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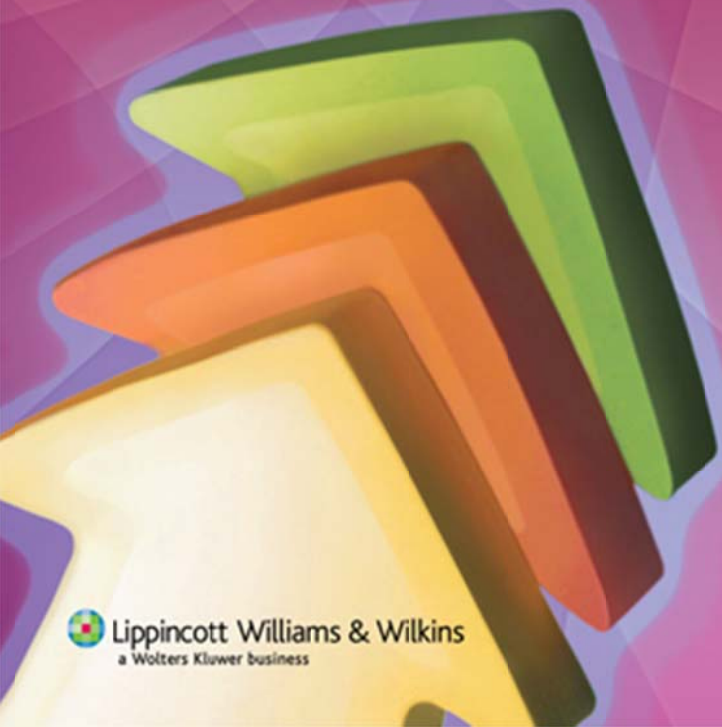
PEDIATRICS RECALL

FOURTH EDITION

Eugene D. McGahren
William G. Wilson



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Fourth Edition

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PEDIATRICS RECALL
Fourth Edition

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Gene McGahren dedicates the fourth edition to
Catherine, Christopher, Caroline, Matthew, and Annie
for their love, understanding, and humor always.

Bill Wilson dedicates the fourth edition to all of the students
and residents who make a career in academic medicine
challenging and rewarding.

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Preface to the Fourth Edition

Pediatrics Recall provides information in a concise question-and-answer format, which allows easy access to material that is essential for medical students during their third-year clinical clerkships in pediatrics. The book covers basic issues in neonatal and pediatric fluid management, blood products, nutrition, emergencies, growth, and intensive care. In addition, an entire chapter is devoted to issues relating to the adolescent patient. Disease entities are organized into chapters according to the systems involved. Descriptions of the individual diseases include signs, symptoms, essentials of pathophysiology, treatments, and possible outcomes. Students may use the question-and-answer format to work through each condition from presentation and diagnosis to therapy and outcome.

Pediatrics Recall is not intended to be used as a primary text. Rather, it allows students to review essential information in an efficient format that is designed to facilitate retention.

Changes in the fourth edition of *Pediatrics Recall* include added disease entities as well as new terminology, techniques, and insights that have evolved in the field of pediatrics since the third edition was published. It also reflects input from students, residents, community physicians, and pediatric generalists and specialists in both the primary writing and review of the text. We hope that this book is useful to you and we welcome your suggestions.

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Contents

Contributors vi
Preface viii
Acknowledgments ix

SECTION I
OVERVIEW AND BACKGROUND INFORMATION

1. Introduction 01
Using the Study Guide 01
Pediatric Notes and Presentations 01
Common Abbreviations 03

2. Pediatric Procedures 09
Venipuncture/IVs 09
Lumbar Puncture 11
Suprapubic Puncture 13
Arterial Puncture 13

3. Fluids and Electrolytes 15
Maintenance Fluid Requirements 15
Dehydration 16

4. Blood and Blood Products 20

5. Pediatric Nutrition 23
Intravenous and Parenteral Alimentation 27

6. Pediatric Emergencies 30
Resuscitation in Children 30
Respiratory Distress 31
Acute Cardiovascular Collapse 33
Acute Neurologic Conditions and Mental Status Changes 41
Environmental Emergencies 47

7. Growth and Development 55
Growth 55
Development 58
Puberty 61

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SECTION II
NEWBORN CARE

8. Perinatal Care and Evaluation of the Newborn 63
Apgar Scores 63
Newborn Resuscitation 64
Evaluation of the Newborn 67

9. Common Clinical Problems 79
Jaundice 79
Neonatal Seizures 81
Neonatal Anuria and Oliguria 83
Failure to Pass Meconium 86
Neonatal Cyanosis 87
Neonatal Respiratory Distress 89
Neonatal Hypotonia 90

10. Diseases of the Newborn 92
Periventricular or Intraventricular Hemorrhage 92
Respiratory Distress Syndrome 94
Transient Tachypnea of the Newborn 96
Meconium Aspiration Syndrome 96
Persistent Pulmonary Hypertension of the Newborn 98
Neonatal Pneumonia 100
Neonatal Sepsis 101
Neonatal Bacterial Meningitis 102
Apnea of Infancy and Sids 104
Neonatal Diarrhea 105
Umbilical Abnormalities 106
Umbilical Hernia 107

11. Newborn Intensive Care: General Considerations . . 109
Respirators 109
Monitors 109
Nutrition 110
Environment 111
Common Questions Parents Ask 112

SECTION III
AMBULATORY PEDIATRICS

12. The Pediatric Physical Examination 113
The Well-Child Visit 113
The School Physical 119
Common Clinical Problems 123
Fever 123
Respiratory Distress 123

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13. Pediatric Sports Medicine 133
General Issues in Sports Medicine 133
Preparticipation Sports Assessment 134
Musculoskeletal Injuries 138
Management of Injuries 138
Head Injuries 142
Eye Injuries 145
Medical Issues Related to Sports Participation 145

14. Adolescent Medicine 155
Puberty and Growth 155
Legal Issues 157
Basic Issues of Teen Hygiene 157
Acne 158
Menstruation 158
Sexually Transmitted Diseases 159
Pregnancy and Contraception 160
Epidemiology of Accidental and Nonaccidental Death 161

SECTION IV
PEDIATRIC DISEASES

15. Hematologic Disorders 163
Nutritional Anemias 163
Hemoglobinopathies 167
Hemolytic Anemias 171
Aplastic Anemia 173
Gaucher Disease 174
Polycythemia 175
Granulocyte Disorders 177
Platelet Disorders 179
Coagulation Defects 182
Heparin-Induced Thrombocytopenia 186
Neonatal Alloimmune Thrombocytopenia 186

16. Pediatric Cardiology 188
Introduction to Pediatric Cardiology 188
Congenital Heart Defects 190
Acquired Heart Disease 205

17. Respiratory and Thoracic Disorders 215
Congenital Diaphragmatic Hernia (CDH) 215
Emphysema 219
Lung Cysts 220
Pneumonia or Pneumonitis 221
Asthma 223
Airway Foreign Body 227
Cystic Fibrosis (CF) 228

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Tracheomalacia 229
Pneumothorax 230
Chylothorax 231
Pectus Deformity 232
Esophageal Duplication Cyst 235
Bronchogenic Cyst 235
Pulmonary Sequestration 236
Congenital Cystic Adenomatoid Malformation (CCAM) 237

18. Head and Neck 239
Epiglottitis 239
Croup 242
Pierre Robin Sequence 244
Choanal Atresia 245
Vocal Cord Paralysis 246
Laryngeal Web 246
Laryngeal Cleft 247
Subglottic Stenosis 247
Branchial Cleft Remnants 248
Thyroglossal Duct Remnant 248
Torticollis 249
Peritonsillar Abscess 250
Craniosynostosis 251

19. Gastrointestinal Disorders 252
Short-Gut (Short-Bowel) Syndrome 252
Necrotizing Enterocolitis 254
Ulcerative Colitis 257
Crohn Disease 260
Antibiotic-Related Colitis (Pseudomembranous Colitis) 263
Celiac Disease (Gluten Enteropathy/Celiac Sprue) 263
Gastroesophageal Reflux in Infants and Children 266
Peptic Ulcer Disease 270
Intussusception 272
Anorectal Malformations (Imperforate Anus) 275
Henoch-Schönlein Purpura 277
Intestinal Transport Defects 281
Appendicitis 282
Pancreatitis 283
Hemolytic Uremic Syndrome 285
Constipation 287
Encopresis 290
Functional Abdominal Pain in Children 291
Hirschsprung Disease 293
Malrotation 294
Congenital Duodenal Obstruction 296
Intestinal Atresia 298
Esophageal Atresia or Tracheoesophageal Fistula 300
Pyloric Stenosis 304

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Intestinal Polyps 305
Bezoar 306
Pica 307
Meckel Diverticulum 308
Perianal and Perirectal Abscess 309

20. Liver and Hepatobiliary Disorders 311
Cholelithiasis (Gallstones) 311
Hepatitis 313
Biliary Atresia 320
Choledochal Cysts 321
Portal Hypertension 323
Congenital Hepatic Fibrosis 325
Alagille Syndrome 325

21. Renal Diseases 328
Acute Kidney Injury 328
Chronic Kidney Disease 330
Nephrotic Syndrome 332
Glomerulonephritis 334
Vesicoureteral Reflux 335
Renal Tubular Acidosis 336
Fanconi Syndrome 338
Diabetes Insipidus 338
Renal Stones 341
Hypertension 342

22. Genitourinary Disorders 344
Hypospadias 344
Cryptorchidism 344
Epididymitis 346
Testicular Torsion 346
Testicular Tumors 347
Ovarian Tumors 349
Vaginal Anomalies 353
Vaginal Tumors 354
Imperforate Hymen 355

23. Hernias and Abdominal Wall Defects 356
Hernias 356
Abdominal Wall Defects 360

24. Endocrine Disorders 364
Diabetes 364
Disorders of Calcium Balance 370
Precocious Puberty 375
Gynecomastia 377
Delayed Puberty 378

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GH Deficiency 380
Panhypopituitarism 382
Thyroid Disorders 383
SIADH 388
Adrenocortical Insufficiency 389
Cushing Disease and Syndrome 390
Pheochromocytoma 391
Ambiguous Genitalia 392

25. Neurologic Diseases 396
Seizure 396
Arnold-Chiari Malformation 397
Cerebral Palsy 398
Intracranial Hemorrhage 399
Guillain-Barré Syndrome 400
Myasthenia Gravis 401
Neural Tube Defects 402
Macrocephaly and Hydrocephalus 403
Common Muscular Dystrophies 404
Miscellaneous Neurologic Conditions 404

26. Neoplastic Diseases 406
Neuroblastoma 406
Wilms Tumor 410
Hodgkin Disease 413
Non-Hodgkin Lymphoma 416
Leukemia 418
Retinoblastoma 421
Rhabdomyosarcoma 422
Osteogenic Sarcoma 424
Ewing Sarcoma 426
Medulloblastoma 427
Astrocytoma 427
Hepatoblastoma 428
Hepatoma 430
Teratoma 431
Melanoma 433

27. Skin, Soft Tissue, Nail, and Hair Disorders 435
Definitions 435
Inflammatory Disorders 436
Bacterial Infections 439
Viral Infections 443
Fungal Infections 445
Parasitic Infections 446
Hyperpigmentation and Hypopigmentation 447
Hypersensitivity Reactions 450
Benign Mass Lesions 452

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Syndromes with Prominent Cutaneous Features 454
Cutaneous Manifestations of Systemic Disease 456
Nail Disorders 458
Hair Disorders 460

28. Infectious Diseases 462
Meningitis 462
Conjunctivitis 467
Otitis Media 469
Thrush 473
Gingivostomatitis 475
Lymphadenitis 476
Gastroenteritis 477
Urinary Tract Infection 479
Pyelonephritis 481
Dactylitis 482
Sexually Transmitted Diseases (STD) 482
Common Viral Syndromes 489
Human Immunodeficiency Virus (HIV) 494
Tuberculosis 497
Pertussis 501
Parasitic Infections and Infestations 503
Atypical Mycobacteria 506

29. Allergic Diseases 507
Atopy 507
Food Allergies 508
Allergic Rhinitis 509
Insect Stings and Bites 510

30. Genetics 512
Definitions 512
Common Genetic Syndromes 513

Index 519

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Section I

Overview and Background Information

Chapter 1

Introduction

USING THE STUDY GUIDE

This study guide was written to accompany the pediatric clerkship, and we welcome any feedback or suggestions for improvement. The objective of the guide is to provide a rapid overview of common pediatric topics, but keep in mind that this is NOT an all-encompassing source (i.e., you will have to consult major textbooks to round out the information in this guide). The guide is organized in a self-study quiz format. By covering the information and answers on the right side of the page with the bookmark, you can attempt to answer the questions on the left to assess your understanding of the information. Keep the guide with you at all times, and when you have even a few minutes (e.g., between patients), hammer out a page or at least a few questions.

PEDIATRIC NOTES AND PRESENTATIONS

DOCUMENTATION AND COMMUNICATION

In addition to documentation and communication functions, the content of admission and progress notes implies much about the ability of the medical student or resident to gather and analyze data and formulate a treatment plan. Therefore, pertinent negative findings (i.e., the normal findings that the student or resident chooses to include in the written notes) convey the student's problem-solving abilities. In addition, many pediatric, medical, or surgical services expect the medical students to write a detailed discussion of the patient's condition at the end of an admission or progress note. Make certain you understand what is expected of you.

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ADMISSION NOTES

The format of an admission note may vary in different institutions, but the note generally contains the following information:

1. Name of the patient
2. Age
3. Sex
4. Reason for admission (presenting complaint or diagnosis)
5. Informant (historian)
6. Referring physician or health professional (if referred)
7. History of present illness
8. Past medical history (including, as indicated, prenatal and newborn history)
9. Immunizations
10. Allergies
11. Current medications
12. Hospitalizations
13. Surgeries
14. Developmental history
15. Review of systems
16. Physical measurements (height, weight, head circumference)
17. Vital signs (usually temperature, pulse, respiratory rate, blood pressure)
18. Physical examination
19. Assessment
20. Plan

The organization of the admission note may vary. Because pediatrics encompasses a broad age range, certain elements may be included or excluded, depending on the situation. For example, the elements of the history for a 3-year-old with seizures and developmental delay may not be the same as those for a 16-year-old with an ankle fracture.

PROGRESS NOTES

Progress notes should be concise and convey information about the patient's status, test results, and current treatments and plans. Editorial comments, criticisms of other services or other health care professionals, and humor should be avoided. Most hospitals use some modification of the **SOAP note** as the standard for progress notes: **s**ubjective (what the patient says he or she feels), **o**bjective (what the physical examination reveals), **a**ssessment (interpretation of the information obtained), **p**lan (treatment plan).

ORAL PRESENTATIONS

The format of an oral presentation depends on the situation. Presentations are usually brief on work and checkout rounds and are more detailed during attending or teaching rounds. You must clearly communicate to your

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colleagues the important information needed for patient care. Presentations during teaching rounds usually allow more time for discussing differential diagnoses and basic science correlations.

Presentations, such as written notes, convey information about the student's ability. A clear, well-organized presentation with discussion of the differential diagnosis and plans for evaluation and treatment implies much about the clinical and analytical skills of the student.

COMMON ABBREVIATIONS

Although abbreviations are a part of the medical culture, they may be misinterpreted. A written note that does not use abbreviations is much less likely to be misinterpreted than the one that is filled with abbreviations. Be careful in using abbreviations, because many of them have several different interpretations, depending on the context and the patient. For example, the abbreviation CP could mean cerebral palsy, cleft palate, chest pain, or carotid pulse. When in doubt, write out a term. Most hospitals have a list of approved abbreviations. Make certain that you use abbreviations that are approved in your hospital. Remember that the purposes of written notes are documentation and communication.

Δ	Change
ā	Before
ABG	Arterial blood gas
As and Bs	Apnea and bradycardia
AD	Autosomal dominant
ADH	Antidiuretic hormone
ADHD	Attention deficit hyperactivity disorder
AGA	Appropriate for gestational age
AIDS	Acquired immunodeficiency syndrome
ALL	Acute lymphocytic leukemia Acute lymphoblastic leukemia
ALTE	Acute life-threatening event
AMA	Against medical advice
AML	Acute myelocytic leukemia
ANC	Absolute neutrophil count
ANLL	Acute nonlymphocytic leukemia
AP	Anterior-posterior
AR	Autosomal recessive
AS	Aortic stenosis
ASD	Atrial septal defect
B	Bilateral
BA	Bone age

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BAER	Brainstem auditory evoked response
BE	Barium enema
bid	Twice a day
BM	Bone marrow (aspirate)
	Bowel movement
BP	Blood pressure
BPD	Bronchopulmonary dysplasia
BUN	Blood urea nitrogen
$\overline{c}$	With
CA	Cancer
	Chronologic age
C & S	Culture and sensitivity
CBC	Complete blood (cell) count
cc	Cubic centimeter
CC	Clinical clerk
CDC	Centers for Disease Control and Prevention
CDH	Congenital diaphragmatic hernia
	Congenital dislocation of the hip
CF	Cystic fibrosis
CHD	Congenital heart disease
CGH	Comparative genomic hybridization
CHF	Congestive heart failure
CMV	Cytomegalovirus
CN	Cranial nerve
CNS	Central nervous system
C/O	Complaint of
CP	Cerebral palsy
	Cleft palate
CPAP	Continuous positive airway pressure
CPR	Cardiopulmonary resuscitation
CSF	Cerebrospinal fluid
C-spine	Cervical spine
CT	Computed tomography
CVA	Cerebrovascular accident (stroke)
CVP	Central venous pressure
C/W	Compatible with
	Consistent with
CX	Culture
CXR	Chest radiograph
D/C (or DC)	Discontinue
	Discharge
DDST	Denver Developmental Screening Test
DI	Diabetes insipidus
DIC	Disseminated intravascular coagulation

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DM	Diabetes mellitus
DNA	Deoxyribonucleic acid
DNS	Dextrose in normal saline
DTaP	Diphtheria/tetanus/acellular pertussis immunization (also known as DTP: diphtheria, tetanus, and pertussis vaccine)
DTRs	deep tendon reflexes
D#W	Dextrose in water (#% glucose in g/dL [e.g., D5W])
DX	Diagnosis
ECG/EKG	Electrocardiogram
ECMO	Extracorporeal membrane oxygenation
EEG	Electroencephalogram
EGA	Estimated gestational age
EMG	Electromyogram
ESR	Erythrocyte sedimentation rate
ESRD	End-stage renal disease
ETT	Endotracheal tube
FiO_2	Fraction of inspired oxygen
FROM	Full range of motion
GBS	Group B streptococcus
G tube	Gastrostomy tube
GU	Genitourinary
HAL	Hyperalimentation
Hct	Hematocrit
HFJV	High-frequency jet ventilation
HFOV	High-frequency oscillatory ventilation
Hgb	Hemoglobin
HiB	*Haemophilus influenzae* B vaccine
HIE	Hypoxic ischemic encephalopathy
HIV	Human immunodeficiency virus
HPV	Human papillomavirus
HMD	Hyaline membrane disease
HSP	Henoch-Schönlein purpura
HSV	Herpes simplex virus
ICH	Intracranial hemorrhage
ICP	Intracranial pressure
IDDM	Insulin-dependent diabetes mellitus
IM	Intramuscular(ly)
I/O	Intake and output
IPV	Inactivated injectable poliovirus vaccine
IRDS	Infantile respiratory distress syndrome
ITP	Idiopathic thrombocytopenic purpura
IUGR	Intrauterine growth retardation

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IV	Intravenous(ly)
IVH	Intraventricular hemorrhage
IVIG	Intravenous immunoglobulin
IVP	Intravenous pyelogram
LA	Left arm
	Left atrium
LE	Lower extremity
LFTs	Liver function tests
LGA	Large for gestational age
LLL	Left lower lobe of lung
LLQ	Left lower quadrant of abdomen
LP	Lumbar puncture
LR	Lactated Ringer's (solution)
LUL	Left upper lobe of lung
LUQ	Left upper quadrant of abdomen
LV	Left ventricle
LVH	Left ventricular hypertrophy
MAP	Mean airway pressure
MD	Medical doctor
	Muscular dystrophy
MMR	Measles, mumps, rubella vaccine
MRSA	Methicillin-resistant *Staphylococcus aureus*
MS	Mitral stenosis
	Morphine sulfate
	Multiple sclerosis
MSPN	Medical student progress note
MVA	Motor vehicle accident
MVC	Motor vehicle crash
NAD	No acute distress
	No apparent distress
NARD	No apparent respiratory distress
ND	Nasoduodenal
NEC	Necrotizing enterocolitis
NG	Nasogastric
NGT	Nasogastric tube
NJ	Nasojejunal tube
NPO	Nothing by mouth
NPR	Nothing per rectum
NSR	Normal sinus rhythm
OFC	Occipitofrontal circumference
OG	Orogastric
OGT	Orogastric tube
OM	Otitis media
OPV	Oral poliovirus vaccine

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OT	Occupational therapy
PA	Posterior-anterior
	Pulmonary artery
PAC	Premature atrial contraction
PDA	Patent ductus arteriosus
	Personal digital assistant
PE	Physical examination
	Pulmonary embolism
PEEP	Positive end-expiratory pressure
PEG	Percutaneous endoscopic gastrostomy
PID	Pelvic inflammatory disease
PIP	Positive inspiratory pressure
PO	By mouth
PPHN	Persistent pulmonary hypertension of the newborn
PR	By rectum
PRN, prn	As needed
PT	Physical therapy
	Patient
qd	Daily
qid	Four times a day
qod	Every other day
RA	Radial artery
	Right atrium
RAM	Rapid alternating movement
RBC	Red blood cell
RDS	Respiratory distress syndrome
RLL	Right lower lobe of lung
RLQ	Right lower quadrant of abdomen
RML	Right middle lobe of lung
ROM	Range of motion
	Rupture of membranes
RSV	Respiratory syncytial virus
RUL	Right upper lobe of lung
RUQ	Right upper quadrant of abdomen
RV	Right ventricle
RVH	Right ventricular hypertrophy
Rx	Treatment
$\overline{s}$	Without
SCT	Sacrococcygeal teratoma
SEM	Systolic ejection murmur
SGA	Small for gestational age
SIADH	Syndrome of inappropriate antidiuretic hormone secretion
SIDS	Sudden infant death syndrome

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S/P	Status post
STD	Sexually transmitted disease
SVC	Superior vena cava
SVT	Supraventricular tachycardia
SX	Sign(s) or symptom(s)
TB	Tuberculosis
TBW	Total body water Total body weight
TEF	Tracheoesophageal fistula
tid	Three times a day
Tmax	Maximum temperature
TOF	Tetralogy of Fallot
TOGV	Transposition of the great vessels (also known as TOGA: transposition of the great arteries)
TPN	Total parenteral nutrition
TRP	Tubular reabsorption of phosphate
TTN	Transient tachypnea of newborn
UAC	Umbilical artery catheter
UPJ	Ureteropelvic junction
URI	Upper respiratory infection
U/S, US	Ultrasound examination
UTD	Up to date
UTI	Urinary tract infection
UVC	Umbilical vein catheter
VCUG	Vesicocystourethrogram Voiding cystourethrogram
VDRL	Venereal Disease Research Laboratory (test)
VER	Visual evoked response
VRE	Vancomycin-resistant enterococcus
VSD	Ventricular septal defect
VUR	Vesicoureteral reflux
WBC	White blood cell (count)
WNL	Within normal limits
$\bar{x}$	Except

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Chapter 2 Pediatric Procedures

VENIPUNCTURE/IVs

What are the 3 indications for peripheral IVs in infants and children?

