscum contagiosum, 171
- Mongolian spots, 156–157
- monocytes, normal values, 55
- moral development theories, 9
- mosquito bites, 176–177
- motor development. *See also* physical growth and development
- infants (birth to 2 years), 11
 - milestones, 12*t*, 14*t*
 - school-aged children, 15
 - toddlers and preschoolers, 14
- mouth
- disorders of, 106–109
 - aphthous ulcers (canker sores), 108
 - cleft lip/palate, 107
 - dental caries, 107–108
 - example questions and answers, 114–117
 - hand, foot, and mouth disease, 109
 - herpangina, 109
 - herpes labialis (cold sore, fever blister), 108–109
 - herpes simplex stomatitis, 108–109
 - oral candidiasis (thrush), 106–107
 - normal findings, 48
- MPS IH (mucopolysaccharidosis I), 365–366
- MSP. *See* Munchausen syndrome by proxy
- mucocutaneous lymph node syndrome (Kawasaki disease), 127–129
- mucopolysaccharidosis I (MPS IH), 365–366
- Multistate Nurse Licensure Compact, 376
- multisystem disorders, 349–372
- about, 349–350
 - cerebral palsy (CP), 366–367
 - congenital diseases, 350–359
 - acquired immune deficiency syndrome (AIDS), 350–351
 - chlamydia trachomatis, 351–352
 - congenital rubella syndrome (CRS), 356
 - congenital syphilis, 353–354
 - cytomegalovirus (CMV), 355
 - fetal alcohol syndrome (FAS) and fetal alcohol spectrum disorders (FASD), 358–359
 - gonococcal infection, 352–353
 - hepatitis B, 356–357
 - herpes, 354–355
 - toxoplasmosis, 357–358
- example questions and answers, 369–371
- genetic syndromes, 359–366
- Down syndrome (DS) (trisomy 21), 360–361
 - Fragile X syndrome (FXS), 361–362
 - Klinefelter's syndrome (XXY karyotype), 363
 - Marfan syndrome, 364–365
 - mucopolysaccharidosis I (MPS IH), 365–366
 - Prader-Willi syndrome, 366
 - Tay-Sachs disease, 363–364
 - trisomy 18 (Edwards Syndrome), 359–360
 - Turner's syndrome (XO karyotype), 362–363
- spina bifida, 367–369
- sudden infant death syndrome (SIDS), 369
- mumps, 221–222
- Munchausen syndrome by proxy (MSP), 22. *See also* abuse and neglect
- murmurs, heart, 49, 122–123
- musculoskeletal disorders, 233–249
- developmental dysplasia of the hip (DDH), 234–235