1. Administration of resuscitative fluids
2. Administration of maintenance fluids
3. Access for medications and parenteralnutrition

What are the appropriate-size IVs

For infants? 24- or 22-gauge

For toddlers? 22-gauge

For school-age children? 22- or 20-gauge

For older children and adolescents? 20-, 18-, or 16-gauge

What are the preferred sites for IVs?

A peripheral location is preferred, usually the dorsum of the hand, the foot, and, in infants, the scalp. The saphenous or antecubital veins are the next options. In children old enough to walk, hand and arm sites are best. IV sites should be carefully secured (especially for infants). However, excessive wrapping with gauze should be avoided so that the IV can be easily inspected and potentially serious problems (e.g., extensive infiltration) can be avoided.

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What is the best technique for placing an IV?

Most health care workers have a technique that works best for them. The extremity is immobilized while placing the IV. (An assistant will be needed for placing IVs in neonates and toddlers.) The skin over the vein is pulled taut, allowing the IV needle to puncture through the skin without creating redundancy. Once a flash of blood is obtained, the needle is advanced about 1 mm to ensure that the plastic catheter is in the vein. The catheter is then advanced over the needle. Have securing materials, including tape and an extremity board, ready when the IV is placed.

List 4 complications of IVs.

1. Infiltration
2. Thrombophlebitis
3. Necrosis of the surrounding soft tissue; usually associated with infiltration of high-concentration calcium solutions or pressors administered through peripheral IVs. Administration of such agents through peripheral IVs should be avoided.
4. In extreme cases of infiltration, compartment syndrome with threatening of the limb may occur.

How are these 4 complications treated?

1. Infiltration: Remove the IV, elevate the extremity, and apply a warm soak.
2. Thrombophlebitis: Same as for infiltration. Occasionally, an associated cellulitis may need to be treated with antibiotics.
3. Necrosis of the soft tissue: Remove the IV. Treat like a burn by applying antibiotic ointments (e.g., silver sulfadiazine [Silvadene], bacitracin). Any eschar should be removed. In rare cases, skin grafting may be necessary.
4. If compartment syndrome is present, escharotomy or fasciotomy will be required.

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How can the pain of IV placement be minimized?

A topical anesthetic cream (e.g., prilocaine cream [EMLA]) can be applied to the IV site before placement. In older, more cooperative children, subcutaneous 1% lidocaine may be an alternative.

LUMBAR PUNCTURE

List 3 indications for LP.

1. Suspicion of meningitis (Ch 28, p. 462)
2. Administration of intrathecal medications
3. Any other need to evaluate CSF

List 4 contraindications to LP.

1. Evidence of increased ICP when positioning the patient for an LP (consider increased ICP in any patient with a brain mass or sign of brain swelling)
2. If positioning the patient for an LP would risk cardiopulmonary compromise
3. Infection of the skin overlying the site of an LP
4. Thrombocytopenia or coagulopathy

Should a contraindication to an LP delay antibiotic treatment or other therapy that may be needed?

No

How is an LP performed?

The patient is in a flexed lateral decubitus position. The skin is sterilized and the L3–4 or L4–5 interspace is identified for the LP. Local anesthesia (e.g., 1% lidocaine) is given subcutaneously at the site of the LP. Usually, a 22-gauge 1½-in. spinal needle with a stylet is used. The needle is advanced slowly until it has entered the CSF space, which usually feels like a small "pop." This sensation may not be felt in a neonate. The stylet is then removed. An opening CSF pressure may be obtained. CSF is then allowed to drip into specimen containers. The needle is withdrawn, and pressure is placed on the site.

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List 4 studies that should be performed on a CSF specimen.

Culture and sensitivity, cell count, and glucose and protein concentrations

What is a normal CSF RBC count?

Normal CSF contains no RBCs. Presence of RBCs suggests a traumatic LP or a subarachnoid hemorrhage.

How may these be differentiated?

RBC count should decline from the first collection sample to the last with a traumatic LP.

What is a normal CSF WBC count?

0–5 WBC/mm^3. The count may be as high as 15 WBC/mm^3 in newborns. Presence of polymorphonuclear neutrophil leukocytes (PMNs) within this count is **abnormal,** although 1–2 PMN/mm^3 may be present in a newborn.

What is a normal CSF protein concentration?

Up to 150 mg/dL in the neonate but falls to a normal range of 10–25 mg/dL by 6–12 weeks of age. It rises to adult range of 20–45 mg/dL during puberty. CSF protein increases approximately 1 mg/dL per 1,000 RBC/mm^3 in a bloody specimen.

What is a "traumatic" LP (a.k.a. "bloody tap")?

An LP contaminated by blood from a blood vessel that has been punctured in the process of performing the LP. Usually, this confounds the neutrophil and protein count. It is best to rely on the bacterial culture of this kind of specimen (instead of adjusting the neutrophil or protein counts to account for the traumatic LP) or to obtain another LP sample at a later time.

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SUPRAPUBIC PUNCTURE

What is an indication for suprapubic puncture?

Requirement for a sterile urine collection; it is usually performed in infants and toddlers because it is difficult to obtain a midstream urine specimen. An attempt to perform an in-and-out catheterization should be made before using a suprapubic technique.

What is the technique for suprapubic puncture?

After adequate hydration, a full bladder can be percussed. The suprapubic skin is disinfected. A 22- or 25-gauge needle is placed through the skin and into the bladder at a site about one fingerbreadth above the pubis. One or two milliliters of fluid is obtained for culture.

ARTERIAL PUNCTURE

What are the 2 indications for arterial puncture?

1. When measurement of an arterial blood gas (ABG) is required
2. When a blood sample is needed and a vein cannot be accessed, an artery provides a good site, particularly in an infant.

List 2 arteries that are preferred for drawing blood in infants.

The **radial** or the **dorsalis pedis** arteries. The femoral, brachial, or axillary arteries are less desirable because thrombosis and arterial insufficiency to the respective limb may occur.

List 3 instances in which an arterial cannula should be placed.

1. For constant monitoring of BP
2. When frequent blood samples are required, especially in an infant or premature baby
3. When frequent ABG measurements are required

What are the best sites for arterial catheters? (List 3)

Radial and dorsalis pedis arteries. In the newborn, an umbilical artery catheter (UAC) can be placed for up to 10–14 days (Ch 8, p. 66).

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What is the technique for blood sampling via an artery?

Usually, a thin needle (25- or 23-gauge) is used. It is placed into the artery at a 45-degree angle facing toward the proximal end of the artery. The practitioner should make one clean pass and then pull the needle back until the tip is within the lumen of the artery. (Avoid "searching" for the artery with the needle.) The sample can then be taken. Pressure should be held after the needle is removed to avoid a hematoma or further blood loss via leak.

What are the best techniques for placement of an arterial catheter?

Similar principles hold for placement of a catheter. Sometimes, an angle of about 30 degrees is better. After blood is obtained, the catheter should pass easily over the needle. If it does not, a small guidewire may be helpful. The needle is removed, the guidewire is passed through the plastic catheter, and the catheter is advanced over the guidewire. Arterial catheters are secured with a tape in neonates and usually with a suture in toddlers and older children.

What are the 2 complications of arterial catheters?

Thrombosis with ischemia to the affected extremity (rare when the radial or the dorsalis pedis arteries are used, because there is usually collateral flow to the palmar and plantar arches in the hand and the foot, respectively)

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Chapter 3 Fluids and Electrolytes

MAINTENANCE FLUID REQUIREMENTS

What are the 3 primary methods for calculating maintenance fluid requirements?

Basing calculation on **body weight** (most common); **body surface area (BSA);** and **caloric requirements** and **expenditures**

How is BSA calculated?

$$\text{BSA } (m^2) = \sqrt{\frac{\text{Height (cm)} \times \text{weight (kg)}}{3{,}600}}$$

What are the approximate maintenance fluid requirements for infants, toddlers, and young children?

Fluids

100 mL/kg per 24 hours for first 10 kg of body weight (4 mL/kg per hour)

50 mL/kg per 24 hours for second 10 kg of body weight (2 mL/kg per hour)

20–25 mL/kg per 24 hours for each subsequent 10 kg of body weight (~1 mL/kg per hour)

This translates to the "4:2:1" rule for hourly rates.

Electrolytes

Sodium: 3 mEq/100 mL

Chloride: 2 mEq/100 mL

Potassium: 2 mEq/100 mL

What are the approximate maintenance fluid requirements for older children (10–14 years of age)?

Water: 1,500 mL/m^2 per 24 hours

Sodium: 30–50 mEq/m^2 per 24 hours

Potassium: 20–40 mEq/m^2 per 24 hours

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DEHYDRATION

What is it?	Depletion of total body water
List 4 common pediatric causes of dehydration.	Gastrointestinal (GI) losses (e.g., from gastroenteritis, diarrhea), inadequate fluid intake, excess renal losses, increased insensible losses (e.g., fever, sweating)
List 6 findings in mild dehydration.	Loss of 3–5% of body weight, normal hemodynamic variables and skin turgor, dry mucous membranes, slight decrease in urine output, and decreased tearing
List 6 findings in moderate dehydration.	Loss of 8–10% of body weight, decreased skin turgor, dry mucous membranes, relatively normal hemodynamic variables, decreased urine output, and slight to moderate increase in heart rate
What are the findings in severe dehydration?	Loss of 10–15% of body weight, abnormal skin turgor and color, dry mucous membranes, rapid heart rate, decreased BP (although it may still be normal!), poor peripheral perfusion, no urine output or tears
What is important to remember regarding the relationship between BP and dehydration in the infant and the child?	Infants and children may maintain a relatively normal BP until a severe degree of dehydration (15–25%) has occurred! The physician must monitor other variables (i.e., heart rate, urine output, skin turgor, mucous membranes, mental status).

ISOTONIC DEHYDRATION

What is it?	Dehydration with maintenance of normal Na^+ concentration; as dehydration worsens, K^+ and BUN tend to increase, and bicarbonate (HCO_3^-) tends to decrease.

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What will happen to urine specific gravity?

It will increase. However, infants have poor ability to concentrate urine; thus, specific gravity may reach only 1.020, even in cases of severe dehydration.

What is the most common cause of isotonic dehydration?

GI losses secondary to viral or bacterial enteritis

What are the electrolyte concentrations of GI fluids?

See Table 3–1.

Table 3–1. Electrolyte Concentrations of GI Fluids

Fluid	Na^+ (mEq/L)	K^+ (mEq/L)	Cl^- (mEq/L)
Gastric	20–80	5–20	100–150
Pancreatic	120–150	5–15	75–120
Small intestine	100–150	5–15	90–130
Bile	120–170	5–15	80–120
Diarrhea	10–130	10–100	10–100

What is the treatment strategy?

Calculate fluid and electrolyte losses using body weight, electrolyte values, and estimated time of dehydration. Then, rehydrate with appropriate fluids for 24–48 hours. **Note:** Replace K^+ more slowly because K^+ needs time to move intracellularly, where it is the predominant electrolyte. K^+ is added after normal renal function is confirmed.

HYPERNATREMIC DEHYDRATION

What is it?

Loss of more body water than solute, or administration of excess sodium, resulting in elevated serum sodium (>145 mEq/L)

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What are the 4 causes of hypernatremic dehydration?

1. **Increased Na^+** (excess Na^+ intake or administration, hyperaldosteronism)
2. **Water loss** (excess respiration and perspiration, diabetes insipidus [DI; **Ch 21, p. 338**])
3. **Water loss that is greater than Na^+ loss** (GI and renal losses)
4. **Abnormal central control of osmotic balance** (essential hypernatremia)

List 5 signs and symptoms.

Lethargy, irritability, muscle weakness, convulsions, coma

Why is hypernatremic dehydration dangerous?

Because losses are more from intracellular than intravascular spaces, the symptoms may be masked until dehydration becomes severe.

How is hypernatremic dehydration treated?

Rehydrate **slowly** with low-sodium fluid to avoid rapid fluid shifts to the intracellular spaces. Usually, the deficit should be replaced over the course of 48 hours.

What may happen if correction is too rapid?

Cerebral edema

HYPONATREMIC DEHYDRATION

What is it?

Relative depletion of sodium compared with total body water loss

What are the 4 common etiologic factors?

GI losses, renal losses (including those caused by diuretics), adrenal insufficiency, third-space losses (e.g., ascites, postsurgical, burns)

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List 11 signs and symptoms.

Anorexia, nausea, muscle cramps, lethargy, disorientation, agitation, diminished or pathologic reflexes, Cheyne-Stokes respiration, hypothermia, pseudobulbar palsy, seizures

What is the treatment?

Isotonic saline, administered at a rate determined by the assessment of fluid and electrolyte losses and adequacy of rehydration. In some cases, judicious administration of hypertonic saline may be beneficial.

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Chapter 4 Blood and Blood Products

What are the uses of "whole blood"?

Sometimes used for volume expansion in emergencies in which there has been acute blood loss. There are few therapeutic indications for using "whole blood," given the availability of component therapy.

What constitutes a unit of packed RBCs?

300 mL (±50 mL) with a hematocrit of 65–80%

List 2 uses of packed RBCs.

Correct anemia and improve oxygen-carrying capacity of blood

By what amount does a packed–red-cell transfusion of 3 mL/kg raise the hematocrit and hemoglobin?

Approximately 3% (hematocrit) and 1 g/dL (hemoglobin)

What are the indications for irradiated blood products?

Situations in which the recipient might be at risk for engraftment by the donor cells (e.g., premature infants and immunocompromised patients)

List 4 commonly used products that need to be irradiated for these patients.

RBCs, platelets, whole blood, leukocytes

What are the indications for the use of CMV-negative blood for transfusion?

CMV-negative blood is used for premature infants, transplant patients who are CMV negative, bone marrow transplant recipients, patients with AIDS, and any others who might be at risk for symptomatic infection because they are immunocompromised.

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List 2 indications for granulocyte transfusion.

Neutropenia (or defective neutrophil function) with an infection; serious infection that is not responding to antibiotics

What constitutes a unit of platelets?

About 5×10^{10} platelets in 40–70 mL of plasma if stored at 20°C–24°C or 20–30 mL of plasma if stored at 1°C–6°C

List 2 indications for platelet transfusions.

Thrombocytopenia and **severe platelet dysfunction** (Ch 15, p. 179, 186)

What volume of a platelet solution, stored at 20°C–24°C, is given to raise the platelet count by 50,000?

10 mL/kg should raise the platelet count by about 5.0×10^4/mL.

What is fresh-frozen plasma (FFP)?

Plasma from whole blood. About 80% of FFP is made up of plasma proteins.

When is FFP used?

FFP is primarily used to replace clotting factors. It is indicated when clotting factors are depleted such as in disseminated intravascular coagulopathy (DIC), or massive blood loss.

Which metabolite should be monitored with infusion of FFP?

Ionized calcium. FFP has a high concentration of citrate that can reduce ionized calcium, especially in infants.

How much FFP is needed to raise clotting factors by 10–20%?

10–15 mL/kg

List 3 of the therapeutic uses for immunoglobulin.

Replacement in immunodeficient patients; treatment of Kawasaki disease; to convey passive immunity to susceptible patients exposed to a variety of specific infections, such as tetanus, hepatitis B (Ch 20, p. 316), rabies, and varicella-zoster (Ch 28, p. 493)

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Name 2 uses of factor VII.

To control severe hemorrhage, including severe bleeding in hemophiliacs who have inhibitors to clotting factors, and to treat bleeding in patients with factor VII deficiency

What is cryoprecipitate?

A plasma preparation containing factor VIII, von Willebrand factor, and fibrinogen (Ch 15, p. 184)

List 4 risks that are associated with the use of blood products.

Risks vary with the type of product, the clinical situation, and the patient, but include infection (e.g., HIV, hepatitis B and C, CMV, and bacteria), sensitization, immune response, graft-versus-host reaction.

What should always be monitored when administering blood products?

Patient temperature. Fever may indicate a reaction of some type. Rapid administration of chilled products should also be avoided.

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Chapter 5 Pediatric Nutrition

Why do nutritional considerations in pediatric patients differ from those in adults?	Anabolic processes (growth, maturation, and development) increase the nutritional needs of children.
What is the caloric content of fat?	9 kcal/g
Of carbohydrates or protein?	4 kcal/g
What are the caloric requirements of a healthy term infant?	On average, 100 kcal/kg per day. More may be required for a premature infant.
What are the fluid requirements if provided by an enteral route?	About 150 mL/kg per day
Recommended protein intake of infants?	Approximately 2–2.2 g/kg per day (may be 3–3.5 g/kg per day for premature infants)
What is the caloric content of breast milk?	It varies; averages about 20 kcal/30 mL
Of commercial infant formula?	Most contain 20 kcal/30 mL
List 5 advantages of breast-feeding	Easily available, inexpensive, promotes mother-child bonding, less immunogenic, contains antibodies (which may reduce the incidence of infection)

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Do breast-fed infants require supplementation?

Yes. Breast-fed infants require **vitamin D, fluoride** (after age 6 months), and **iron** (after age 4–6 months) supplementation.

List 4 contraindications to breast-feeding.

1. Certain medications the mother may be taking (e.g., antimetabolites, chloramphenicol)
2. Certain maternal infections (e.g., HIV, active TB in the mother)
3. Maternal substance abuse
4. Abnormal gag reflex or swallowing in the infant

What is the best regimen for breast-feeding?

On-demand feeding until the milk supply has been established and feeding is going well. Intervals between feedings can be gradually increased as the duration of each feeding increases.

At what age does breast-feeding usually stop?

It varies. Some parents wean the child around 9 months of age, as the child learns to drink from a cup. Usually, breast-feeding does not extend beyond 18 months of age.

What are essential amino acids?

Amino acids that cannot be synthesized and must be acquired through the diet

List 8 essential amino acids.

Isoleucine, leucine, lysine, phenylalanine, threonine, tryptophan, valine, methionine

What other amino acids may be essential under certain conditions?

Histidine (infancy)

Tyrosine, cysteine, proline (possibly in premature infants)

Glutamine, arginine (possibly in stress and excess energy demands)

What are essential fatty acids?

Fatty acids that cannot be synthesized and must be obtained from dietary sources

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Name 2 essential fatty acids.	Linoleic acid and linolenic acid
List 4 symptoms of essential fatty acid deficiency.	Diarrhea, dermatitis, hair loss, and skin abnormalities (e.g., poor wound healing)
What is MCT oil?	A medium-chain triglyceride preparation that may be used as a calorie supplement
Name the fat-soluble vitamins.	Vitamins A, D, E, and K
Why is vitamin K given to newborns?	Newborns may be deficient in vitamin K, and administration of the vitamin helps prevent hemorrhagic complications caused by the deficiency of vitamin K–dependent coagulation proteins.
What is the primary carbohydrate in breast milk and in cow's-milk–based formulas?	Lactose
List 3 indications for using a lactose-free formula.	Galactosemia; lactose intolerance (either temporary or persistent); formula intolerance
When are "special" formulas used?	A variety is available for specific patients and clinical problems, including malabsorption, inborn errors of metabolism, protein allergies, and nutritional deficiencies.
When are solid foods introduced?	Usually 4–6 months of age. This varies widely depending on the preferences of the parents and the physician.
What solid food is introduced first?	Usually an iron-fortified single-grain cereal
Why use single-grain cereals?	A single grain allows easier identification of specific foods or ingredients that may not be tolerated by the infant.

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What kinds of foods should be avoided for young children?

Foods that are easily aspirated or need to be chewed by molar teeth. Examples include nuts, popcorn with kernels, hard candy, grapes, meat or chicken chunks.

Do older infants need vitamin supplementation?

Children on a well-balanced diet probably do not need vitamin supplements.

Do vitamin supplements have a role in treating children with mental retardation?

There are few studies to support their general use in these children. Children whose diets are inadequate or who have specific nutritional needs (or deficiencies) may benefit from specific supplements.

What is marasmus?

Wasting of muscle and subcutaneous fat from malnutrition

What is kwashiorkor?

Malnutrition in which there is relative protein deficiency; affected children are usually edematous.

Are vegetarian diets safe for children?

A well-planned vegetarian diet that contains all the essential amino acids is probably safe for children.

Will a vegetarian diet provide an adequate amount of vitamin B_{12}?

Because vitamin B_{12} comes from animal sources, strict vegetarians may be at risk for vitamin B_{12} deficiency.

How are caloric needs calculated for older children?

100 kcal/kg per day for first 10 kg

50 kcal/kg per day for second 10 kg

20–25 kcal/kg per day for each subsequent 10 kg

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INTRAVENOUS AND PARENTERAL ALIMENTATION

Note: Most hospitals have established intravenous alimentation protocols, many using computer templates for calculation of components. Students and house officers should familiarize themselves with these protocols.

What are the indications for intravenous alimentation?

Generally, an inability to maintain adequate fluid or nutritional balance by enteric (oral or feeding tube) fluid or nutrient intake

What is TPN (a.k.a. "central hyperalimentation")?

Total **p**arenteral **n**utrition—implies parenteral administration of sufficient calories and nutrients for growth and weight gain. TPN is administered via a central venous route.

List 6 components of TPN.

Nitrogen source (amino acids), calories (primarily from glucose), electrolytes, vitamins, minerals, water (lipids are administered in a separate preparation)

What are the 4 indications for TPN?

Severe GI disease, extensive bowel resection, inflammatory bowel disease, conditions that necessitate bowel rest or prohibit oral or enteric intake for an extended period of time

When should total (central) parenteral nutrition be used?

When a period of >2 weeks of intravenous alimentation is anticipated

When should peripheral hyperalimentation be used? (a.k.a. "peripheral parenteral nutrition" [PPN])

Usually, when a short-term need is anticipated, or when the peripheral alimentation is used as a supplement to enteral nutrition

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What is the main limiting factor of peripheral hyperalimentation?

The highly osmotic content of hyperalimentation solutions may be irritating to peripheral veins. Generally, 10–12.5% dextrose is the upper limit for peripheral infusion. Therefore, it is not usually possible to administer TPN via a peripheral vein.

What is "D10"?

10% dextrose (in water or another solution). This means 10 g of dextrose per 100 mL of fluid.

What is the caloric density of dextrose monohydrate?

3.4 kcal/g; for D10, this means 34 kcal/100 mL fluid

What is Intralipid?

A fat emulsion that serves as a source of fatty acids and calories. A 10% solution of Intralipid contains 1.1 kcal/mL.

How is Intralipid administered?

Intralipid cannot be mixed in the hyperalimentation solution and is usually given parenterally via a Y-connector.

What is the source of protein in TPN?

Usually, a commercially prepared amino acid or protein solution

How is centrally administered TPN initiated?

It usually begins with a 10% dextrose solution (with electrolytes) as the maintenance fluid, with incremental increases (by 2.5% per day) as tolerated, up to a 20% dextrose solution. The amino acid mixture is then added, and the infusion rate is increased until the desired intake is achieved. Intralipid is administered in an amount to provide appropriate balance of carbohydrate, protein, and fat calories.

How are vitamins administered?

Usually, in a mixture prepared for this purpose and added to the TPN

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How are TPN patients monitored?

Each shift: Glucose and ketones in urine—test each shift until regimen is established, then daily

Daily:

1. Weight
2. Strict intake and output measurements
3. Electrolytes and glucose—daily until regimen is established, then every 3 days (or weekly), depending on the protocol of the institution
4. Lipemia—visual checks

Weekly: CBC, total protein, calcium, magnesium, phosphorus, hepatocellular enzymes, bilirubin, creatinine, and serum triglycerides (if using lipids); obtain sample immediately before infusion

Monthly: Zinc, copper, and iron levels

List 12 complications of TPN. Infection (particularly line infections), hyperglycemia, hypoglycemia (if TPN is stopped too quickly), acidosis, abnormal liver function (particularly from cholestasis), hypocalcemia, hypomagnesemia, trace metal deficiency, hyperlipemia, hyperammonemia, thrombosis, hyperbilirubinemia

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Chapter 6 Pediatric Emergencies

RESUSCITATION IN CHILDREN

What principle should be applied first in all pediatric emergencies?

Remember the ABCs: airway, **b**reathing, and **c**irculation must be confirmed or established before further interventions. This may require placement of an oral airway or ETT and will almost always require an IV or intraosseous catheter.

In what 4 ways does the pediatric airway differ from that of adults?

The pediatric airway diameter is smaller; the larynx is cephalad; the tongue is relatively larger; and the narrowest part of the airway is the subglottic region. In addition, a child's occiput is larger, relative to body size, than that of an adult and thus affects body positioning when the airway is being treated.

Where is the easiest place to palpate a pulse in an infant?

The brachial artery—over the medial distal humerus

How does pediatric cardiopulmonary arrest differ from adult cardiopulmonary arrest?

Cardiopulmonary arrest typically begins with respiratory arrest in children and cardiac arrest in adults.

How should a newborn be stimulated to determine responsiveness?

Rub the infant's back or the soles of his or her feet.

What is the minimum resuscitation effort needed for every newborn?

Warm and dry the infant. Consider suctioning the oral and nasal secretions.

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What is the most common cause of pathologic bradycardia in the newborn?	Respiratory insufficiency
What historical information is important in resuscitation?	An **AMPLE** history: **a**llergies, **m**edications, **p**ast medical history, **l**ast meal, **e**vents of this illness

RESPIRATORY DISTRESS

What is respiratory distress?	A set of clinical signs observed when increased breathing work is required to compensate for hypoxia, hypercarbia, or airway obstruction
List 6 common clinical signs of early respiratory distress.	Anxiety, irritability, use of accessory muscles of respiration (e.g., nasal flaring, sternocleidomastoid, intercostals, abdominal musculature), tachypnea, tachycardia, hypertension
Describe the classic difference in physical examination findings distinguishing intrathoracic airway obstruction from extrathoracic airway obstruction.	Intrathoracic obstruction causes wheezing and a prolonged expiratory phase. Extrathoracic obstruction causes inspiratory stridor.
Are there any absolute laboratory or radiographic findings that define respiratory distress or failure?	No. Laboratory studies may help determine the cause or show trends, but they must be placed in the context of the physical findings and history.
List 6 common postoperative causes of respiratory distress.	Atelectasis, pulmonary edema, pleural effusion, pneumothorax, malpositioned ETT, aspiration
Posttraumatic causes? (List 5)	Pulmonary contusion; pneumothorax, hemothorax, or both; disruption of an airway; injury to the diaphragm

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Infectious causes? (List 6)

Pneumonia, pleural effusion, empyema, croup, epiglottitis, bacterial tracheitis

Causes from primary lung disease? (List 2)

Asthma, CF

Other causes? (List 2)

Aspiration of a foreign body, anatomic airway anomaly

What is the differential diagnosis of acute stridor in a child?

Croup, epiglottitis, bacterial tracheitis, peritonsillar abscess, retropharyngeal abscess, foreign body, laryngeal fracture, angioedema, caustic ingestion, vascular ring, diphtheria, subglottic stenosis

What happens if respiratory distress is not treated?

The patient may progress to **respiratory failure.**

List 4 criteria for evaluating the pediatric airway when determining the need for intervention.

Airway patency, airway anatomy, adequacy of respiratory effort, oxygenation

What 3 maneuvers can be done to achieve airway patency?

1. Elevate the child's shoulders (remember the large occiput causes neck flexion).
2. Position the child's jaw (i.e., chin thrust) to eliminate the occlusion of the airway by the tongue.
3. Suction any secretions.

What is the most useful medication in pediatric resuscitation?

Oxygen

When should intubation be undertaken?

When child is showing clinical signs of progression toward respiratory failure despite simple interventions, **even if laboratory values of ABG are acceptable** (Ch 8, p. 64, intubation in neonates)

What happens if respiratory failure is not treated?

Cardiac arrest

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How does the practitioner estimate the appropriate size (mm inner diameter) for an ETT?

1. Use the following formula:

$$\frac{\text{Child's age in years} + 16}{4}$$

2. Estimate the tube size based on the size (width) of the child's fifth finger

List 7 options for airway control.

Nasopharyngeal airway, oropharyngeal airway, orotracheal intubation, nasotracheal intubation, laryngeal mask airway, cricothyroidotomy, percutaneous transtracheal ventilation

Name 3 syndromes that would make a child difficult to intubate.

Down syndrome, Pierre Robin malformation complex, Treacher Collins syndrome

List 3 anomalies associated with Down syndrome that may make intubation more difficult.

A child with Down syndrome has macroglossia, a small trachea, and may have C1–C2 ligamentous instability.

ACUTE CARDIOVASCULAR COLLAPSE

CARDIAC ARREST

What is cardiac arrest?

Pulseless cardiac arrest is a clinical diagnosis based on the absence of a palpable central (femoral, brachial) pulse. It is accompanied by apnea. It may exist in the presence of electrocardiographic complexes.

What are the common causes of cardiac arrest in children?

Most pulseless arrests in children are the result of **severe hypoxemia** and **acidosis,** secondary to **respiratory failure** or **shock** (septic, most commonly).

What are the 7 causes of electromechanical dissociation (electrocardiographic complexes without cardiac contractions)?

Hypoxia, blood or fluid loss, tension pneumothorax, cardiac tamponade, electrolyte imbalance, profound hypothermia, drug overdose

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What is the initial approach to—and treatment of—cardiopulmonary arrest?

1. Confirm cardiopulmonary arrest and begin CPR. **(Remember, airway is always secured first! Airway, breathing, circulation—the ABCs!)**
2. Apply monitoring leads and confirm heart rhythm.
3. Obtain venous or intraosseous access.
4. Identify and treat causes.
5. Perform repeated cardiopulmonary assessments and respond to changes accordingly. **[See decision tree in Pediatric Advanced Life Support (PALS) manual.]**

What ECG syndromes must be recognized in order to identify the increased subsequent risk of ventricular tachycardia?

Brugada syndrome, prolonged QT syndrome, Wolff-Parkinson-White syndrome

When transport is necessary, what 4 important tasks should be included in the arrangements?

Contact the receiving unit. Copy and retain pertinent records. Obtain permission to transport. Remain with the patient while awaiting transport to perform and respond to continuing assessments.

What is the outcome of pediatric cardiac arrests?

Survival with an intact neurologic status after out-of-hospital cardiopulmonary arrest is rare. Organ systems other than the CNS are more resilient and may recover if the brain survives. Witnessed in-hospital arrest has a better, but still poor, prognosis for intact survival.

Why is the outcome of pediatric cardiac arrests so poor?

Children do not die of rapidly treatable cardiac arrhythmias. Children usually go into cardiac arrest as a complication of another end-organ illness, typically respiratory causes.

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SHOCK

What is shock?

A clinical state in which delivery of oxygen and metabolic substrates to tissues is inadequate to meet tissues' metabolic demands

What are the 2 stages of shock?

1. **Compensated shock:** BP is maintained within a normal range for age.
2. **Uncompensated shock:** Hypotension, with or without low cardiac output, is present.

Describe "THE MISFITS" pneumonic for the differential diagnosis of critically ill newborns.

T—trauma (including nonaccidental trauma)

H—heart disease

E—endocrine (CAH)

M—metabolic disturbances (hypoglycemia, hyponatremia)

I—inborn errors of metabolism

S—sepsis

F—formula dilution or overconcentration

I—intestinal catastrophes

T—toxins (home remedies)

S—seizures

What is hypovolemic shock?

Shock from loss of blood or body fluid

What are the causes of hypovolemic shock?

Blood loss from trauma or excessive bleeding; body fluid depletion from diarrhea, vomiting, or poor fluid or food intake

What is the earliest sign of hypovolemic shock in children?

Tachycardia

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What are the causes of distributive shock?

Total body fluid may be adequate but has left the intravascular space. Distributive shock may result from sepsis, anaphylaxis, or tissue or bowel swelling after surgery ("third spacing").

Name 4 common causes of anaphylaxis.

Hymenoptera envenomation (bee stings), peanuts, antibiotics, latex-containing medical equipment

Name 5 medications used to treat anaphylaxis.

IV fluids, diphenhydramine, methylprednisolone, epinephrine, albuterol

What is cardiogenic shock?

Cardiac pump failure; volume status may be low, adequate, or excessive.

List 3 common causes of pediatric cardiac pump failure leading to cardiogenic shock.

1. **Intrinsic cardiac disease:** including cardiomyopathy, myocarditis, structural heart disease, and dysrhythmia resulting in poor output
2. **Toxin-mediated failure:** including drug-induced state and sepsis-related mediators
3. **Hypoxia,** resulting in poor cardiac contractility

What are the clinical signs of compensated shock in children?

Cardiovascular signs? (List 3)

1. BP normal or high (Children maintain BP well until late in shock!)
2. Heart rate usually above normal range for age
3. Decreased peripheral perfusion (peripheral pulses thready or absent; capillary refill time > 2–3 seconds; skin temperature cool)

Respiratory signs? (List 2)

1. Respiratory rate is often increased as compensation for metabolic acidosis.
2. Work of breathing may be increased.

Renal signs?

Decreased urine output

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CNS signs?

Child may be agitated, combative, or lethargic

What are the clinical signs of uncompensated shock?

Cardiovascular signs? (List 3)

1. BP below normal range for age
2. Heart rate remains elevated although bradycardia may occur in late shock.
3. Decreased peripheral and central perfusion (peripheral signs of hypoperfusion combined with thready central pulses)

Respiratory signs? (List 2)

1. Respiratory rate may remain elevated or may have fallen despite metabolic acidosis.
2. Work of breathing may be at an elevated, or inappropriately low, rate.

Renal signs?

Decreased urine output

CNS signs?

Child is usually combative or lethargic; may fail to recognize a parent, appear apathetic even with painful stimuli, or be unresponsive or comatose.

What are the 2 laboratory signs of shock in children?

1. **Metabolic acidosis,** with or without respiratory compensation; elevated lactate implies poor tissue perfusion.
2. **Hypoglycemia** may be secondary to shock or cause shock.

What are the physiologic consequences of shock?

Inadequate tissue perfusion causes end-organ dysfunction and eventually **irreversible organ damage if untreated.**

What is the differential diagnosis of shock in an infant?

Sepsis, meningitis, pneumonia, heart disease (acquired or congenital), metabolic disorder, toxic ingestion, congenital adrenal hypoplasia, dehydration, posterior urethral valves, malrotation with midgut volvulus

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What are the 2 primary electrolyte abnormalities seen in congenital adrenal hypoplasia?

Hyperkalemia and hyponatremia

What is the most common category of symptoms in infants with infections, including meningitis?

Respiratory symptoms

List 2 common symptoms in children with pulmonary edema or cardiogenic shock.

Tachypnea; difficulty or sweating with feeding

What are the 5 cyanotic congenital heart lesions?

1-2-3-4-5 Ts:

Truncus arteriosus (**1** vessel)

Transposition of the (**2**) great vessels

Tricuspid atresia

Tetralogy of Fallot

Total anomalous pulmonary venous return (has **5** words)

Distinguish central cyanosis from peripheral cyanosis.

Central cyanosis describes arterial hypoxia (examination may reveal blue color around lips—perioral cyanosis). Peripheral cyanosis describes cyanosis as a result of poor perfusion—as may occur in the extremities during hypovolemic shock.

What is the initial treatment of shock caused by transposition of the great vessels?

Infusion of prostaglandin E_1

List 3 complications of prostaglandin E_1 infusion.

Apnea, agitation, hyperthermia

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What is the diagnostic approach for shock?

Identification of shock should be **clinical,** based on the interpretation of the physical findings outlined earlier. Determination of the cause is useful in later treatment but should not delay initial stabilization effort.

What is the reason cardiogenic shock must be differentiated from other forms of shock?

It **requires a different treatment approach** after initial interventions than other forms of shock. In cases of cardiogenic shock, the heart may have adequate preload (end-diastolic pressure); thus, IV fluid boluses may be ineffective or deleterious.

Treatment

List 2 approaches for definitive treatment of shock.

1. Resuscitation (should be used in all cases of shock)
2. Identification and treatment of the underlying cause of shock

What are the basic steps of resuscitation?

Remember the ABCs!

1. Immediately provide **oxygen** through an **adequate airway** (native or artificial).
2. If bradycardia and poor perfusion present, proceed as for cardiac arrest.
3. Achieve vascular access when shock is recognized.
4. Provide an **initial fluid bolus of 20 mL/kg,** using NS or LR. In infants and children, administer fluid using a large syringe and stopcock (not a "wide open" infusion) to allow rapid administration. Repeat as indicated, while avoiding inadvertent overresuscitation.
5. Obtain a bedside **glucose** measurement and treat with 0.5–1.0 g/kg of IV dextrose if patient is hypoglycemic. Consider obtaining other diagnostic studies (e.g., blood studies or radiographs), **but ABCs are always first!**

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6. Reassess **peripheral perfusion** (pulse strength and capillary refill time), **fluid balance** (liver size, urine output), **breath sounds** (work of breathing), and **vital signs.** Repeat fluid boluses with frequent reassessment until patient is no longer in shock or until signs of fluid overload are noted. Colloid may be required as a fluid replacement if colloid (e.g., blood) has been lost.

Remain aware that shock may continue until the predisposing condition has been corrected.

What medications can be used for resuscitation from shock (especially cardiogenic shock) if preload is adequate?

Initiate inotropic support via intraosseous or central venous access.

Doses:

Dopamine: 5–20 μg/kg per minute

Epinephrine: 0.05–1 μg/kg per minute

List 2 complications of treatment of shock.

1. Worsened end-organ damage may occur because of inadequate resuscitation.
2. Overzealous resuscitation may cause fluid overload with resulting pulmonary edema.

What fluid should be administered to a child with seizures due to hyponatremia?

3% saline (hypertonic saline). The amount given should be carefully calculated to avoid rapid fluid shifts.

What is the most feared complication in cases of hyponatremic dehydration?

Central pontine myelinolysis

What may result from overly aggressive treatment of hypertonic dehydration?