- duchenne muscular dystrophy (DMD), 245–246
 example questions and answers, 246–249
 genu valgum (knock-knee), 237
 genu varum (bowleg), 236–237
 growing pains, 238
 juvenile idiopathic arthritis, 242–244
 legg-calvé-perthes disease (LCPD), 238
 metatarsus adductus/metatarsus varus, 235–236
 Osgood-Schlatter disease, 239
 osteomyelitis, 245
 scoliosis (idiopathic), 239–240
 slipped capital femoral/1nlepiphyysis, 241
 sports injuries, 240–241
 systemic lupus erythematosus, 244–245
 talipes equinovarus congenita (clubfoot), 235
 tibial torsion (internal), 236
 torticollis (wry neck), 45, 233–234
 transient (toxic) synovitis of the hip, 237
 musculoskeletal system, normal findings, 51–52
 myocardial inflammatory disease, acute, 129–130
 myoclonic seizures, 252
 myopia (nearsightedness), 92
- N**
 nasal polyps, 106
 nasopharyngitis, acute (common cold), 111
 National Association of Pediatric Nurse Practitioners, 374
 nature theories of human growth and development, 7–8
 nearsightedness (myopia), 92
 neck
 normal findings, 44–45
 torticollis (wry neck), 45, 233–234
 needs, hierarchy of (Maslow), 9
 negatively formatted test questions, 5
 negativism, 71
 negligence, 379
 Neisseria gonorrhoeae (GC). *See* gonoccal infection
 neo-Freudian theories of development, 8
 neonatal hyperbilirubinemia, 300–301
 nephroblastoma (Wilms' tumor), 313
 nephrogenic diabetes insipidus (DI), 326–328
 neuroblastoma, 311
 neurofibromatosis (NF1) (Von Recklinghausen disease), 257–258
 neurogenic diabetes insipidus (DI), 326–328
 neurologic system, normal findings, 52
 neurological disorders, 251–267
 brain tumors, 263–264
 example questions and answers, 264–267
 febrile seizures, 255
 head injuries, 260–262
 headaches, 255–257
 meningitis, 262–263
 neurofibromatosis (NF1) (Von Recklinghausen disease), 257–258
 seizure disorders/epilepsy, 251–255
 tic disorders, 258–260
 tuberous sclerosis (TS), 258
 neutrophils, normal values, 55
 nevi (birthmarks), 53
 capillary hemangioma (strawberry nevus), 155–156
 port-wine stain, 155
 salmon patch (nevus simplex), 154–155
 Newborn Behavioral Assessment Scale (NBAS), 32
 newborn examination and screening, 57–58, 59–60
 newborn exanthema, 153–154
 cutis marmorata, 153
 erythema toxicum neonatorum, 153–154
 milia, 154
 newborns. *See* infants
 NF1 (neurofibromatosis), 257–258
 NHL (non-Hodgkin's lymphoma), 312–313
 nicotine, 30
 nitrites, normal values, 56
 non-Hodgkin's lymphoma (NHL), 312–313
 non-insulin-dependent diabetes mellitus (NIDDM), 336–339
 norovirus (Norwalk virus), 195
 norwalk, 195*t*
 nose
 disorders of, 103–106
 allergic rhinitis, 103–104
 chronic rhinitis, 104
 epistaxis, 104–105
 example questions and answers, 114–117
 foreign body in nose, 104
 nasal polyps, 106
 sinusitis, 105–106
 normal findings, 47–48
 nursing. *See* advanced practice nursing
 Nursing's Agenda for Health Care Reform, 384
 nurture theories of human growth and development, 8
 nutrition. *See also* breastfeeding
 cholesterol. *See* cholesterol
 guidance
 adolescents, 75–76
 infants, 68–69
 school-aged children, 74
 toddlers and preschoolers, 71, 72
 history, 40
 malabsorption, 201–202
 obesity, 343–344
 nystagmus, 91
- O**
 obesity, 21–22, 343–344
 obstructive sleep apnea (OSA), 146
 oligoarthritis, 242
 OME. *See* otitis media with effusion
 oncologic disorders. *See* cancers
 operant conditioning theory, 8
 ophthalmia neonatorum (conjunctivitis of the newborn), 84–85
 ophthalmoscopic examination, 47
 opiates, 31
 oppositional-defiant disorder, 25
 oral candidiasis (thrush), 106–107
 orbital/periorbital cellulitis, 88–89
 organizations, professional, 382
 Ortolani test, 52
 Ortolani's sign, 234
 Osgood-Schlatter disease, 239
 osteomyelitis, 245
 osteosarcoma, 313–314
 otitis media, 96–99
 acute (AOM), 97–98
 antibiotic therapy for, 99*t*
 with effusion (OME), 99
 external (EOM), 96–97
- P**
 palate
 cleft lip/palate, 107
 normal findings, 48
 papulosquamous conditions, 159–161
 pityriasis rosea, 159–160
 psoriasis, 160–161
 paraphimosis, 274
 parent-child relationship
 attachment, 9, 13
 maternal deprivation experiments, 8
 parent-infant interaction, 9, 13
 screening and assessment tests, 32
 parenting styles and discipline, 14
 guidance, 74, 76
 partial seizures, 251–252
 past medical history (PMH), 40
 patent ductus arteriosus (PDA), 122*t*
 pediculosis infestation (lice), 180–181
 pelvis, examination of, 61–62
 perforations, ear, 100
 perinatal history, 40
 periodontal disease, 108
 periorbital cellulitis, 88–89
 periorbital edema, 46
 peritonsillar abscess, 112
 permissive parenting, 14
 personal fable, 17
 person-place-process model, 10
 pertussis (whooping cough), 215–216
 pH, urine, 56
 pharyngitis and tonsillitis, 110–111
 phenobarbital, 254*t*
 phenytoin, 254*t*
 phimosis and paraphimosis, 274
 physical abuse. *See* abuse and neglect
 physical domain, 7
 physical examination, 42–44
 physical growth and development
 abuse and neglect, 22–23
 adolescents, 16–17

- aggression, defiance, and disruptive behaviors, 25–26
- Attention Deficit Hyperactivity Disorder (ADHD), 23–24
- Autism Spectrum Disorder (ASD), 19–21
- childhood depression, 28–29
- deviations in, 18–31
- failure-to-thrive (FTT), 18–19
- eating disorders, 27–28
- head, normal, 81*t*
- health supervision, 31–32
- in history, 41
- infants (birth to 2 years), 10–11
- learning disabilities (LD), 26–27
- obesity, 21–22
- school-aged children, 15
- screening and assessment tests, 32
- substance abuse, 30–31
- suicidal behavior, 29–30
- toddlers and preschoolers, 13–14
- Piaget, Jean, 8
- pigmentation conditions, 156–159
- albinism, 157–158
- café au lait spots, 156
- malignant melanoma, 157
- Mongolian spots, 156–157
- pityriasis alba, 159
- pityriasis rosea, 159–160
- vitiligo, 158–159
- pinworms (*enterobiasis vermicularis*), 199
- pituitary disorder—diabetes insipidus (DI), 326–328
- pityriasis alba, 159
- pityriasis rosea, 159–160
- planning for certification test, 2
- maximizing study time, 5–6
- Plasmodium*. *See* malaria
- platelets, 55
- play (in human development), 15
- plumbism (lead poisoning), 305–306
- PMH. *See* past medical history
- PMI. *See* point of maximal impulse
- PNCB examination, overview of, 3*t*
- pneumonia, 138–140
- point of maximal impulse (PMI), 49
- Point of Service Plans (POSS), 385
- poliomyelitis, 226–227
- polyarthritis, 242
- polyps, nasal, 106
- port-wine stain, 155
- position statements on nurse practitioners, 374
- positive predictive value (PPV), physical growth, 31
- postnatal history, 40
- potassium, 55–56
- potential test item traps, 5
- PPV. *See* positive predictive value
- practice issues, 377–382
- practice-based research, 382
- Prader-Willi syndrome, 366
- precocious puberty, 339–341
- Preferred Provider Organizations (PPOs), 385
- pregnancy, 290–293. *See also* contraception
- example questions and answers, 293–298
- prehypertension, 125
- premature atrial contractions (PAC), 124
- premature ventricular contractions (PVC), 124
- prenatal breastfeeding guidance, 63–64
- prenatal health supervision visits, 62–63
- preschoolers. *See* toddlers and preschoolers
- prescriptive authority, 376
- pretend or dramatic play, 15
- primary health care, 384–385
- prioritizing issues, questions on, 4–5
- privacy. *See* confidentiality issues
- problem-oriented health records, 62
- professional organizations, 382
- progestins, long-acting, 289
- Prolonged QT Syndrome, 124
- protein (CSF test), 56
- protein (urine test), 56
- psoriasis, 160–161
- psychological and social domain, 7
- psychological birth, 8
- psychosexual development, stage theory of, 8
- psychosocial development
- adolescents, 17
- infants (birth to 2 years), 13
- school-aged children, 15–16
- stage theory (Erikson), 8
- toddlers and preschoolers, 14–15
- puberty, 16–17
- disorders of pubertal development, 339–341
- pubic hair development, 16–17
- pulse, norms, 43, 43*t*
- pupils, normal findings, 46–47
- pyloric stenosis, 194–195
- Q**
- quality improvement in advanced nursing practice, 378
- questions (test), 2–5. *See also* example questions and answers
- R**
- RAFFT Tool for Adolescents, 42
- rash. *See* dermatologic conditions
- RBC. *See* red blood cells
- reaction range, 10
- receptive language, 14
- recurrent abdominal pain, 203–204
- red blood cells (RBC), 56. *See also* hematologic disorders
- normal values, 54
- red measles (rubeola), 218–219
- reflexes, 52*t*
- refractive errors of eye, 91–92
- astigmatism, 92
- hyperopia (farsightedness), 91–92
- myopia (nearsightedness), 92
- regulation of advanced nursing practice, 376–377
- reimbursement, about, 379, 386
- renal tubular acidosis (RTA), 280–281
- reproductive history. *See* sexual/reproductive history
- reproductive system
- menstrual disorders (amenorrhea), 342–343
- normal findings, 51
- requirements for certification, reviewing, 2
- research in advanced nursing practice, 382
- resiliency model of development, 10
- respiration norms, 43, 43*t*
- reticulocyte count, 55
- retinoblastoma, 91, 311–312
- retropharyngeal abscess, 112
- review of symptoms (ROS), 41
- Rh incompatibility, 299–300
- rheumatic fever/heart disease, 130–131
- rhinitis, 103–104
- Rickettsia rickettsii*. *See* Rocky Mountain spotted fever
- ringworm
- of the body (*tinea corporis*), 173–174
- of the feet (*tinea pedis*), 175–176
- of the scalp (*tinea capitis*), 172–173
- risk management, 378
- RMSF (Rocky Mountain spotted fever), 223
- Rocky Mountain spotted fever (RMSF), 223
- Rogers, Carl, 9
- roles of nurse practitioners, 374
- roseola (exanthem subitum), 219
- rotavirus, 195, 195*t*
- rough-and-tumble play, 15
- Rousseau, Jacque, 7
- Rovsing's sign, 203
- RTA (renal tubular acidosis), 280–281
- rubella (german measles), 217–218
- congenital rubella syndrome (CRS), 356
- rubeola (red measles), 218–219
- Rutter, Michael, 10
- S**
- safety
- guidance
- adolescents, 76
- infants, 69
- school-aged children, 74–75
- toddlers and preschoolers, 71–72, 73
- salmon patch (nevus simplex), 154–155
- Salmonella, 196*t*, 197, 198
- Salmonella typhi, 196*t*, 197
- Sameroff, Arnold, 10
- scabies infestation, 179–180
- scalp injuries, 260
- schedule of child health supervision visits, 62–76
- adolescents, 75–76
- infants (birth to 2 years), 65–71
- prenatal, 62–64