Cerebral edema with increased ICP

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ACUTE NEUROLOGIC CONDITIONS AND MENTAL STATUS CHANGES

HEAD TRAUMA

What kind of injury is most responsible for childhood deaths beyond 1 year of age?	Head trauma
What percentage of children with head trauma have a skull fracture?	Approximately 35%
What percentage of children with skull fractures have an ICH?	Approximately 30%
How should a child with a severe head injury be approached?	**ABCs**—**a**irway, **b**reathing, and **c**irculation—are the top 3 priorities. **Cervical spine immobilization** must be maintained.
What is the most common cause of head trauma in children?	Motor vehicle crashes
What findings suggest "shaken baby" head injury?	Subdural hematoma (classically parafalcine) and retinal hemorrhages
What is a concussion?	Impact to the head with any alteration in consciousness with or without amnesia of the events immediately surrounding the injury (Ch 13, p. 142)
What is a subdural hematoma?	A collection of blood between the dura and cerebral mantle
What is an epidural hematoma?	A collection of blood in the extradural space, usually caused by a rupture of the middle meningeal artery or tears in the dural vein; therefore, **blood usually collects rapidly.**

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What is a typical course of symptoms after an epidural hematoma in an awake patient?

A child will have suffered a **concussion (possibly very brief!).** Often there is a **lucid interval** before the onset of vomiting, headache, and focal neurologic signs.

What is the treatment for a subdural or an epidural hematoma?

Surgical decompression

Why is cerebral edema harmful?

It causes an increase in ICP, thus compromising cerebral blood flow, resulting in further cerebral ischemia.

What is the formula for cerebral perfusion pressure?

Cerebral perfusion pressure (CPP) = mean arterial pressure (MAP) − ICP

List 2 common causes of increased ICP.

1. Cerebral edema (resulting from cell death, i.e., hypoxia or inadequate perfusion, or cellular swelling after overaggressive fluid resuscitation in hyperosmolar conditions, i.e., hypernatremic dehydration)
2. Mass effect (diffuse, i.e., meningitis, or focal, i.e., tumor or ICH)

Either cause may be induced by trauma or by medical conditions.

List 6 specific interventions to treat cerebral edema.

(These interventions must be individualized to the needs of the patient.) Oxygenation, elevation of the head of the bed, judicious use of IV fluids, adequate ventilation or hyperventilation, monitoring of ICP, and administration of mannitol

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What must ultimately be optimized when treating cerebral edema?	**CPP.** Many maneuvers designed to reduce ICP and cerebral edema may also reduce cerebral blood flow (e.g., hyperventilation, diuresis with mannitol, phenobarbital). Appropriate resuscitation is mandatory and pressors may be necessary to maintain appropriate CPP (>50 mm Hg in younger children and >60 mm Hg in older children).
What is the most important determinant of neurologic outcome after head injury?	Duration of coma
How does the outcome for children compare with that of adults if the brain injuries are similar?	Children generally do better.
How does the outcome for children younger than 2 years compare with that of older children if the brain injuries are similar?	Children younger than 2 years generally do worse.

SEIZURES

What is a seizure?	Clinical manifestation of synchronized electrical discharges of CNS neurons
What are the signs and symptoms of motor seizures in children?	They are characterized by patterns of movement that relate to the patterns of electrical discharges and are usually, but not always, accompanied by impaired consciousness.
Of nonmotor seizures?	They may be manifested only by the loss of responsiveness to the environment and may be mistaken for coma without seizures.
Name 4 other conditions that may mimic seizures	Breath-holding spells, reflux esophagitis (Sandifer syndrome), cardiogenic syncope, pseudoseizures

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How are seizures classified? See Table 6–1

Table 6–1. International Classification of Epileptic Seizures

Classification	Subclassification
Partial (focal, local)	
Simple partial seizure	With motor signs
	With somatosensory or special sensory symptoms
	With autonomic signs or symptoms
	With psychic symptoms
Complex partial seizure	Simple partial onset followed by impairment of consciousness
	With impairment of consciousness at onset
Partial seizures evolving to secondarily generalized seizures	Simple partial seizures evolving to generalized seizures
	Complex partial seizures evolving to generalized seizures
	Simple partial seizures evolving to complex partial seizures evolving to generalized seizures
Generalized (convulsive and nonconvulsive)	
Absence seizure	Typical absence
Myoclonic seizure	Atypical absence
Clonic seizure	
Tonic seizure	
Tonic-clonic seizure	
Atonic (astatic) seizure	
Unclassified epileptic seizure	

What are the physiologic results of brief seizures (seconds to a few minutes)? It is believed that brief seizures do not harm brain tissue, but the child may sustain secondary injury (e.g., may aspirate, strike head, drown, become hypothermic) through loss of protective reflexes.

What are the physiologic results of prolonged seizures (>30 minutes)? They may cause direct injury to the brain tissue through electrolyte shifts, increased cerebral blood flow, and elevated ICP.

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What are the common pathologic causes?

See Table 6–2

Table 6–2. Common Pathologic Causes of Seizure

Fever
Infection
Encephalitis
Meningitis
Abscess
Traumatic brain injury
Hypoxic-ischemic injury
Metabolic disorders
Acute electrolyte abnormalities
Genetic metabolic disease
Drugs or toxins
Infantile spasms
Brain malformation or tumors
Primary or idiopathic seizure disorders
Benign neonatal seizure
Absence epilepsy
Generalized tonic-clonic seizure
Myoclonic seizure

Overdose of what medication may result in refractory seizures and what is the treatment?

Isoniazid-induced seizures should be treated with pyridoxine (vitamin B_6).

List 5 characteristics of a simple febrile seizure.

Generalized tonic-clonic, lasts <15 minutes, occurs in children 3 months to 5 years of age, occurs on day 1 of illness with high fever, and there may be a family history of febrile seizures.

List 5 characteristics of a complex febrile seizure.

Lasts ≥15 minutes; partial or focal seizures may occur; multiple seizures in 1 day may occur; neurologic deficits or developmental delay may be predispositions; family history may include nonfebrile seizures.

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Treatment

What is the main goal of treatment?

Stopping seizure activity rapidly without compromising the patient

List 3 immediate priorities in caring for a child who is having a seizure.

1. Evaluate and support airway and breathing.
2. Obtain blood for chemistries (e.g., calcium, sodium, glucose), drug levels, and suspected toxins that may require immediate intervention.
3. Gain venous access.

What are the next treatment priorities (5–20 minutes after presentation)?

Administer anticonvulsants while monitoring cardiorespiratory status:

1. **Rapid-onset, short-acting agents** (e.g., lorazepam). Be prepared to intubate the patient if respiratory depression occurs.
2. **Delayed-onset, long-acting agents** (e.g., phenytoin, phenobarbital)

List 3 diagnostic options that are used to evaluate a patient with seizures during and after initial treatment.

1. **Physical examination** during the episode may identify the type of seizure, but not of the cause.
2. **Electroencephalography,** with or without video monitoring, may be useful but should not delay treatment.
3. **Laboratory and radiographic studies** (such as serum electrolytes and glucose, MRI, lumbar puncture) identifying infection, trauma, toxins, and so forth may be helpful, depending on the circumstances.

What is the outcome?

Brief seizure—good if no complications occur; damage may still occur because of the underlying cause.

Prolonged seizure—variable, but should be anticipated to be worse than that from a brief seizure

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UNEXPLAINED COMA

What is coma?

A state of unconsciousness from which one cannot be aroused by stimulation of any magnitude

What is the differential diagnosis of coma in an infant or child?

AEIOU TIPS

Alcohol/Abuse

Epilepsy/Encephalopathy

Infection/Inborn errors of metabolism

Opiates

Uremia

Trauma/Tumor

Insulin (hypoglycemia)/Intussusception (with severe bowel compromise)

Poisoning (toxicology)

Shock

What initial laboratory and imaging studies are appropriate?

The choice of laboratory studies and imaging studies should be guided by the findings of the physical examination and by the patient's history combined with the differential diagnosis listed earlier.

What role does rapid CT of the brain play in the treatment of the child in coma?

Head CT will rule out rapidly progressive CNS mass lesions that may result in decreased CPP or a cerebral herniation syndrome.

ENVIRONMENTAL EMERGENCIES

DROWNING

What is near drowning?

A submersion incident followed by survival for at least 24 hours, regardless of the ultimate outcome

What is drowning?

A submersion incident that results in death within the first 24 hours

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List 4 epidemiologic characteristics of near drowning in children and infants.

1. Near drowning is more common in boys than in girls.
2. High-risk groups: toddlers, adolescent males, and children with seizure disorders
3. Most drowning or near drowning in small children occurs during brief periods (<5 minutes) without supervision, not as a result of neglect.
4. Most near-drowning incidents occur in residential swimming pools but can occur in any available body of water, including lakes, rivers, 5-gallon buckets (often used for storing liquids), bathtubs, toilet bowls, hot tubs, and standing water on pool covers.

List 3 appropriate initial interventions (before child is in the emergency department).

1. **On-site basic life support, including ABCs with stabilization of the cervical spine,** is initially appropriate for all cold-water (temperature < 5°C) submersion victims—and arguably for all warm-water submersion victims—unless the submersion is known to be exceptionally prolonged.
2. Avoid excessive airway manipulation or pressure on the abdomen to avoid emesis and aspiration.
3. Escalate intervention to advanced life-support measures if needed, with continuation until the patient is evaluated in the emergency department. Many clinicians recommend continuing resuscitative efforts until the patient's core temperature exceeds 32°C, because existence or maintenance of a spontaneous cardiac rhythm may not occur below this temperature.

How long should cardiopulmonary resuscitation be continued in a submersion victim who arrives in the emergency department without spontaneous circulation?

This issue is controversial. Many clinicians recommend discontinuing efforts if advanced life-support measures fail to restore spontaneous circulation once the patient has reached a core temperature of 28°C; others suggest using 32°C as a temperature goal.

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List 4 indicators of poor outcome after near drowning.

1. Documented submersion > 5 minutes
2. The need for CPR in the emergency setting
3. A serum pH < 7 or fixed and dilated pupils in the emergency setting
4. Need for cardiotonic drugs during resuscitation

However, the only factor consistently predicting poor outcome is the need for continued CPR in the emergency department for nonhypothermic patients.

What is the role of prevention?

Because near-drowning outcomes are largely determined by the degree of the initial hypoxic insult, prevention is of paramount importance. Current areas of focus include improved legislation for barrier requirements around pools, education of the public about drowning risks in the home and around natural bodies of water, and encouragement of CPR training for pool owners.

HYPOTHERMIA

What is hypothermia?

Core body temperature < 36°C

List 2 reasons for infants being at higher risk for hypothermia than adults.

1. Their ratio of body surface area to weight is higher than that of adults.
2. Infants do not have the motor control to cover themselves.

Define mild, moderate, and severe hypothermia.

Mild: Core temperature 32°C–35°C

Moderate: Core temperature 28°C–32°C

Severe: Core temperature < 28°C

Describe the symptoms of mild, moderate, and severe hypothermia.

Mild: Shivering, loss of fine motor control, confusion

Moderate: Delirium, slowed reflexes

Severe: Cardiac arrhythmias common, coma

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List 3 common causes of hypothermia.

Induced hypothermia (e.g., intraoperative heat loss, infusion of large volumes of cold fluid), exposure (especially after trauma or immersion), CNS depression (hypothalamic dysfunction, infection)

How is hypothermia diagnosed?

By rectal or other core temperature measurement (e.g., esophageal probe)

List 2 ways it is treated.

1. Eliminate ongoing heat loss.
2. Gradually rewarm the patient at approximately 1°C/h.

List 3 methods for rewarming.

1. **Surface rewarming:** blankets, heat lamp
2. **Core rewarming:** warm inhaled gases, warm IV fluids, peritoneal lavage
3. **Extracorporeal techniques:** cardiopulmonary bypass

What must be carefully monitored during rewarming?

Electrolytes and **acid-base status;** acidosis and associated hyperkalemia may worsen during rewarming. **Avoid burning tissue with heat lamps.**

How long should a patient with accidental hypothermia be resuscitated?

This issue is controversial. Neurologic signs are absent at <25°C–27°C; defibrillation is difficult at temperatures < 30°C. Exercise clinical judgment. A usual rule of thumb: **A patient is not dead until he is warm and dead!**

MAJOR TRAUMA: PEDIATRIC ASPECTS

What are the 3 major points to remember in any pediatric trauma situation?

The **ABCs: a**irway, **b**reathing, **c**irculation

What is the best way to establish access for circulatory resuscitation in the event of trauma?

Establishment of **2 large-bore peripheral IV lines.** The size of the IV line should be appropriate for the size of the infant or child (Ch 2, p. 9).

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What access route is used if IVs are unsuccessful?

An intraosseous line—a 16-gauge IV needle, or a bone marrow aspiration needle, or a needle specific for intraosseous infusion is placed directly into the bone marrow.

What are the 2 preferred sites for the placement of an intraosseous line?

1. **Proximal tibia** (most preferred) approximately 1 fingerbreadth below the tibial tuberosity
2. **Distal femur** (second preferred) approximately 1 fingerbreadth above the knee

How long may intraosseous lines stay in place?

No longer than **6 hours**

What is the circulating blood volume of an infant or toddler?

80 mL/kg

What resuscitative strategy should be used for the infant or child who has experienced trauma?

The first IV bolus should be **20 mL/kg of isotonic crystalloid.** Next, administer a 2nd 20-mL/kg bolus if necessary. If additional resuscitation is necessary, follow this with a bolus of 20 mL/kg of packed red blood cells. **All IV fluids and blood products should be warmed!**

What are the 5 most likely sites of ongoing blood loss after trauma?

Abdomen, chest, retroperitoneum, femur fracture, intracranial bleeding in infants **(Note: In all patients except infants, intracranial bleeding alone does not account for hemodynamic instability.)**

What is characteristic about the child's ability to compensate for intravascular volume loss?

Generally, BP is maintained until 25–40% of intravascular volume is lost. Therefore, watch for **tachycardia** and **decreased urine volume. When BP begins falling, the child may be exhibiting circulatory collapse.**

Why can children maintain BP with a greater degree of hemorrhage than adults can?

Pediatric cardiac output is more heart rate related. Adult cardiac output is more preload dependent.

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List the 3 best signs of adequate circulatory resuscitation.

Adequate or appropriate urine output, pulse, and BP

Why do children have pulmonary contusions more frequently than adults?

Relatively softer ribs and compliant rib cage do not absorb the energy of impact.

Give 2 reasons that children suffer spleen and liver injuries more commonly than adults.

1. Small iliac crests; automobile lap belts restrain the abdomen and not the pelvis.
2. Large spleens and livers, relative to their body size, protected by soft ribs with compliant rib cage and relatively thin abdominal muscles.

How can many of these injuries be prevented?

Education about the proper use of car seats and boosters can decrease the risk of severe injury.

Describe the Salter-Harris classification (I–IV) of fractures

I. **S**ame: Fracture of the growth plate along the cartilage of the physis
II. **A**bove: The fracture is through and *above* the physis.
III. **L**ower: The fracture is through and *below* the physis in the epiphysis.
IV. **T**hrough: The fracture is *through* the metaphysis, physis, and epiphysis.
V. **ER**ased (crushed): The physis is *crushed*.

What is the significance of fractures through the physis?

Growth arrest may occur.

Name 5 fracture patterns associated with nonaccidental trauma?

Spiral fractures of long bones (except "toddler's fractures"), posterior rib fractures, metaphyseal fractures (corner or "buckethandle" fractures), hand fractures, compression fractures of the thoracic or lumbar spines

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What is the role of the health care provider in cases of nonaccidental trauma?

Most states have laws requiring health care providers to report cases of suspected nonaccidental trauma to appropriate social service organizations. The health care worker should determine whether a particular injury is consistent with the mechanism of injury as described by the caretaker.

TOXICOLOGY

Name 7 agents that may result in hypotension.

Anticholinergics, cyclic antidepressants, iron, theophylline, β-adrenergic antagonists, calcium-channel blockers, diuretics

Name 3 agents that may result in hypertension.

Anticholinergics, amphetamines, sympathomimetics

Name 3 agents that may result in bradycardia.

β-adrenergic antagonists, calcium-channel blockers, organophosphates

Name 4 agents that may result in tachycardia.

Anticholinergics, cyclic antidepressants, ethylene glycol, iron

Name 4 agents that may result in tachypnea.

Ethylene glycol, methanol, salicylates, sympathomimetics

Name 4 agents that may result in hyperthermia.

Amphetamines, anticholinergics, cyclic antidepressants, salicylates

Name 3 agents that may result in hypothermia.

Ethanol, oral hypoglycemics, opioids

Name 3 agents that may result in miosis.

Opioids, organophosphates, clonidine

Name 4 agents that may result in mydriasis.

Antihistamines, belladonna alkaloids, cyclic antidepressants, sympathomimetics (phenylephrine)

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Describe the MUDPILES acronym for increased anion gap metabolic acidosis.

Methanol

Uremia

Diabetic ketoacidosis

Paraldehyde

Iron or isoniazid

Lactic acid

Ethylene glycol

Salicylates

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Chapter 7 Growth and Development

GROWTH

What is the most important feature to remember about growth?	Growth is a **dynamic,** not a static, process.
What is the best way to evaluate growth?	Longitudinally along a time line, either by direct observation or by evaluation of accurate historical data
Why are growth charts important?	They are a record of data points, allowing easy comparisons with previous points and standard growth patterns to facilitate growth data analysis. (Samples of selected growth charts, as published by the NCHS, in collaboration with the National Center for Chronic Disease and Health Promotion [Centers for Disease Control and Prevention—CDC] are shown in Figs. 7–1 and 7–2.)
What is growth rate?	Change in a growth variable with time. When evaluating growth abnormalities, the growth rate is frequently more important than an isolated data point.
When is growth most rapid?	Relative growth is most rapid during fetal development. Adolescence is the time of greatest postnatal growth.
What is the normal rate of weight gain for infants?	After the initial postnatal water loss (about 5–10% of birth weight) during the first few days after birth, an infant should gain about 1 oz (30 g) per day.

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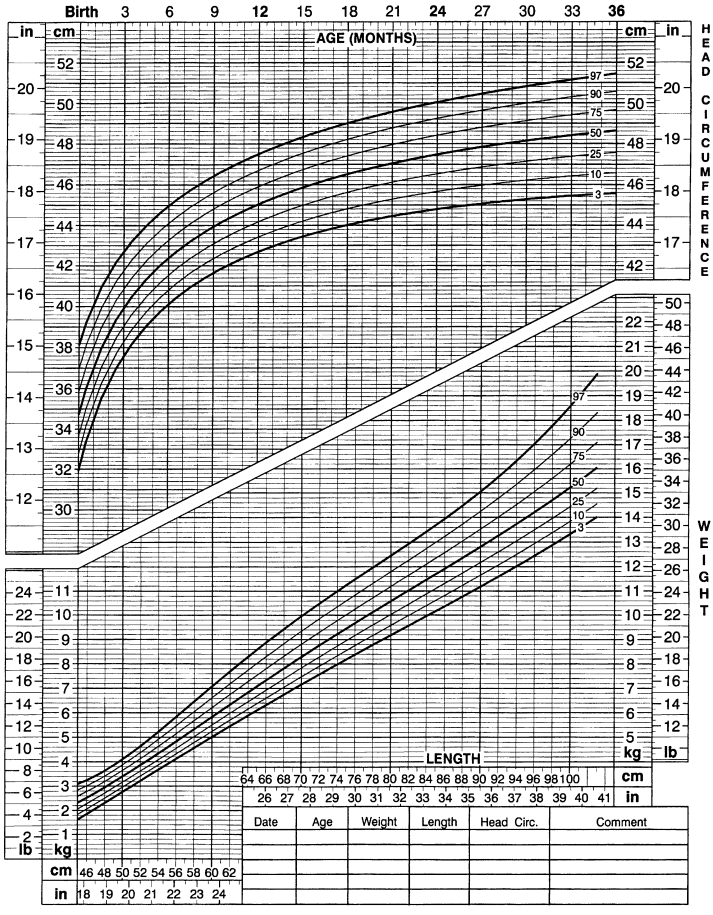
Birth to 36 months: Girls
Head circumference-for-age and
Weight-for-length percentiles

NAME _____

RECORD # _____



SOURCE: Developed by the National Center for Health Statistics in collaboration with the National Center for Chronic Disease Prevention and Health Promotion (2000). http://www.cdc.gov/growthcharts

Figure 7–1. Growth chart for girls from birth to 36 months: head circumference-for-age and weight-for-length percentiles. (Source: National Center for Health Statistics in Collaboration with the National Center for Chronic Disease Prevention and Health Promotion. Revised November 21, 2000. http://www.cdc.gov/growthcharts.)