- school-aged children, 73–75
 toddlers and preschoolers, 70–73
 school-aged children
 physical examination, 43, 43*t*, 44*t*
 schedule of visits, 73–75
 school-aged development, 15–16
 sclera, 46
 scoliosis, 52
 scoliosis (idiopathic), 239–240
 scope of advanced nursing practice, 375
 screening and assessment tests
 child health supervision, 32
 health promotion, 57–59
 physical growth and development, 31
 seborrhea dermatitis, 164
 seizure disorders, 251–255
 common antiepileptic drugs, 254*t*
 self-assessment for certification, 1
 sensitive periods, theory of, 8
 sensitivity, physical growth and development, 31
 sensorimotor development, 12–13
 sensorineural hearing loss, 101–102
 separation anxiety, 69
 sepsis (septicemia), 213–214
 septicemia (sepsis), 213–214
 sex education, 73, 74
 sexual abuse. *See also* abuse and neglect
 signs and symptoms, 51
 sexually transmitted infections (STIs), 281–288
 acquired syphilis, 283–285
 bacterial vaginosis (BV), 287–288
 chlamydia, 282–283
 genital herpes simplex virus (HSV), 285–286
 genital warts (condylomata acuminata), 286–287
 gonorrhea, 281–282
 screening, 59
 trichomoniasis, 287
 sexual/reproductive history, 42
 Shigella, 196*t*, 197, 198
 short stature, 328–330
 Shuler's model of NP practice, 374
 sibling rivalry, 71
 sickle cell disease, 303–305, 303*t*, 304*t*
 SIDS (sudden infant death syndrome), 369
 Simon, Theophile, 7
 single ventricle physiology, 121*t*, 122*t*
 sinus headache, 256
 sinuses, normal findings, 47–48
 sinusitis, 105–106
 skeletal issues. *See* musculoskeletal disorders
 skin. *See also* dermatologic system
 normal findings, 52–53
 tuberculin skin test (TST), 146
 Skinner, B. F., 8
 skull fracture, 261
 sleep
 guidance
 infants, 69
 toddlers and preschoolers, 71, 72
 history, 40
 slipped capital femoral/epiphysis, 241
 social and psychological domain, 7
 social learning theory, 8
 sodium chloride, normal values, 55
 special examinations, 59–62
 specificity, physical growth and development, 31
 speech. *See* language development
 spermicides, vaginal, 288
 spider bites, 177–178
 spina bifida, 367–369
 spine. *See* musculoskeletal disorders; spina bifida
 spirometry, 143
 spontaneous termination of pregnancy, 291
 sports evaluation, 60–61
 sports injuries, 240–241
 sprains, 240–241
 Stroufe, Alan, 10
 stage theory of life span psychosocial development, 8
 stage theory of psychosexual development, 8
 standards of advanced nursing practice, 375
 staphylococcal scalded skin syndrome, 167–168
 Staphylococcus aureus, 196*t*, 197
 Stevens-Johnson syndrome (erythema multiforme major), 183
 Still's murmur, 123
 stings, insect, 178–179
 STIs. *See* sexually transmitted infections
 strabismus, 90–91
 “strange situation” paradigm, 9
 strawberry nevus (capillary hemangioma), 155–156
 structure of American family, 17
 study plan, 2, 4*f*
 study time, maximizing, 5–6
 stuttering, 19
 stye (hordeolum), 88
 subdural hematoma (head), 260–261
 substance abuse, 30–31
 adolescent pregnancy and, 292
 history in adolescents, 42
 substance dependence, 30
 sudden infant death syndrome (SIDS), 369
 suicidal behavior, 29–30
 sunburns, 165–166
 supraglottitis (epiglottitis), 113–114
 supraventricular tachycardia (SVT), 124
 surveillance, physical growth and development, 31
 synechia vulvae, 275–276
 syphilis
 acquired syphilis, 283–285
 congenital syphilis, 353–354
 systemic lupus erythematosus, 244–245
T
tabula rasa, child as, 8
 tachycardia, 124
 talipes equinovarus congenita (clubfoot), 235
 Tanner staging, 16–17
 Tay-Sachs disease, 363–364
 TB. *See* tuberculosis
 teeth. *See* dental development
 telephone history, 43
 television, guidance on, 72
 temperament, 13
 temperature measurement, 43
 temporomandibular joint (TMJ), 48
 tension headaches, 256
 termination of pregnancy, 291–292
 test day, preparing for, 6
 test plan for certification, 2
 test questions, about, 2–5. *See also* example questions and answers
 test taking strategies, 1–6
 testicular torsion, 275
 tests. *See* laboratory tests and values; screening and assessment tests
 tetanus (lockjaw), 227–228
 Tetralogy of Fallot (TOF), 121*t*
 throat
 disorders of, 110–114
 acute nasopharyngitis (common cold), 111
 cervical lymphadenitis, 113
 epiglottitis (supraglottitis), 113–114
 example questions and answers, 114–117
 peritonsillar abscess, 112
 pharyngitis and tonsillitis, 110–111
 retropharyngeal abscess, 112
 normal findings, 48
 thrush (oral candidiasis), 106–107
 thumb sucking, 71
 thymic hypoplasia (DiGeorge syndrome), 315
 thyroglossal duct cyst, 45
 thyroid, enlarged, 45
 thyroid disorders, 321–326
 hyperthyroidism, 323–325
 hypothyroidism, 321–323
 thyroiditis, 325–326
 thyroiditis, 325–326
 tibial torsion (internal), 236
 tic disorders, 258–260
 tick bites. *See* lyme disease; Rocky Mountain spotted fever
 tinea capitis (ringworm of the scalp), 172–173
 tinea corporis (ringworm of the body), 173–174

tinea cruris (jock itch), 174–175
 tinea pedis (athlete's feet or ringworm of the feet), 175–176
 TM. *See* tympanic membrane
 TMJ. *See* temporomandibular joint
 tobacco. *See* substance abuse
 toddler growth and development, 13–15
 toddlers and preschoolers
 physical examination, 42–43, 43*t*, 44*t*
 schedule of visits, 70–73
 toilet training, 71
 tonic seizures, 252
 tonic-clonic (grand mal) seizures, 255
 tonic-clonic seizures, 252
 tonsillar abscess, 112
 tonsillitis and pharyngitis, 110–111
 tonsils, 48
 torticollis (wry neck), 45, 233–234
 total anomalous pulmonary venous connection, 121*t*
 Tourette syndrome, 259–260
 toxic transient synovitis of the hip, 237
 toxoplasmosis, 357–358
 trachea. *See* lower respiratory disorders
 transactional developmental theories and theorists, 9–10
 transient (toxic) synovitis of the hip, 237
 transposition of the great arteries, 121*t*
 Trendelenburg test, 52
 trends in advanced practice nursing, 375
Treponema pallidum, 353–354
 trichomoniasis, 287
 trisomy 18 (Edwards Syndrome), 359–360
 trisomy 21 (Down syndrome), 360–361
 truncus arteriosus, 121*t*
 TS (tuberous sclerosis), 258
 tuberculin skin test (TST), 146
 tuberculosis (TB), 147–149
 screening, 58
 tuberous sclerosis (TS), 258
 Turner's syndrome (XO karyotype), 362–363
 tympanic membrane (TM), 47
 tympanostomy tubes, 99–100
 type 2 diabetes, 336–339