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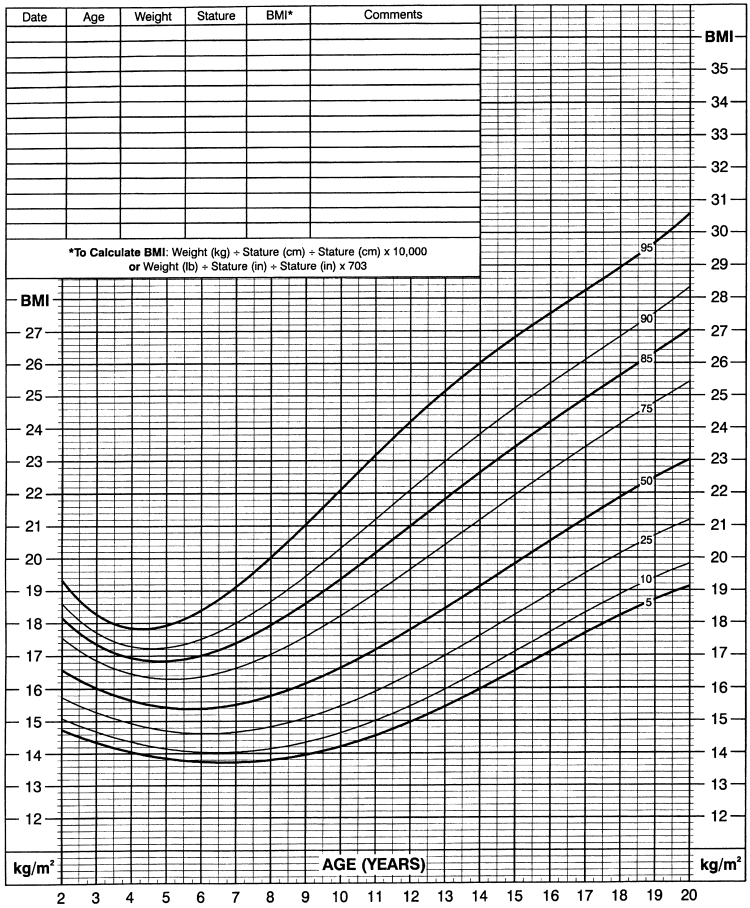
2 to 20 years: Boys
Body mass index-for-age percentiles

NAME _____

RECORD # _____

Date	Age	Weight	Stature	BMI*	Comments

***To Calculate BMI**: Weight (kg) ÷ Stature (cm) ÷ Stature (cm) x 10,000
or Weight (lb) ÷ Stature (in) ÷ Stature (in) x 703



SOURCE: Developed by the National Center for Health Statistics in collaboration with the National Center for Chronic Disease Prevention and Health Promotion (2000).
http://www.cdc.gov/growthcharts

Figure 7–2. Growth chart for boys 2–20 years of age: body mass index-for-age percentiles. (Source: National Center for Health Statistics in Collaboration with the National Center for Chronic Disease Prevention and Health Promotion, 2000. http://www.cdc.gov/growthcharts.)

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At what age does an infant double his birth weight?	Usually at 6 months of age
What height and weight should a normal child be at 4 years of age?	About 40 inches tall and 40 pounds
What is meant by the "upper:lower segment ratio"?	The ratio of the length of the upper segment (i.e., distance from top of the pubis to top of the head) to the length of the lower segment (i.e., distance from the top of the pubis to the bottom of the feet)
How do upper:lower segment ratios vary with age?	Infants and young children have relatively short lower limbs and relatively large heads, so the upper:lower ratio is higher in young children than in adults.
What is the normal upper:lower segment ratio at birth?	1.7:1
What is the normal upper:lower segment ratio at 10 years of age?	1:1
What other variables are important in interpreting upper:lower segment ratios?	Normal values for upper:lower segment ratios can vary with sex and with race or ethnicity.
At what age do first teeth usually erupt?	About 5–8 months; central mandibular incisors usually appear first.
Which permanent teeth usually appear first and when?	First molars ("6-year molars") around 5–7 years

DEVELOPMENT

Why is development an important pediatrics issue?	It reflects neurologic maturation and social and sensory development in the child. Abnormal development may reflect a neurologic, medical, or social problem.

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How does sensory impairment affect development?	Children with undetected hearing or visual deficits may not develop certain skills at the appropriate age.
What areas of development are monitored?	Gross motor, fine motor, social, and language development (should be included in the routine physical examination)
What are the representative gross motor milestones at the following ages:	
1 month?	Lifts head from prone position
2 months?	Holds head upright without wobble
3 months?	Regards hand
4 months?	Purposeful grasp; rolls front to back
6 months?	Beginning to sit without support; rolls back to front
9 months?	Sits well without support; up on all fours; crawls
8–10 months?	Pulls to standing; walks with support
13–15 months?	Walks independently
18 months?	Walks up and down stairs; begins to run
24 months?	Jumps in air
What are the representative fine motor milestones at age:	
2 months?	Follows visually past midline
3–4 months?	Grasps objects and brings to mouth
6 months?	Places objects carefully rather than dropping them, transfers hand-to-hand
9 months?	Clasps hands
9–10 months?	Demonstrates pincer grasp

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15 months?	Scribbles; stacks 2 cubes
18 months?	Stacks 4 cubes
24 months?	Stacks 8 cubes

What are the representative social milestones at age:

6–8 weeks?	Smiles responsively
2 months?	Smiles when seeing mother
4 months?	Smiles spontaneously
6 months?	Copies facial expressions
9 months?	Fears strangers; shows separation anxiety; plays interactively (peekaboo, patty-cake)
12 months?	Drinks from cup and finger—feeds self
15 months?	Uses spoon; imitates adult actions
18 months?	Removes clothing; uses cup
24 months?	Begins toilet training; puts on clothing

What are representative language milestones at age:

2 months?	Coos responsively
4 months?	Social laughter
6 months?	Makes nonspecific vowel sounds
9 months?	"Dada" and "Mama" (nonspecific)
12 months?	"Dada" and "Mama" plus 2 other words
15 months?	Several more words
18 months?	Combines 2 words into phrases
24 months?	Combines 3 or more words

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36 months?	Most speech clear to strangers
48 months?	Toddler stuttering subsiding
What is the Denver Developmental Screening Test?	A screening test developed for the quick assessment of developmental milestones
Does developmental delay imply later mental retardation?	No. Mental retardation usually refers to cognitive and problem-solving deficits, whereas developmental delay includes a wide range of skills that may be adversely affected by medical or neurologic problems that may not affect cognition. Mental retardation implies a permanent deficit, whereas aspects of developmental delay may be temporary or permanent depending on the causes.

PUBERTY

(Also see Chapter 14)

What is puberty?	The development of secondary sexual characteristics and the maturation of gonadal function
When does puberty usually begin in boys and what is the usual progression?	Around 11.5 years of age; begins with enlargement of the testes, followed by appearance of pubic hair and linear growth spurt
What is the normal age range of onset of puberty in boys?	Approximately 9.5–13.5 years
When does puberty usually begin in girls and what is the usual progression?	Around 10.5 years of age; usually begins with breast buds (thelarche), followed by appearance of pubic hair, growth spurt, then menarche
What is the normal age range of onset in girls?	8–13 years

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When does the maximum growth velocity occur?	Usually 1.5 years after the beginning of puberty
When does menarche occur?	Usually 2 years after appearance of breast buds
What is precocious puberty?	Onset of puberty before age 8 (females) or 9 (males)
Is precocious puberty more common in boys or girls?	Girls
What is the most common cause of precocious puberty in girls?	Idiopathic precocious puberty (Ch 24, p. 375)
What is delayed puberty?	If there has been no development of secondary sexual characteristics by age 13 (girls) or 14 (boys) (Ch 24, p. 379)

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Section II Newborn Care

Chapter 8 Perinatal Care and Evaluation of the Newborn

APGAR SCORES

What is the Apgar score?	A method of evaluating a newborn introduced in 1953 by Dr. Virginia Apgar. **Five** physical signs are identified, and a score of **0, 1,** or **2** is given to each sign at 1 minute and 5 minutes after birth.
What are the 5 signs evaluated, and what constitutes a score of 0, 1, and 2 for each sign?	See Table 8–1 for the signs and Apgar evaluation scoring.
What is the APGAR mnemonic?	**A**ppearance, **p**ulse, **g**rimace, **a**ctivity, and **r**espirations
What do the 1- and 5-minute scores imply?	The 1-minute score indicates the infant's initial condition. The 5-minute score indicates the infant's improvement, continued well-being, or subsequent decline (depending on the initial score).
Should additional Apgar assessments be made beyond 5 minutes?	If the Apgar score is **<7 at 5 minutes,** checking of Apgar scores every 5 minutes for the subsequent 20 minutes is helpful to assess resuscitation efforts.

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Table 8–1. The Apgar Evaluation Scoring

	Scores		
Sign	**0**	**1**	**2**
Color	Central cyanosis	Peripheral cyanosis	Completely pink
Heart rate	Absent	<100 bpm	>100 bpm
Reflex irritability	No response	Grimace	Cough or sneeze
Muscle tone	None	With some flexion	Well flexed or spontaneous movement
Respiratory effort	Absent	Irregular or weak cry	Regular or strong cry

NEWBORN RESUSCITATION

INITIAL ASSESSMENT

List 3 components of initial assessment and management of a newborn.

1. Gentle suction of the mouth, nose, and pharynx with a bulb syringe or suction catheter
2. Drying of the infant to minimize evaporative heat loss with warming via radiant heat
3. Evaluation of the infant's color, respiratory effort, and heart rate

What are the potential risks of suctioning?

Deep suctioning should be avoided in the initial resuscitation because this may induce laryngeal spasm, increase vagal tone resulting in apnea and bradycardia, or result in trauma to the pharynx or esophagus.

INTUBATION

List 5 signs of inadequate oxygenation and ventilation.

1. Poor color
2. Poor (or lack of) responsiveness
3. Lack of movement of chest with bag-and-mask ventilation
4. Falling oxygen saturation
5. Falling heart rate (normal newborn heart rate 120–160 beats/min [bpm])

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What is the appropriate positioning for bag-and-mask ventilation?

Placement of the mask over the mouth and nose, with the head and neck slightly extended

What is the primary indication for intubation?

Inability to oxygenate and ventilate an infant adequately via bag-and-mask ventilation

What sizes of ETTs are appropriate for infants?

Uncuffed tubes with internal diameters of 2.5, 3.0, or 3.5 mm, depending on the infant's size (Ch 6, p. 33)

List 6 important anatomic and position considerations during intubation.

1. The infant's head and neck should be slightly extended.
2. The larynx is more anterior and caudad in the neonate than in the older child or adult.
3. The epiglottis tends to hang over the vocal cords.
4. A straight-blade laryngoscope with a Miller 0 or 1 blade is usually most useful for visualizing the airway.
5. The ETT should be placed only 1–1.5 cm beyond the cords to avoid mainstem (usually right-sided) intubation. A weight-to-distance correlation of 1–2–3 kg:7–8–9 cm (i.e., tube marking at infant's lip) is extremely useful for determining the appropriate distance for tube placement.
6. The chest should be auscultated for symmetric breath sounds over both lung fields and absence of sounds over the stomach. There should be symmetric chest rise, and ideally the infant's heart rate and color will improve. End-tidal CO_2 monitor or CO_2 detector can confirm tracheal intubation. Chest x-ray confirms the appropriate placement of the tube.

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UMBILICAL VESSEL CATHETERIZATION

When are UVCs used?	Primarily in premature and acutely ill infants
List 3 purposes of a UVC.	1. Resuscitation—for emergency vascular access for administration of drugs and fluid 2. Administration of intravenous fluids, hyperalimentation, and medications 3. Exchange transfusion; sampling blood when needed
Where should the tip of the UVC lie?	The junction of the right atrium and the inferior vena cava (level of T10–11)
List 7 potential complications of a UVC.	Vascular embolization, vascular spasm, vascular perforation, infection, hemorrhage, venous congestion of the lower extremities, and thrombosis (e.g., thrombosis of the portal vein, resulting in portal hypertension)
List 3 uses of a UAC.	1. Monitoring pulse and BP 2. Access for blood samples and ABG 3. Administering of fluids, medications, and hyperalimentation
Where should the tip of a UAC be placed?	Either just above the bifurcation of aorta (level of L3–5) or above the celiac axis (level of T6–10)
List 9 complications of a UAC.	Vascular embolization, thrombosis, vascular spasm, vascular perforation, ischemic or chemical necrosis of abdominal viscera, infection, hemorrhage, impaired circulation to the leg, and renovascular hypertension
How long may UVCs or UACs remain in place?	UVCs should be removed within 7–10 days and UACs within 7–14 days.

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EVALUATION OF THE NEWBORN

How are newborns classified?	By **gestational age** and **size**
List the 3 gestational ages and the span each includes.	**Preterm:** <37 weeks' gestation **Term:** 37 weeks to <42 weeks **Postterm:** 42 weeks and beyond
What is SGA?	**S**mall for **g**estational **a**ge; defined as less than the 5th or 10th percentile for gestational age (Fig. 8–1)
List 5 concerns about SGA.	Increased caloric needs (relative to their weight), a higher mortality rate, increased risk of malformations, hypoglycemia, and congenital infections
What is symmetric growth retardation?	Growth retardation of length, weight, and head circumference
List 3 causes of symmetric growth retardation.	1. An early insult or chronic condition in the pregnancy (e.g., teratogen, maternal disease) 2. A malformation syndrome (including chromosome abnormalities) 3. Early infection Alternatively, the parents may be small in stature.
Why may asymmetric growth retardation occur?	If weight is disproportionately low (relative to length and head circumference), it implies an insult later in pregnancy (e.g., maternal hypertension, placental insufficiency).
Do all cases of SGA have an identifiable cause?	No. As many as 40% of cases will have **no** identifiable etiologic factors.
What is LGA?	**L**arge for **g**estational **a**ge

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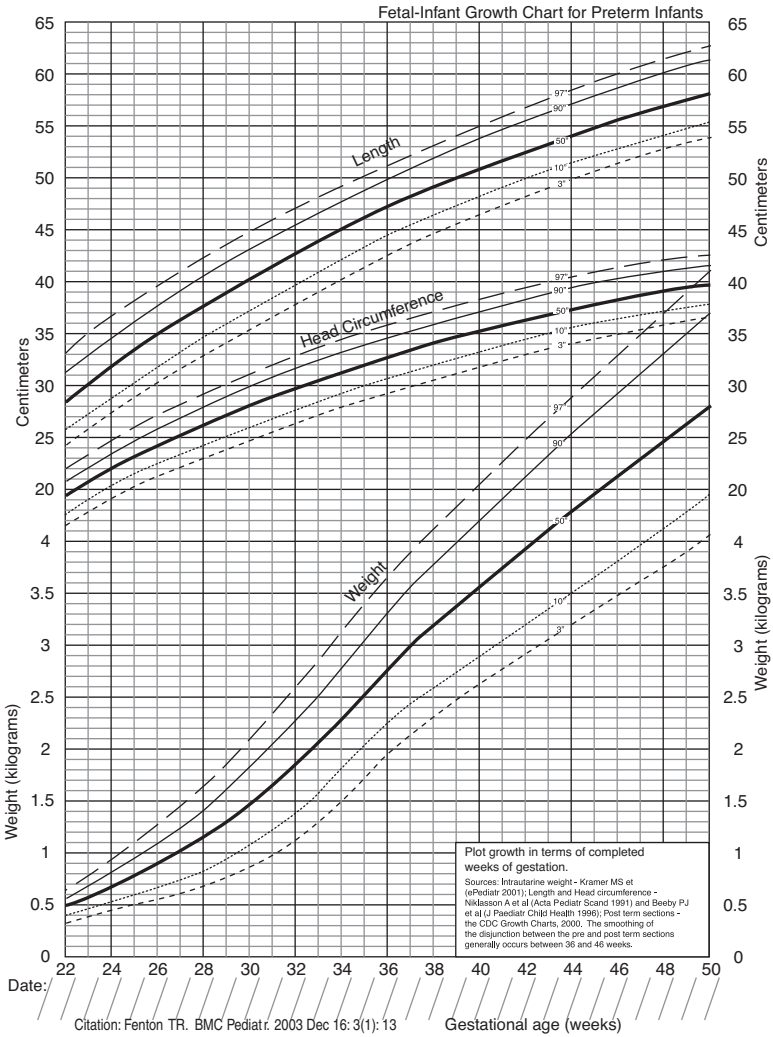

Figure 8–1. Fetal-infant growth chart for preterm infants. (From Fenton TR. BMC Pediatrics 2003;3:13. Used with permission of licensee BioMed Central Ltd.)

List 3 causes of LGA.

1. Maternal diabetes (including gestational diabetes)
2. Beckwith-Wiedemann syndrome
3. Twin-twin transfusion (recipient twin LGA)

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Do all cases of LGA have an identifiable cause?	No
List 3 complications of LGA.	1. Hypoglycemia 2. Increased incidence of injury at birth 3. Complications related to underlying cause
What is an IDM?	**I**nfant of a **d**iabetic **m**other
List 7 conditions that IDMs are at risk for.	Hypoglycemia, hypocalcemia, polycythemia, respiratory distress, renal vein thrombosis, small left colon syndrome, and malformations—particularly CHD and variants of caudal regression syndrome
What test assesses gestational age of the newborn?	New Ballard Score (based on the earlier Dubowitz examination), which uses physical and neurologic criteria correlated to gestational age
What is the significance of meconium staining?	It may reflect in utero stress and places the infant at risk for meconium aspiration (Ch 10, p. 96).
What is the normal newborn heart rate?	>100 bpm (usually 120–160 beats/min)
When should a newborn be examined?	In the delivery room. A complete physical examination should be done within 12 hours of birth.
List 4 purposes of the delivery room assessment.	1. Determine whether the infant will need resuscitation. 2. Assess for obvious malformations or abnormalities. 3. Estimate gestational age. 4. Assess the infant's cardiopulmonary transition from the intrauterine environment to the outside world.

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List clinical information important for the evaluation of the newborn.

1. Pregnancy history: gestational age, maternal disease, maternal medications, other complications
2. Labor and delivery history: type of delivery, time of rupture of membranes, medications during the labor, bleeding
3. Maternal laboratory data: blood type; antibody screen; screens for syphilis, hepatitis B, and HIV

List 3 purposes of the later, more complete examination.

1. Evaluate infant for malformations.
2. Establish normalcy of growth and function.
3. Document physical findings.

What 3 physical measurements are routinely done?

1. Head circumference (using greatest occipital-to-frontal diameter)
2. Weight
3. Crown-to-heel length

All measurements should be analyzed, using standard curves, for appropriateness for gestational age.

What is the normal newborn respiratory rate?

40–60 breaths/min

How should the complete physical examination of the newborn proceed?

The examiner should note the vital signs and general appearance and take advantage of the infant's current state. If the infant is sleeping or is quiet, auscultation of the heart and lungs and palpation and auscultation of the abdomen are done first, followed by a head-to-toe inspection and palpation. Unique aspects of the physical examination of a newborn include evaluation of patency of the nasal passages, palpation of the kidneys, and evaluation of the hips for dislocation or laxity. Ophthalmologic examination, including evaluation of the red reflex, is performed later in the examination. The formal part of the neurologic examination completes the physical examination.

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What is vernix caseosa?	A white greasy coating on the skin of newborns; more common in preterm infants
What is lanugo?	Fine hair that covers the body of infants; more common in premature infants
What are mongolian spots?	Bluish discoloration of the skin, usually over the buttocks and lower back; more common in racial groups with darker skin pigment
What are milia?	Tiny white papules that form over the surface of sebaceous glands; often present over the nose
What are miliaria?	Clear (crystallina) or inflamed-appearing (rubra) vesicles that form over obstructed sweat glands; often seen with overheating
What is erythema toxicum?	A benign, splotchy pattern of erythema and pustules filled with eosinophils. Typically appears on face, trunk, and extremities. **Must be distinguished from staphylococcal rashes or herpes.**
List 3 characteristics of staphylococcal rashes.	1. Pustules 2. Generalized erythema 3. Bullous eruptions (referred to as staphylococcal scalded skin syndrome, toxic epidermal necrosis, or Ritter disease)
What is a characteristic of herpes simplex rashes?	Vesiculobullous eruptions (may be only a few vesicles) on an erythematous base
What is a "blueberry muffin" rash?	Macular, raised, purple lesions—indicates congenital rubella (Ch 27, p. 443) or cytomegalovirus
Is palpable breast tissue normal in newborns?	Yes. About 1 cm of palpable breast tissue may be present in males or females because of maternal estrogens.

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Are heart murmurs common in newborns?

Soft, short systolic murmurs are common. Loud or harsh murmurs should arouse suspicion.

Name 5 heart defects associated with cyanosis.