U
 ulcerative colitis (UC). *See* inflammatory bowel disease
 umbilical hernia, 208
 undescended testes (cryptorchidism), 272–273
 urinary tract infection (UTI), 269–271
 urine tests, 56, 59
 urticaria (hives), 184
 UTI (urinary tract infection), 269–271
 utilization of health policy, 384

V
 vaginal spermicides, 288
 valproic acid, 254*t*
 varicella-zoster virus (VZV) (chickenpox), 220–221
 vascular lesions, 154–156
 capillary hemangioma (strawberry nevus), 155–156
 port-wine stain, 155
 salmon patch (nevus simplex), 154–155
 vasopressin-resistant diabetes insipidus (DI), 326–328
 vasopressin-sensitive diabetes insipidus (DI), 326–328
 ventricular septal defect, 122*t*
 verruca vulgaris (warts), 172
 viral dermatologic conditions, 170–172
 herpes simplex (common cold sore), 108–109, 170–171
 molluscum contagiosum, 171
 verruca vulgaris (warts), 172
 vision. *See also* eyes
 development of, 11
 normal findings, 46–47
 screening, 57
 visits for child health supervision, 62–76
 adolescents, 75–76
 infants (birth to 2 years), 65–71
 prenatal, 62–64
 school-aged children, 73–75
 toddlers and preschoolers, 70–73
 vital signs, measurement of, 43

vital signs, measurement of, 43
 vitiligo, 158–159
 voice, normal findings, 48
 volatile substances, 31
 Von Recklinghausen disease (neurofibromatosis), 257–258
 von Willebrand Disease (VWD), 308–309
 vulvovaginitis, 276–277
 VWD (von Willebrand Disease), 308–309
 Vygotsky, L. S., 9
 VZV (varicella-zoster virus), 220–221

W
 warts
 genital (condylomata acuminata), 286–287
 verruca vulgaris, 172
 WAS (Wiskott-Aldrich syndrome), 315–316
 wasp stings, 178–179
 water safety. *See also* safety guidance, 70
 Watson, John B., 8
 WBC. *See* white blood cells
 webbed neck, 45
 Werner, Emmy, 10
 West syndrome (infantile spasms), 252
 white blood cells (WBC), 54, 56
 whooping cough (pertussis), 215–216
 Wilms' tumor (nephroblastoma), 313
 Wiskott-Aldrich syndrome (WAS), 315–316

X
 X-linked agammaglobulinemia, 314–315
 XO karyotype (Turner's syndrome), 362–363
 XXY karyotype (Klinefelter's syndrome), 363

Y
 yellow jacket stings, 178–179
 yellow sclera, 46

Z
 zone of proximal development (ZPD), 9