The "5 Ts" (Ch 16):
1. Tetralogy of Fallot
2. Total anomalous pulmonary venous return
3. Truncus arteriosus
4. Transposition of the great vessels
5. Tricuspid atresia

What 3 conditions are associated with diminished or absent femoral pulses?

Coarctation of the aorta (Ch 16, p. 195), interrupted aortic arch, and hypoplastic left heart syndrome

How is the fontanel measured?

Anteroposterior and side-to-side dimensions (largest measurement of each)

List 4 disorders that are associated with an enlarged anterior fontanel.

Hydrocephalus, hypothyroidism, hypophosphatasia, and skeletal dysplasias (e.g., osteogenesis imperfecta)

List 3 kinds of disorders that are associated with a small fontanel.

Microcephaly, craniostenosis, and craniosynostosis syndromes

What is molding?

Temporary misshaping of the cranium, usually related to infant's position during the latter part of pregnancy and labor

What is a cephalohematoma?

A hematoma beneath the periosteum of the cranium

What is caput succedaneum?

Edema of the soft tissues of the scalp

How can cephalohematoma and caput succedaneum be differentiated?

Cephalohematomas do not cross suture lines.

What is ocular hypertelorism?

True ocular hypertelorism is an increased distance between the orbits.

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What is the red reflex?	The red reflection of the retina through the lens of the eye; a normal red reflex implies that there are no large lens opacities (e.g., cataracts), corneal clouding (e.g., congenital glaucoma), or large retinal tumors.
Why is a catheter passed through both sides of the nose?	To rule out choanal atresia or stenosis
Why not do this immediately after birth?	It may cause a vagal response with bradycardia.
Why is it important to have patent choanae?	Infants are obligate nose breathers, so choanal atresia can cause respiratory distress.
What are "Epstein pearls"?	Small, white, epithelial inclusion cysts seen in the midline of the roof of the mouth. Similar lesions may be seen in the gums. They resolve spontaneously.
What is the most common fracture during delivery?	Fracture of the clavicle
How large is the newborn liver?	The normal newborn liver may be palpable 1–2 cm below the right costal margin.
What is the normal number of vessels in the umbilical cord?	3 vessels (2 arteries and 1 vein)
When does the umbilical cord usually dry and fall off?	Usually before 3 weeks of age
What is acrocyanosis?	Bluish discoloration of the hands and feet; not rare in newborns but may be abnormal in older infants
What is harlequin color change?	Reddening of 1 side of the infant, with a sharp line of demarcation at the midline; may be related to autonomic factors; benign and self-limited

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What is cutis marmorata?	Reticulated mottling of the skin; may be seen transiently in infants who are cold; also seen as a more persistent finding in infants with Down syndrome or other specific disorders
What is the normal penis length for a term male infant?	About 3–4 cm shaft length
What is hypospadias?	Abnormal location of penile urethral meatus along the ventral aspect of the shaft
What is chordee?	Bowing or bending of the penile shaft
How do the labia minora vary with gestational age?	The labia minora are prominent in preterm females, usually protruding beyond the labia majora. They gradually become covered by the enlarging labia majora.
What is the significance of hair, swelling, or reddish discoloration in the lumbosacral area?	This is sometimes associated with spina bifida occulta, tethered cord.
What is the significance of a dimple at the base of the spine?	Dimples located **below the gluteal crease** are usually insignificant. Dimples **higher along the spine** require investigation for occult spina bifida or tethered cord.
What is syndactyly?	Cutaneous fusion of the digits
What is polydactyly?	Presence of extra digits
What is developmental dysplasia of the hip?	This term is sometimes used synonymously with congenital dysplasia of the hip; however, it is also a broader term that includes hip dislocation that may not be evident immediately at birth.

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List 2 ways to screen for it.

1. Observation for asymmetry of fat folds and discrepancy in leg length
2. Passively abducting the hips (i.e., Ortolani maneuver) or adducting and rotating each hip (i.e., Barlow test)

Which gender is affected more often?

Females

What if the neonate's feet or legs turn in?

Newborns may exhibit a variety of findings related to in utero positioning. If the foot or leg can be easily brought to a neutral or beyond-neutral position, the finding should resolve in several weeks. Otherwise, orthopaedic consultation may be required for forefoot, hindfoot, tibia, or hips.

List 4 key components of the neurologic examination of a newborn.

Mental status, cranial nerve function, motor function, and reflexes

How can mental status be examined?

Generally, awake infants are in a state of "quiet alertness." Abnormalities of mental status may manifest as unusual irritability or lethargy.

List 10 components of the examination of the cranial nerves.

Examine for:

1. Blinking when light is shone in eyes (II, VII)
2. Fixation on and tracking of objects (II, III, IV, VI)
3. Pupillary reaction (II, III)
4. Extraocular movements (III, IV, VI)
5. Corneal reflex and withdrawal on pinprick to face (V)
6. Facial movement and symmetry (VII)
7. Hearing (e.g., response to hand clap; VIII)
8. Sucking (V, VII, XII)
9. Swallowing (IX, X)
10. Strength and integrity of sternocleidomastoid (XI) and tongue (XII)

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What are the 2 things to look for when examining motor function?

1. Spontaneous and symmetric movement
2. Muscle tone, both active and passive (e.g., posture, resistance to passive motion)

What are primary (primitive) reflexes?

Inherent reflexes present during the first few months of life

Why are they important?

Absence of primary reflexes suggests CNS depression.

List 4 commonly tested primary (primitive) reflexes.

1. Moro reflex: Rapid abduction and extension of arms in response to the sense of falling (the examiner lifts the infant slightly off bed and releases)
2. Finger grasp: The infant flexes fingers to grasp when the examiner's index finger is placed in hand and slight traction is applied.
3. Automatic walking: The infant tries to support with feet when held upright; moves feet to "walk" when tilted forward.
4. Suck-swallow reflex: The infant sucks and swallows when the examiner's finger (clean) is placed in the infant's mouth.

Why give a newborn vitamin K?

Newborns may be deficient in vitamin K; the vitamin prevents hemorrhagic disease caused by deficiency of vitamin K–dependent coagulation factors.

Why put prophylactic antibiotics in the infant's eyes?

To prevent gonococcal eye infection (ophthalmia neonatorum)

What antibiotic is used?

0.5% erythromycin or 1% tetracycline ophthalmic ointments or 1% silver nitrate solution

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Is circumcision of males routine?

No. The indications for circumcision have traditionally been primarily cultural and religious. However, there are some studies suggesting medical benefits to circumcision including decreased risk of UTI in infancy and of HIV, other STDs, and penile cancer in adulthood. Parents should be appropriately informed about the pros and cons of the procedure.

What percentage of males are circumcised?

60–70%. Varies dramatically, based on cultural factors

Should local anesthesia be used for circumcision?

Yes (usually 1% lidocaine) as a dorsal penile or ring block. Topical EMLA is also effective.

Is nonretractile foreskin a cause for concern?

Foreskin typically does not retract completely in the newborn, but this condition usually improves with age.

List 6 contraindications for circumcision.

Hypospadias (Ch 22, p. 344), chordee, micropenis, exposure to herpes simplex during labor or delivery, ambiguous genitalia (Ch 24, p. 392), and bleeding disorder or family history of a bleeding disorder (a contraindication until the child has been tested and either is found to be unaffected or is successfully treated; Ch 15, p. 182)

When can feedings begin for a newborn?

Breast: Breast-feed immediately after delivery and every 3–4 hours or on demand.

Formula by bottle: Begin when respiratory status is stable and every 4 hours or on demand.

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What laboratory studies should typically be obtained in the newborn?

1. Capillary Hct and bilirubin at 4 hours when Rh sensitization is known or newborn is pale in color, twin gestation, IDM, or SGA. Investigate if Hct < 40% or >70% and bilirubin > 4 mg/dL.
2. Glucose screen within 1 hour and prn if weight < 2,500 g or >90th percentile for gestational age (LGA); or if newborn is jittery or has respiratory distress, unstable temperature, or apnea. Investigate if value < 50 mg/dL.
3. At discharge, state-required "newborn screening tests." (These vary from state to state and may include tests for hypothyroidism, tyrosinemia, phenylketonuria, galactosemia, maple syrup urine disease, homocystinuria, biotinidase deficiency, adrenal hyperplasia, hemoglobinopathies, and CF, among others.)
4. Hearing screen before discharge
5. Total bilirubin determination, using either serum bilirubin or noninvasive photometric method. Timed result can be used to determine the risk for significant hyperbilirubinemia and need for follow-up after discharge.

Is a car seat required for all infants for discharge home?

Yes, infants born before 37 weeks of gestation should be monitored in their car seats for apnea, bradycardia, or desaturation for a minimum of 1.5 hours or however long the ride home is.

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Chapter 9 Common Clinical Problems

JAUNDICE

What is it?	Accumulation of bilirubin in the epidermal tissues of the body, resulting in a yellowish tinge to the skin, sclera, and mucosa
At what bilirubin level is jaundice usually evident in the newborn?	Serum levels > 5.0 mg/dL
What type of bilirubin is most commonly elevated in the neonate?	Unconjugated bilirubin
What is the physiology of elevated bilirubin?	**Unconjugated** (indirect) hyperbilirubinemia is secondary to increased production of bilirubin (e.g., excess RBC destruction), decreased hepatic conjugation of bilirubin, increased absorption of bilirubin from the intestine (enterohepatic circulation), or decreased hepatic uptake of bilirubin. **Conjugated** (direct) hyperbilirubinemia is caused by hepatobiliary dysfunction or extrahepatic biliary obstruction.
What are the complications of hyperbilirubinemia?	Persistent and pathologic elevation of bilirubin in the newborn may cause an excess of free bilirubin (unconjugated bilirubin not bound to albumin or other serum proteins). This potential neuro-toxin may cause **kernicterus,** an often-irreversible phenomenon characterized by alteration of neurobehavioral status and injury to the brain. Long-term sequelae of kernicterus may include deafness, cerebral palsy, or death.

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What level of bilirubin is excessive in neonatal jaundice?

Controversial. In healthy term infants, an unconjugated bilirubin concentration > 20 mg/dL is potentially a cause for concern. Prematurity, hemolysis, acidosis, and other conditions may lower the threshold at which hyperbilirubinemia may cause damage.

What is the differential diagnosis?

Unconjugated hyperbilirubinemia may be physiologic (i.e., caused by immature hepatic enzyme pathways in the newborn) or may be associated with breast-feeding. More pathologic causes include:

1. Rh, ABO, or other RBC isoimmunization conditions
2. RBC membrane defects (e.g., congenital spherocytosis)
3. RBC biochemical defects (e.g., G6PD deficiency)
4. Deficiency in glucuronyl transferase (e.g., Crigler-Najjar syndrome)
5. Hypothyroidism
6. Bruising
7. Bacterial or viral sepsis
8. Resolving cephalohematoma or caput succedaneum with subgaleal hemorrhage

Conjugated hyperbilirubinemia may be caused by direct hepatic insult from asphyxia, sepsis, or congenital metabolic toxins, cholestasis from hyperalimentation, or intrahepatic or extrahepatic biliary obstruction.

List 5 important components of the clinical evaluation.

1. Pertinent history should identify maternal complications with pregnancy or delivery; maternal and neonatal blood types, and direct Coombs' results; feeding history; time of onset and duration of jaundice.
2. Clinical examination should be thorough and should include a neurobehavioral examination and evaluation for signs of sepsis.

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3. Pertinent studies include fractionated bilirubin level (direct and indirect bilirubin levels), Hct, and evaluation of a blood smear for evidence of hemolysis; reticulocyte count and G6PD assay may be helpful. Evaluation of liver function and hepatocellular integrity is necessary with conjugated hyperbilirubinemia.
4. With conjugated hyperbilirubinemia, metabolic and infectious evaluation is necessary. Ultrasound of the liver and biliary tree or nuclear medicine excretion studies may be needed to rule out anatomic abnormalities (e.g., biliary atresia).
5. Liver biopsy and cholangiogram may be needed in certain cases.

What is the treatment?

Unconjugated hyperbilirubinemia: Prophylactic or therapeutic phototherapy. IVIg may be useful for infants with Rh or ABO incompatibility and hemolysis. Infants at high risk of kernicterus may need exchange transfusions of whole blood.

Conjugated hyperbilirubinemia: Treat the underlying liver disease or other disease process.

NEONATAL SEIZURES (SEE CH 6, P. 43 AND CH 25, P. 396)

What is the incidence of seizures in neonates?

0.8–1% of all live births; more common in preterm infants

What do seizures reflect?

Seizures may indicate underlying illness or metabolic derangement. All seizures require prompt evaluation!

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What are the common manifestations of seizures in a neonate?

Seizure activity in the newborn may be focal, clonic, multifocal clonic, tonic, or myoclonic movements.

Subtle seizures may manifest with sudden onset of apnea; intermittent vasomotor phenomena; oromotor, ocular, or facial tics; or repetitive motion of facial muscle groups.

List 2 complications of seizures.

1. Respiratory compromise or apnea
2. Prolonged seizure activity may cause permanent brain injury.

What is the most common cause of seizures?

Hypoxic-ischemic encephalopathy, which is caused by intrauterine or birth-related insult

List 7 other common causes of seizures.

1. Infection: bacterial or viral infection or toxoplasmosis
2. Imbalances in the blood concentrations of glucose, Na^+, Ca^{2+}, and Mg^{2+}
3. ICH
4. CNS malformation
5. Drug withdrawal
6. Inborn errors of metabolism
7. Benign familial neonatal seizures

List 4 important factors in the history when evaluating a seizure.

1. Complications during pregnancy or delivery
2. Maternal drug use
3. Family history of seizures
4. Risk factors for sepsis

What 4 diagnostic studies should be performed?

1. Blood chemistries (e.g., electrolytes, calcium, magnesium, glucose, ABG)
2. Evaluation for sepsis, including LP
3. CT or MRI to evaluate for hemorrhage, ischemia, or CNS malformation
4. EEG for detecting subtle seizures

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What do skin vesicles or mucosal lesions suggest?

Herpes simplex encephalitis may present after the first week of age with seizures, lethargy, irritability, or a sepsis-like syndrome.

What is the treatment for neonatal seizures?

Treat the underlying cause.
Also:
1. Attention to airway and respiratory support
2. Electrolyte replacement as needed
3. Antibiotics and acyclovir therapy as clinically indicated
4. Anticonvulsant therapy (e.g., phenobarbital and phenytoin) titrated for effect
5. EEG monitoring for detection of subclinical seizures

What is the prognosis?

The prognosis depends on the etiologic factors.

List 3 signs of poor prognosis.

1. Seizures caused by hypoxic-ischemic encephalopathy or CNS malformation
2. Seizures initially occurring <12 hours after birth
3. Refractory seizures persisting beyond 24 hours after birth, requiring multiple drugs or high serum drug levels for control

What percentage of infants with seizures will have neurodevelopmental problems?

15–20%; the risk depends on etiology.

NEONATAL ANURIA AND OLIGURIA

What is neonatal oliguria?

Urine output of <15–20 mL/kg per day in the first 24–48 hours after birth. Many infants may only void once in the first 24 hours after birth (see Ch 21, p. 328).

What is neonatal anuria?

Failure to void within the first 48 hours after birth

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What percentage of newborns void within 24 hours?

92%. Voiding in the delivery room "counts."

Within 48 hours?

99%

What is the normal urine output for an infant after 24–48 hours?

~2 mL/kg per hour

What are the causes of oliguria?

Prerenal in origin until proven otherwise! Oliguria is usually secondary to hemodynamic compromise caused by sepsis, CHD, dehydration, hypoxia, or (rarely) renovascular accident.

What are the 2 causes of oliguria as a result of urinary retention?

1. Renal malformation
2. Obstruction of urinary outflow tract (e.g., posterior urethral valves)

List 6 causes of intrauterine anuria.

Bilateral renal agenesis, severe congenital polycystic kidney disease, posterior urethral valves, maternal or neonatal drug exposure, fetal cardiovascular compromise (e.g., placental insufficiency, severe intrauterine growth restriction)

List 2 complications of intrauterine anuria.

Oligohydramnios, pulmonary hypoplasia

What is the incidence of renal malformations, and what is the most common one?

5–6 per 1,000 live births; horseshoe kidney is most common.

List 4 primary renal causes of anuria.

Bilateral renal agenesis, multicystic renal disease, congenital polycystic kidney disease, exposure to nephrotoxins

List 4 important obstructive lesions.

1. Ureteropelvic junction obstruction
2. Posterior urethral valves
3. Duplication of the calyces, renal pelvis, or ureters
4. Urethral atresia

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Why is family history important?

Some congenital renal disorders are inherited.

What are the important aspects of prenatal and intrapartum history?

1. Maternal history of oligohydramnios
2. History consistent with a hypoxic-ischemic event

Many urinary anomalies are now diagnosed antenatally by ultrasound.

List 2 ways of evaluating prerenal compromise in the neonate.

1. Fluid bolus challenge of 10–20 mL/kg
2. Single-dose furosemide therapy (1 mg/kg) after a bolus dose of isotonic fluids may help rule out more severe disease.

List 2 important components of the clinical evaluation of oliguria or anuria.

1. Evaluation of perfusion and BP
2. Abdominal examination with careful palpation of kidneys; inability to palpate may suggest agenesis or absence of kidney; palpation of mass may suggest polycystic kidney or hydronephrosis as a result of urinary obstruction.

List 2 important laboratory values in evaluating a neonate with oliguria or anuria.

1. Blood chemistries including BUN and creatinine. (IMPORTANT: Neonatal electrolytes and chemistry values typically reflect the mother's values in the first 24 hours of the newborn's life.)
2. Urinalysis

What are the 2 most helpful imaging studies?

1. Abdominal ultrasound—The kidneys and bladder should be evaluated in any infant with multiple congenital anomalies.
2. Nuclear medicine excretion studies may help assess the relative function of each kidney.

List 3 treatments that may be included in the management of neonatal oliguria or anuria.

1. Judicious use of IV fluids to maintain adequate urine output
2. Maintenance of normal BP; renovascular accidents, renal dysplasia, and postobstructive disorders are associated with severe hypertension
3. Peritoneal or arteriovenous dialysis if renal failure ensues

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What is the outcome?

Varies, depending on the cause. Prompt diagnosis and treatment of oliguria and anuria prevent continued renal deterioration.

FAILURE TO PASS MECONIUM

When is meconium normally passed?

95–97% of healthy term newborns pass meconium in the first 24 hours. Sick term infants and healthy preterm infants may not pass meconium for 3–5 days.

List 3 signs associated with failure to pass meconium.

Abdominal distension, feeding intolerance, emesis. Any bilious emesis requires prompt evaluation and should be considered volvulus until proven otherwise.

What is the differential diagnosis?

1. Hirschsprung disease (Ch 19, p. 293)
2. Meconium ileus
3. Meconium plug
4. Anorectal malformation or atresia (i.e., imperforate anus; Ch 19, p. 275)
5. Malrotation of the bowel (Ch 19, p. 294)
6. Small left colon syndrome in the infant of a diabetic mother
7. Maternal medication, especially magnesium sulfate
8. Atresia of the duodenum, jejunum, ileum, or colon (Ch 19, p. 296)
9. Intestinal duplication

In any of these conditions except meconium ileus or imperforate anus, meconium may still pass appropriately!

List 4 important aspects of the clinical evaluation.

1. Confirm patency of rectum with digital examination or passage of soft catheter.
2. Water-soluble contrast study of lower and upper gastrointestinal tract to determine whether obstruction is present
3. Rectal biopsy if Hirschsprung disease is suspected
4. Assessment for CF

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List 2 possibilities for treatment.

1. Water-soluble contrast enema may alleviate meconium plug, meconium ileus, or small left colon syndrome. (Examiner must consider Hirschsprung disease or CF in infants with meconium plug or meconium ileus.)
2. Surgical exploration and repair may be necessary when an obstructing disorder exists. Preoperative abdominal decompression and maintenance of fluid and electrolyte homeostasis and hemodynamic status are imperative in these cases.

NEONATAL CYANOSIS

What is it?

Bluish tint of the skin: reflects the presence of 3–5 g/dL of reduced Hgb in the blood

What must the O_2 saturation be for an infant with polycythemia to demonstrate cyanosis?

<88%

An anemic infant?

<70%

List 5 common causes of cyanosis.

Primary lung disease, poor cardiac output, congenital cyanotic heart disease, pulmonary hypertension in the newborn, methemoglobinemia

Many healthy newborn infants may have cyanosis of the extremities (acrocyanosis) with a normal pink color centrally, as a result of cooling, immature regulation of skin blood flow, or both.

Why are prenatal and perinatal histories important?

Intrauterine or birth-related complications may indicate sepsis, asphyxia, or pulmonary insult as causes of cyanosis.

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List 3 significant findings on clinical examination.

Heart murmurs, absent distal pulses, respiratory distress

Why is a chest radiograph important? (List 2 reasons.)

1. Evaluation of the lung fields for evidence of primary lung disease
2. Evaluation of cardiac size and shape for evidence of congenital heart disease

What are the 2 significant findings for ABG?

1. PaO_2 level > 60 mm Hg on room air virtually excludes cyanotic heart disease.
2. Failure to demonstrate a rise of PaO_2 >100 mm Hg in response to oxygen suggests a cardiac rather than pulmonary etiologic factor.

What is the significance of chocolate-colored blood?

It may indicate methemoglobinemia.

What condition may cause a 5–10% difference in O_2 saturation between the arms and legs?

Pulmonary hypertension

Why?

Pulmonary hypertension may cause right-to-left shunting through a PDA. Blood supply to the right arm is preductal and to the legs is postductal.

What is the treatment for cyanosis?

Treat the underlying disease. Oxygen and ventilatory support may be required for primary lung disease. Antibiotics are important in fighting infection. Nitric oxide or ECMO therapy, or both, may be needed for refractory pulmonary disease, sepsis, and persistent pulmonary hypertension. Prostaglandin E_1 infusion may improve oxygenation in the infant with cyanotic heart disease or perfusion in the infant with left-sided obstructive lesions.

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NEONATAL RESPIRATORY DISTRESS

What is the most common reason for admitting a newborn to a level II or III neonatal intensive care unit?	Respiratory distress
List 4 features of respiratory distress.	Tachypnea (i.e., >60 breaths/min in any infant); use of accessory muscles of respiration (e.g., nasal flaring, intercostal retractions, grunting); hypoxia; hypercapnia
What is the most common cause of neonatal respiratory distress?	Primary lung disease
List 3 types of restrictive (poor lung compliance) conditions.	Pneumonia, surfactant deficiency (RDS; Ch 10, p. 94), malformation of the lung or chest wall
Give an example of a type of obstructive (normal compliance) condition.	Aspiration syndromes—for example, aspiration of blood, amniotic fluid, meconium (Ch 10, p. 96), or gastric contents
List 12 other common causes of respiratory distress.	CNS injury; obstruction of upper airway by nasopharyngeal tissues (as in choanal stenosis or atresia) or the tongue (as in severe micrognathia or macroglossia); primary malformations of lung tissue; pulmonary edema; retained fetal lung fluid after precipitous vaginal delivery or cesarean section; pleural effusion; severe abdominal distension; diaphragmatic hernia; hypoplastic lungs (caused by renal dysfunction or other causes of oligohydramnios); infection; polycythemia; TTN (Ch 10, p. 96)
List 6 important factors in evaluation.	Clinical history and examination, chest radiograph, ABG, Hct, assessment for cardiac disease, evaluation for sepsis

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NEONATAL HYPOTONIA

What is the normal resting position for a newborn?

Elbows and knees flexed; hands most often in the fisted position

For a premature newborn?

Flexed and fisted less frequently

List 5 characteristics of hypotonia.

1. Extension of extremities
2. Open hands
3. Occasionally exaggerated "frog-leg" position when supine
4. Child not withdrawing into flexion with noxious stimuli
5. Diminished primitive reflexes because of poor truncal tone

What causes hypotonia?

Conditions that are **genetic** or **acquired** in the intrauterine environment or during the birth process. May be systemic or primarily neuromuscular. Neuromuscular conditions may be at any level: cerebral cortex to peripheral nerve to neuromuscular junction to muscle.

List 4 causes of hypotonia without a primary neuromuscular etiology.

Maternal disease or medication; sepsis; severe cardiorespiratory disease; hypoglycemia

List 6 causes of hypotonia due to CNS or neuromuscular pathology.

1. Hypoxic ischemic encephalopathy
2. Abnormal cortical development: for example, Prader-Willi, Trisomy 21
3. Spinal cord injury
4. Anterior horn cell degeneration (e.g., infantile spinal muscular atrophy)
5. Variants of congenital myasthenia gravis
6. Myotonic dystrophy and other muscular dystrophies

Why is family history important?

Because of possible genetic etiologic factors. In addition, infants with congenital myotonic dystrophy may show no clinical or electrical signs of myotonia; however, often the mother will.

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List 5 important laboratory studies.	Blood chemistries, sepsis evaluation, acid-base status, plasma ammonia concentration, CPK levels
What imaging studies are helpful?	Cranial and spinal MRI or CT
Why are the electroencephalogram and electromyogram important?	They can help rule out seizures and abnormalities of skeletal muscle innervation.
What is another important test?	Muscle biopsy
What consultants are commonly needed?	Neurologists and genetic consultants provide workups of specific congenital metabolic disorders.
What is the prognosis?	The outcome depends on the specific disease present; genetic counseling may be helpful in certain cases.

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Chapter 10 Diseases of the Newborn

PERIVENTRICULAR OR INTRAVENTRICULAR HEMORRHAGE

What is it?

Intracranial bleeding that most commonly arises from the capillary network of the subependymal germinal matrix layer; arises less frequently from the choroid plexus or the roof of the fourth ventricle (Ch 25, p. 399)

List the 4 grades (classifications) of these hemorrhages.

Grade I: isolated germinal matrix hemorrhage

Grade II: IVH with normal ventricular size

Grade III: IVH with acute ventricular dilatation

Grade IV: IVH with associated parenchymal hemorrhagic infarct

What is the incidence?

Approximately 30% of infants who weigh <1,500 g and are <35 weeks' gestation have some degree of hemorrhage. Most hemorrhages occur within the first week of life, usually within the first 2 days. Although essentially a condition of premature neonates, it is seen occasionally in full-term infants.

What is the physiology of this condition?

The germinal matrix is a periventricular structure, containing a rich vascular network of primitive vessels most prominent at 26–34 weeks from conception. Bleeding from the matrix ruptures into the lateral ventricles.

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List 9 predisposing conditions and events.

Most common: Prematurity, acute respiratory failure requiring mechanical ventilation

Others: Pneumothorax, hypotension, acidosis, coagulopathy, volume expansion, bicarbonate infusion, birth trauma

List 4 signs. Hypotension, apnea, metabolic acidosis, bulging of the anterior fontanel in severe cases

At least 50% of infants with hemorrhage have no clinical symptoms.

List 3 methods that are used for diagnosis. Ultrasound (usually preferred), MRI, or CT scans

What is the treatment? Supportive care. Anticonvulsants may be required for seizures or seizure prophylaxis. If **posthemorrhagic hydrocephalus** ensues, treatment is controversial but options include:

1. Serial LPs
2. Drugs (e.g., acetazolamide) that decrease the production of CSF
3. Ventricular reservoir with serial tapping or ventriculostomy
4. Placement of ventriculoperitoneal shunt

What is the prognosis? Grade 1 and grade 2 hemorrhage do not increase the risk for developmental delay or cerebral palsy. Grade 3, especially if there is progressive hydrocephalus, and grade 4 hemorrhage are associated with increased risk.

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RESPIRATORY DISTRESS SYNDROME

What is it?	Pulmonary disease associated with prematurity and surfactant deficiency; previously called "hyaline membrane disease"
What is the incidence?	Age dependent: 60% at 29 weeks' gestation. Decreases to <1% by 39 weeks' gestation
List 5 circumstances that increase the risk of RDS.	1. Infants of diabetic mothers (IDMs) 2. Infants who have siblings who had RDS 3. Males 4. Infants born by cesarean section without labor 5. Infants who experience perinatal asphyxia **Note:** Infants who had prolonged ROM or IUGR, or whose mothers experienced physiologic stress, are relatively spared.
What are the classic features?	Onset of grunting respirations, chest retractions, and increased oxygen requirements. Characteristic radiographic changes (reticular, granular pattern with air bronchograms) occur within 6 hours of the onset of symptoms.
List 4 acute and 3 long-term complications.	
Acute:	Alveolar rupture (leading to pneumothorax, pneumomediastinum, pneumopericardium, or interstitial emphysema), infections, intracranial hemorrhage, PDA
Long-term:	Chronic lung disease (CLD; also called bronchopulmonary dysplasia [BPD]), retinopathy of prematurity, possible neurodevelopmental impairment

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How is pulmonary immaturity detected prenatally?

Because fetal lung fluid enters the amniotic cavity, **amniocentesis** provides a means for assessing pulmonary maturity via analysis of phospholipids in the amniotic fluid. Lecithin:sphingomyelin (L:S) ratio of <2:1, absence of phosphatidylglycerol, and a saturated phosphatidylcholine (SPD) concentration of <500 are associated with insufficient surfactant and, therefore, with potential RDS.

List 6 components of treatment.

Stabilization of the premature infant, including:

1. Administration of oxygen to keep PaO_2 at 50–80 mm Hg
2. Early surfactant with additional dosing (up to 3–4 doses) as required
3. Lung expansion, using continuous positive airway pressure or intubation and mechanical ventilation with PEEP
4. Thermal neutrality, usually a skin temperature of 36.5°C
5. Cardiovascular support
6. Acid-base and electrolyte therapy as indicated

What is CLD?

A condition characterized by inflammation or scarring of immature lung tissue exposed to high-pressure ventilation, oxygen, or infection

List 3 preventive measures against CLD.

Administration of prenatal glucocorticoids to mother; surfactant therapy to infant with immature lungs at birth; minimization of ventilator injury from pressure and oxygen

List 5 components of treatment for CLD.

Treatment is mainly supportive. Therapies include judicious fluid and diuretic management, prevention of infection, bronchodilators, and nutrition. Glucocorticoids may improve the pulmonary status, but their use is controversial because of reports of increased risk of neurologic impairment.

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What is the long-term outcome?

Some children may experience chronic respiratory insufficiency, but most outgrow their CLD and have the potential for normal lives from a respiratory standpoint.

TRANSIENT TACHYPNEA OF THE NEWBORN

What is it?

Early onset of mild respiratory distress

What causes TTN?

Delivery before the normal physiologic decrease in pulmonary fluid production or any circumstance delaying the clearance of lung liquid by the lymphatics, including elevation of central venous pressure by late clamping of the cord. Often associated with cesarean section with limited or no labor

List 7 features of the classic clinical presentation.

Tachypneic infant with 60–120 shallow respirations per minute, mild cyanosis, mild grunting, nasal flaring, chest retractions, mild respiratory acidosis, and mild to moderate hypoxemia

List 4 radiographic findings.

Hyperaeration of the lungs, fluid in the interlobar fissures, prominent pulmonary vascular markings, and mild cardiomegaly. These signs usually resolve within 24–48 hours.

What is the treatment?

Supportive care with supplemental oxygen. Diuretics are not helpful. TTN is self-limited and often resolves within 72 hours.

MECONIUM ASPIRATION SYNDROME

What is meconium?

A thick, blackish green material that begins to accumulate in the fetal intestines by the end of the first trimester. It is the accumulation of debris from the developing gastrointestinal tract.

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What 3 conditions can be caused by meconium aspiration?

Significant **pneumonia, pneumonitis, pneumothorax**

What conditions are commonly associated with meconium aspiration syndrome?

Pulmonary hypertension, hypoxic-ischemic systemic injury caused by antenatal, intrapartum, and neonatal stress

What 2 groups of infants are particularly at risk?

SGA infants and post-term infants. (Meconium-stained fluid is seen in 8–20% of all deliveries.) Meconium staining rarely occurs before 34 weeks gestation.

How can this syndrome be prevented?

Maternal management: Amnioinfusion of normal saline to relieve cord compression if fetal distress is evident

Infant management: Immediate endotracheal intubation and suctioning before the initiation of spontaneous respirations if the infant is depressed or has respiratory distress. The infant may have aspirated the meconium-stained fluid deep into the lung before birth due to fetal gasping.

What are the key components of treatment?

1. If respiratory distress develops, supportive care, especially directed at oxygenation, is established. Infants may require intubation, mechanical ventilation, and sedation. Surfactant therapy may be beneficial because of the inactivation of the native surfactant by meconium. Inhaled nitric oxide or ECMO may be needed in most severe cases.
2. Antibiotics are given after bacterial cultures are obtained, because meconium may promote bacterial growth, and sepsis may have contributed to the initial passage of meconium.

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What are the outcomes?

Historically, meconium aspiration syndrome has had a high mortality rate—up to 30% of infants who require mechanical ventilation. However, with improved ventilation techniques, nitric oxide, and ECMO, outcomes have improved. ECMO saves 85–95% of infants who would probably otherwise die. CLD may result in a few cases. Outcome is complicated by the comorbidities of pulmonary hypertension and systemic hypoxic-ischemic insults.

PERSISTENT PULMONARY HYPERTENSION OF THE NEWBORN

What is it?

PPHN, or persistent fetal circulation, implies pulmonary hypertension, right-to-left shunting at the level of the PDA and patent foramen ovale (PFO), and a structurally normal heart, often with depressed myocardial function and systemic hypotension. The constellation causes **severe hypoxemia.**

List 8 etiologic factors.

Primary (idiopathic) or secondary to: meconium aspiration or other aspiration syndromes; hyperviscosity of blood; neonatal sepsis (see the following text); intrauterine or perinatal asphyxia; myocardial dysfunction; CDH; neonatal pulmonary disease

List 5 symptoms.

Respiratory distress; cyanosis; blowing murmur of tricuspid regurgitation; shock (Ch 6, p. 35); heart failure

What does the chest radiograph show?

May reveal underlying lung disease; may show diminished pulmonary markings; or may be normal

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Differential diagnosis?

1. Cyanotic CHD (Ch 16), including transposition of the great vessels, total anomalous pulmonary venous return, pulmonic stenosis or atresia, and Ebstein anomaly
2. Severe pulmonary disease including CDH, pulmonary hypoplasia, and pneumonia

What is the treatment?

Goal is to decrease pulmonary vascular resistance and right-to-left shunting.

1. Supportive management is directed at correcting acidosis and hypoxemia. Liberal use of oxygen is recommended.
2. If conservative therapy fails, intubation, mechanical ventilation, and sedation are initiated. Mild alkalosis (either metabolic or respiratory) aids in pulmonary vasodilation. Some clinicians hyperventilate the infant to Pco_2 30 mm Hg and pH 7.45–7.55. Severe hyperventilation should be avoided because of its effect on cerebral blood flow and risk of barotrauma/volume trauma to the lungs.
3. Inhaled nitric oxide is an effective pulmonary vasodilator in some infants. Inhaled or intravenous prostacyclin (PGI) and the cGMP phosphodiesterase inhibitor, sildenafil, may also be effective.
4. Pressors and inotropic agents may be required for systemic hypotension.
5. ECMO may be indicated if conventional therapy fails.

What are the outcomes?

Mortality rate ranges from 20% to 40%. The incidence of neurologic sequelae is 12–25% for survivors. Neurosensory hearing loss has been reported in up to 20% of survivors.

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NEONATAL PNEUMONIA

What is the incidence?	Up to 0.5% of live births
What are the etiologic factors?	Bacterial or viral; may be acquired transplacentally, through the birth canal, or after delivery
What is the most common bacterial agent?	Group B *streptococcus*, affecting 1–4 in 1,000 live births
List 8 other common bacterial agents.	*Escherichia coli* (*E. coli*), *Klebsiella* species, Group D *streptococcus*, *Listeria* species, and Pneumococci. *Staphylococcus* and *Pseudomonas* species can cause slightly later-onset disease, and *Chlamydia trachomatis* can cause pneumonia as late as 3–4 weeks after birth.
What are the common viral agents?	Prenatally or postnatally acquired CMV, as well as postnatally acquired influenza, RSV, and adenovirus. The latter are associated with significant morbidity and mortality rates in infants.
List 4 predisposing factors.	Premature labor; rupture of membranes before the onset of labor, or prolonged rupture of membranes; prolonged active labor with cervical dilatation
List 8 signs and symptoms of neonatal pneumonia.	**Nonpulmonary symptoms:** Lethargy, thermal instability, apnea, abdominal distension, jaundice **Pulmonary symptoms:** Tachypnea, cyanosis, respiratory distress
What are the lung field findings on the chest radiograph?	Vary from streaky density, to diffuse opacification, to a granular appearance. May be confused with RDS or TTN
List 4 important studies to be conducted during evaluation.	Blood and CSF cultures; complete blood count with differential; nasopharyngeal culture for chlamydia or viruses for the older infant; tracheal aspirate for Gram stain and culture if intubated

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What is the treatment?

Initial treatment includes a penicillin (usually ampicillin) and an aminoglycoside because broad-spectrum coverage is indicated for early-onset condition. Treatment for later-onset pneumonia should also include coverage for *staphylococcus* organisms. When the causative organism is identified, treatment may be narrowed and continued for a minimum of 10 days.

NEONATAL SEPSIS

What is it?

A generalized bacterial infection in a clinically ill infant with a positive blood culture during the first month of life

What is the incidence?

It occurs in 1 in 500 to 1 in 600 live births and is influenced by maternal factors, including active infection at delivery and neonatal (especially prematurity) and environmental factors.

When can infection occur?

1. Before labor, it can occur transplacentally or through the amniotic fluid with or without intact membranes.
2. During delivery, as the infant passes through the birth canal
3. After delivery

List the 7 most common bacterial agents.

Group B *streptococcus*; *E. coli* and other gram-negative rods; *Listeria monocytogenes*; group A *streptococcus*; *Enterococcus*; *Streptococcus viridans*; *Staphylococcus* species. Viral agents, especially herpes simplex, may present in this manner.

List some potential symptoms of neonatal sepsis.

Lethargy, irritability, poor feeding, temperature instability, possible fever if fulminant, tachypnea, hypotension, cyanosis, apnea, tachycardia, seizures, vomiting, diarrhea, hepatomegaly, jaundice, petechiae, and bleeding

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Differential diagnosis?

Differential diagnosis includes heart disease, inborn errors of metabolism, hypovolemic shock, intracranial hemorrhage, RDS, pneumonia, gastrointestinal anomalies, or hypoglycemia.

Which blood test is a helpful indicator for sepsis?

WBC with differential. Best indicator is an abnormal ratio of bands to neutrophils (i.e., >20%) on differential. (Normal WBC for an infant is 8,000–20,000 WBC/mm^3.) Platelet count is often low.

List 3 possible components of treatment.

1. After cultures (blood and CSF) have been obtained, broad-spectrum coverage is initiated with ampicillin and an aminoglycoside, usually gentamicin. When an organism has been identified, coverage is narrowed based on sensitivities and a 7- to 10-day course can be completed. LP should be performed when infant is stable (see meningitis in the following text).
2. Granulocyte colony-stimulating factor may be required for desperately ill newborns with neutropenia.
3. ECMO may be used for infants exhibiting pulmonary failure secondary to neonatal sepsis.

What is the outcome?

Mortality rate can be as high as 13–50%.

NEONATAL BACTERIAL MENINGITIS

(See also Ch 28 for discussion of meningitis, including bacterial meningitis.)

What is the incidence?

Approximately 2–10 of 10,000 live births and is responsible for 1–4 of 100 neonatal deaths

List 3 of the most common infecting agents.

Overall, the same as those discussed in neonatal sepsis (see the preceding text), but most common are group B *streptococcus*, *E. coli*, and *L. monocytogenes*

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What are the risk factors?

They are the same as those associated with neonatal sepsis (see the preceding text). Meningitis is associated with up to 33% of cases. Local infections of the skin, respiratory tract, and urinary tract can cause bacteremia and thus meningitis. Premature infants and infants with meningomyelocele are at increased risk. Certain strains of bacteria are associated with increased risk—specifically, *E. coli* containing capsular polysaccharide K1, group B *streptococcus* serotype III, and *L. monocytogenes* type IV.

What are the signs and symptoms?

Same as those for sepsis (see the preceding text). In addition:

Seizures, lethargy or irritability, abnormal cry, focal neurologic signs, bulging fontanel (may be a late sign)

Stiff neck and positive Kernig or Brudzinski signs are rarely seen in this age group.

List 2 important factors in evaluation.

1. Every infant with subtle signs of sepsis requires an **LP** with Gram stain, cell count, protein and glucose analysis, and culture of the CSF.
2. Blood and urine cultures should also be obtained.

What is the treatment?

Broad-spectrum antibiotic therapy (ampicillin and an aminoglycoside) is initiated and tailored to an identified organism and sensitivity. **Gram-positive** meningitis is treated for a minimum of 14 days, whereas **gram-negative** meningitis is treated for 2 weeks after the infection is cleared or 3 weeks minimum, whichever is longer.

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What are the outcomes?

Mortality rate ranges from 20% to 50%. Morbidity is substantial and includes hydrocephalus, subdural effusions, ventriculitis, deafness, and blindness. Neurologic impairment is evident in 40–50% of survivors. All survivors require audiologic and neurologic follow-up.

APNEA OF INFANCY AND SIDS

What is apnea of infancy?

Pause in breathing, usually ranging from 5 to >20 seconds in duration, which arises from a central event, an obstructive event, or a combination of both.

What is the normal physiology of infantile breathing?

While the respiratory system matures, an infant <6 months of age may experience periodic breathing or isolated, asymptomatic apnea of 5–15 seconds in duration.

When are apneic periods of clinical importance?

When unexplained apnea lasts >20 seconds or is symptomatic

What is an ALTE?

Prolonged apnea, resulting in bradycardia and color change, that requires vigorous stimulation or positive pressure ventilation (it was previously also called a "near-SIDS event"). The manifestations are typically frightening to the caretaker.

List 11 causes of ALTE.

Apnea, airway obstruction, inborn errors of metabolism, seizures, sepsis, heart disease, apnea of prematurity, breath holding, gastroesophageal reflux, poisoning, and Munchausen syndrome by proxy or child abuse

What are the treatment options?

1. Up to 30% of cases may be resolved by treating the primary cause.
2. For remaining cases, **apnea monitoring** and **CPR training** of the parents may be recommended.

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What is SIDS?

Sudden **i**nfant **d**eath **s**yndrome: The **sudden death** of an infant **under 1 year of age,** which remains unexplained after a thorough **case investigation,** including performance of a complete **autopsy,** examination of the **death scene,** and a review of the **clinical history** (WHO definition)

What is the incidence?

1–2/1,000 live births; results in 6,000–10,000 deaths yearly, with a peak incidence at 2–4 months of age

What are the risk factors?

1. Infants who have had an ALTE—these infants are at increased risk of dying of SIDS. However, over 95% of infants who die of SIDS have never had such an event.
2. Premature infants
3. Infants of substance-abusing mothers
4. Infants put to sleep on their abdomens
5. Parental smoking

List 6 rules for parents to prevent SIDS.

1. Placing infants **"back to sleep"** (i.e., in a supine sleeping position) is recommended nationally.
2. Have infants on **firm** rather than soft bedding.
3. Avoid overly warm sleeping quarters.
4. Avoid overbundling.
5. Avoid placing objects such as stuffed toys in the cradle or crib.
6. Refrain from smoking around infants.

Use of a pacifier and breast-feeding may also decrease the risk of SIDS.

NEONATAL DIARRHEA

What is it?

Abnormally frequent, loose stools in an infant; may be associated with dehydration, failure to thrive, or systemic illness

List 2 complications.

Profound dehydration and nutritional deprivation (dangerous for the developing nervous system)

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List 7 common causes.

1. Infections (the most common causes), especially **rotavirus** (after 4 months), but also *Salmonella*, *Campylobacter*, and *E. coli* (during the first 2 months)
2. Primary or secondary carbohydrate malabsorption
3. Fat malabsorption
4. Congenital malformations of the intestines
5. Acquired defects of the bowel (e.g., short-gut syndrome—Ch 19, p. 252)
6. Hormonal abnormalities (e.g., thyrotoxicosis, congenital adrenal hyperplasia)
7. Allergic conditions (e.g., intolerance to cow's milk protein)

List 3 ways neonatal diarrhea should be evaluated.

1. **History:** should include family history, birth history, and a chronology of the illness
2. **Physical examination** should be detailed—after an initial assessment of hemodynamic stability and hydration status
3. **Laboratory tests:** serum electrolytes, BUN, and creatinine; CBC to assess for an associated anemia; and stool studies for culture, rotavirus antigen, WBC and occult blood, and reducing substances

What is the treatment?

After fluid and electrolyte stabilization, therapy is determined by the underlying condition. Antimotility agents are **NOT** used in infants and have a significant morbidity rate in this age group.

UMBILICAL ABNORMALITIES

OMPHALITIS

What is it?

Infection of and around the umbilicus and the retained umbilical remnant in the infant

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What are the signs and symptoms?

Fever and possibly signs of sepsis

What is the appearance of the umbilicus with omphalitis?

Erythema around umbilicus, often extending in a streak up the abdomen along the umbilical vein—infection may spread within hours to become a **frank fasciitis** with **crepitus** and tissue **necrosis.**

List 3 components of treatment.

1. Broad-spectrum antibiotics
2. Umbilical remnant may need to be removed.
3. Aggressive surgical debridement if fasciitis is present

List 2 ways it can be prevented.

Hand washing, asepsis in handling of fresh cord

UMBILICAL HERNIA (SEE CH 23, P. 359)

UMBILICAL GRANULOMA

What is it?

Persistent granulation tissue on umbilicus after separation of umbilical cord

What are the signs and symptoms?

Persistent discharge or oozing at the site of granuloma. Cellulitis may develop around the site, leading to omphalitis.

List 3 components of treatment.

1. Mild cases respond to 1–2 applications of silver nitrate.
2. Some granulomas require surgical excision.
3. If cellulitis begins, infant should be treated with intravenous antibiotics and observed for any progression of infection.

PERSISTENT URACHAL REMNANT

It is a remnant of what structure?

The embryologic connection of the umbilicus to the bladder (allantois); it may take various forms: urachal cyst, urachal sinus, or urachal fistula.

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What are the signs and symptoms?

1. Persistent discharge of mucus, pus, or frank urine from umbilicus
2. Cellulitis or sepsis if remnant becomes infected
3. Possible mass (may be tender) in infraumbilical midline position

How is it diagnosed?

Usually by physical examination. Sinogram (contrast study through the draining umbilical site) or cystogram may reveal sinus or fistula. An ultrasound may be useful to diagnose a cyst.

What is the treatment?

Surgical excision

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Chapter 11

Newborn Intensive Care: General Considerations

RESPIRATORS

List 4 basic modalities of oxygen therapy.

1. Nasal cannula with oxygen flow
2. Head box with humidified, heated oxygen to prevent excessive heat loss in the infant, with continuous monitoring of FiO_2
3. **CPAP: C**ontinuous **p**ositive **a**irway **p**ressure administered through nasal mask, nasal cannulae, or endotracheal tube
4. Endotracheal intubation with mechanical ventilatory support

List 2 commonly used types of respirators.

1. **Conventional:** oxygen is delivered 20–40 cycles/min and may be pressure limited, time cycled, or volume limited.
2. **High-frequency:** facilitated diffusion of gases in the lungs at 600–900 cycles/min; may be a jet ventilator or oscillator

MONITORS

List 5 ways infants are monitored in the intensive care setting.

1. **Cardiorespiratory (CR) monitor:** detects apnea, bradycardia
2. **Arterial catheters:** catheters placed in an umbilical or peripheral artery for blood gas or chemistry sampling and for continuous BP monitoring

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3. **Pulse oximetry:** continuous transcutaneous monitoring of arterial oxygen saturation
4. **Transcutaneous oxygen monitoring ($TCpO_2$):** electrode applied to the skin measures oxygen crossing the skin membrane; continuous measurement; correlation with arterial PO_2 varies with infant; good for monitoring trends; unreliable with poor perfusion
5. **Transcutaneous carbon dioxide monitoring $TCpCO_2$:** same issues as $TCpO_2$

NUTRITION

What IV fluids are appropriate for the newborn infant?

$D_{10}W$ in first 24 hours, except for the very low-birth-weight infant who may require only D_5W; add "1/4 normal" saline after 24 hours. In the low-birth-weight infant, amino acid infusion should begin shortly after birth.

At what rate for IV fluids?

80–100 mL/kg per day

When is hyperalimentation used?

For the low-birth-weight infant or ill term infant

What are the goals of hyperalimentation?

Goal: ~80–120 kcal/kg per day, including 3–4 g/kg amino acids, 3–4 g/kg fat; calcium, phosphorus, and other electrolytes and vitamins are added

List 2 administration routes for hyperalimentation.

Peripheral IV (limited to 12.5% dextrose) and central venous catheter (up to 25% dextrose)

List 4 routes of enteral feeding.

1. Breast- or bottle-feeding: Premature infants are usually able to begin to PO feed between 32 and 33 weeks. For premature infants, feedings are increased over 5–7 days.
2. Gavage: feedings through an orogastric or nasogastric tube every 2–4 hours or by continuous infusion

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3. Nasoduodenal: continuous feeding through transpyloric duodenal tube
4. Gastrostomy: G-tube may be placed when an infant undergoes abdominal surgery and a quick return to oral feeding is not anticipated, or when infant's feeding skills are poor.

List 4 basic categories of enteral feeds.

1. **Breast milk:** (~20 cal/oz) the ideal nutritional source for most infants. Improved absorption and anti-infection properties. Caloric density, protein, and mineral content can be increased with additives for the low-birth-weight infant.
2. **Standard formula:** 20 kcal/oz iron-fortified for term infants (same caloric concentration as breast milk)
3. **Premature formula:** 24 kcal/oz (increased sodium, calcium, phosphate, and vitamins for improved growth and bone mineralization)
4. **Elemental formula:** blend of hydrolyzed protein and modified fat for improved absorption in infants with malabsorption or feeding intolerance

Formulas may be milk- or soy-based. (**Note:** 1 ounce equals 30 mL.)

ENVIRONMENT

Why must a premature baby be kept in an incubator or a bed with an overhead warmer (i.e., a controlled environment)?

The infant's body surface area is large relative to body mass, and developmentally, he or she cannot regulate body temperature adequately. Metabolic demands are increased if the infant is kept too warm or too cool.

When can a premature infant regulate his or her own body temperature effectively?

Usually, when a weight of 1,600–2,000 g is attained

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What is the most likely cause when a premature infant's body temperature is too high or too low?

The temperature of the infant's environment. If appropriate, other reasons for the infant's altered temperature (especially sepsis) must be sought.

COMMON QUESTIONS PARENTS ASK

What are the survival rates among premature babies?

For infants with birth weights:

<500 g: <20–30%

500–750 g: 50–70%

750–1,250 g: 85–90%

>1,250 g: 95–98%

Congenital malformations and chromosomal anomalies affect these data.

List 8 common complications of prematurity.

Cerebral palsy, developmental delay, visual impairment, hearing impairment, learning disability, chronic lung disease, behavioral problems, and mental retardation

What is the incidence of severe complications among premature infants?

For infants with birth weights:

<1,000 g: 25–30%

>1,000 g: 15–20%

When can an otherwise healthy premature baby be discharged?

Without major complications, a premature infant is expected to be discharged shortly before its term due date.

Can a mother still provide breast milk for a premature baby?

Yes. Although the infant may not be able to directly breast-feed initially, the mother should pump her breasts at least every 3 hours beginning shortly after delivery. Breast milk offers significant nutritional and immunologic advantages to the premature infant.

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Section III

Ambulatory Pediatrics

Chapter 12

The Pediatric Physical Examination

THE WELL-CHILD VISIT

What is the purpose of the well-child visit?

To identify physical, psychosocial, and developmental problems; prevent disease; provide guidance and advice to parents

List 7 components of the well-child visit.

1. Identify and respond to parental concerns
2. Historical assessment of physical and psychosocial growth and development
3. History of family-child interactions and problems
4. Age-specific physical examination to look for previously undiagnosed problems, assess previously identified problem areas, and assess normal neurologic development
5. Screening laboratory tests
6. Immunizations
7. Anticipatory guidance

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At what ages should visits be scheduled?	3–5 days of age, 1 month, 2 months, 4 months, 6 months, 9 months, 1 year, 15 months, 18 months, 2 years, 30 months, 3 years, and every year thereafter. A newborn discharged from the nursery before 48 hours of age should be examined within 48 hours of discharge. These are recommendations of the American Academy of Pediatrics (AAP).
List some common parental concerns for an infant:	
Newborn to 2 months of age?	Sleep schedules, feeding, crying
2–3 months of age?	Sleep, interpreting cries, initiation of solid foods, effect of mother going to work
4–6 months of age?	Sleep, scheduling naps, initiation of solid foods, effects of day care
6–9 months of age?	Motor development, child's tolerance of solid foods, patterns of discipline
9–12 months of age?	Motor development, temper tantrums, fear of strangers
12–18 months of age?	Temper, limit setting, night walking
18–24 months of age?	Temper and violence toward other children, limit setting, language abilities
24 months of age?	Toilet training, playing with others
36 months of age?	Social skills
List 4 physical measurements that should be recorded and give the frequency.	1. **Weight:** at every visit (unclothed) 2. **Length:** at every visit until the child can stand cooperatively; then height is measured and recorded

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3. **Head circumference** at each visit. Record maximum occipitofrontal circumference.
4. **BP:** in all 4 extremities at 1 month of age to assess for coarctation of the aorta; routine single extremity BP at every visit beginning at 3 years of age

How are the growth variables tracked?	Using standardized growth charts, with growth curves expressed in percentiles
When is the physical growth considered abnormal?	If the child's growth pattern deviates by more than 1 standard deviation from its previous percentile or is more than 2 standard deviations from the mean
What 4 areas of development are monitored?	Gross motor, fine motor, social, and language development (See Ch 7, p. 59, for the milestones, by age, for these areas of development.)
What is the most commonly used developmental screening test?	The Denver Developmental Screening Test
At what age should malformations of fetal development or stigmata of syndromes be assessed?	At birth
At what ages should intra-abdominal masses be looked for?	Birth to school age
At what age should retinoblastoma be looked for?	Throughout the first 3–4 years
At what age should cardiac murmurs be looked for?	At each well-child visit

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At what age should the visual function be assessed?

By 3–4 months

At what age should the congenital hip dysplasia be looked for?

At each visit, until child is walking

When does the normal tooth eruption begin?

At 6 months; normal range is up to 15 months

List 2 early milestones of normal hearing.

Responds to sound by 2 months; orients to sound by 4 months

List 3 types of office screening for hearing.

1. Play audiometry at 3 years
2. Pure tone audiometry at 4 years
3. Brainstem-evoked audiometry by age 6 months if child is in a high-risk group or if a newborn screen is abnormal

When does the normal development of secondary sexual characteristics begin?

Girls: between 8 and 12 years of age

Boys: between 10 and 14 years

When does scoliosis commonly become apparent?

Beginning with the onset of puberty through Tanner stage 4

SCREENING LABORATORY TESTS

List 6 common screening laboratory tests, and when they are performed.

1. Screening for a variety of **metabolic diseases** (e.g., PKU, hypothyroidism, sickle cell disease, medium-chain acyl-CoA dehydrogenase [MCAD] deficiency, galactosemia) is performed at birth on a state-by-state basis; these tests may need to be repeated if the child is discharged from the nursery early, or if the child received a blood transfusion prior to testing.
2. Screening for **sickle cell disease** (based on ethnicity) at 9 months of age, if not done at birth

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3. Screening for **anemia** between 9 and 12 months of age, at entry to school, once in mid-childhood, and once in adolescence. Earlier testing may be indicated if there are neonatal problems.
4. Screening for elevated **lead levels** as per current AAP and CDC protocols
5. **Urinalysis** is controversial, but children are often screened once in infancy and again at school entry.
6. Screening for **tuberculosis**—most children are screened at 12 or 15 months of age and again at school entry. Frequency of screening depends on the area of the country and the child's background (check cross reference) (see Ch 28, p. 497, for the Mantoux tuberculin skin test and childhood tuberculosis in general).

IMMUNIZATIONS

List 12 diseases against which children are routinely immunized.

Diphtheria, pertussis, tetanus, polio (inactivated), measles, mumps, rubella, *Haemophilus influenza* type B, varicella, hepatitis A and B, and *Streptococcus pneumoniae*

What source is the authority on immunization schedules?

The AAP's *Redbook*; it is updated every 3 years.

List 2 other vaccines that may be administered.

Influenza and *Neisseria meningitidis*

ANTICIPATORY GUIDANCE

List 6 topics on which anticipatory guidance may be given to parents.

Injury prevention, poison prevention, development stimulation, nutrition, behavioral development, child's adjustment to family disruptions (e.g., new siblings, moves, illness)

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What guidance about injury and poison prevention should be given? (List 7 topics)

1. Use of a proper **infant car seat** from birth to 4 years of age (or 40 pounds), followed by **seat belt** use thereafter (a lift for the seat ["booster seat"] is recommended for children of young school-age years until the adult seat belt fits correctly without using the booster seat)
2. **Poison prevention**—beginning at 6 months, with reinforcement thereafter. Parents should be advised to conduct a formal safety check of the home before 6 months; discuss drugs, household chemicals, and plants.
3. **Firearm safety** in the home beginning at birth
4. **Swimming lessons** beginning by 3 years of age
5. **Bike helmet** use beginning with first tricycle
6. **Burn prevention** throughout childhood; advise parents to adjust water-heater temperature to maximum of 125°F at birth of first child.
7. **Avoid the use of walkers.**

Guidance about child's development? (List 3)

Discuss milestones achieved and those to be achieved in the interval before the next checkup. Discuss ways to help the child achieve the next milestone in a fun way.

Guidance and advice about nutrition? (List 8)

1. Breast-feed until 1 year; use formula if breast milk is unavailable or if there is a contraindication to breast-feeding (Ch 5, p. 23).
2. Start solid foods about 4–6 months of age (preferably 6 months).
3. Review the principles of a balanced diet regularly.
4. Review the elements of a heart-healthy diet.

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5. Review proper elements of food preparation and storage to avoid food poisoning.
6. Avoid constant overfeeding.
7. Discuss fluoride supplementation when necessary.
8. Monitor for nutritional practices that lead to iron deficiency.

Advice about smoking and tobacco products?

Begin discouraging parental smoking at the prenatal visit and every visit thereafter.

List 7 major family stresses that the primary pediatrician should be alert for.

Divorce; separation; absence of a parent (e.g., at work) for prolonged periods; parental or sibling illness or disability; a move from one house to another; death of a family member, close friend, or pet; natural disasters affecting the family, such as fire, flood, or hurricane

THE SCHOOL PHYSICAL

What is a "school physical"?

The formal assessment of a child entering kindergarten

When is it performed?

Usually within 6 months of school entry (children are usually 4.5–6 years of age); individual state laws vary

List 3 ways the school physical is different from the well-child visit.

1. Focuses on health and development as it relates to school readiness
2. Seeks to identify problems that may impair educational functioning of the child
3. May be considered (by some parents) as the last of the mandatory well-child examinations; therefore, special care should be taken to identify problem areas.

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List 8 general components of the school physical.

1. Immunization record review and completion
2. Developmental history and assessment
3. Screening hearing test
4. Screening vision test
5. Screening laboratory tests
6. Complete physical examination
7. Discussion of school placement and the child's potential strengths and weaknesses with the parents
8. Completion of the school form giving direction and advice to the school system about the child's unique needs, if any

List 3 immunizations that are given at the school physical.

1. Final dose of **polio** vaccine
2. Final dose of **DTP** vaccine, either regular DTP or DTaP vaccine
3. Second dose of **MMR** vaccine and varicella vaccine (if not previously administered)

Note: State laws vary and dictate which immunizations are required for school entry.

What certification must the physician sign?

Usually, the physician must certify that the child has received all appropriate immunizations, the child has a medical or religious contraindication, or (if the child is not fully immunized) a plan is in place to complete the required vaccines by an identified date.

List 4 screening laboratory tests that may be performed.

Usually **hemoglobin** (Hgb), **urinalysis,** and a **tuberculosis (TB) skin test;** in addition, **lead level** should be tested if there is any history of lead exposure, developmental delay, or anemia.

What hearing test is used?

A pure tone audiometry test in a quiet place

Describe this test.

Each ear is tested individually over a frequency range of 500–4,000 Hz, with a minimal threshold of 15–20 dB for the child to "pass" the test.

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List 3 components of the vision test.

1. Visual **acuity** is tested in each eye separately and in both eyes together using a Titmus vision testing machine or a method of equal accuracy.
2. **Color vision** should be tested.
3. An assessment of **strabismus,** including a cover test, is also important.

What are the key components of speech evaluation?

The child should **speak clearly** (i.e., physician readily understands) with little hesitation, using **complete sentences.** Occasional **stutter** may be normal, particularly if the child is excited, but facial grimacing, explosive speech, or frustration associated with the stammering is not. The **vocabulary** should exceed several hundred words, and the child should know colors and body parts and identify most objects to which the tester points.

What are the standards for math readiness?

1. Should understand the concept of "more" versus "less" and "above" versus "below"
2. Should be able to count and correctly identify the number of raised fingers (up to 5)
3. Should be able to recite back to the examiner **3-**number sequences presented orally

What should the physician do if concerned about the child based on the school physical?

Medical problems should be addressed, and the physician should alert the school to the potential problem so the school can perform an evaluation at the earliest possible date.

List 3 areas of consideration related to the child's size.

1. A child who is unusually small or large may be subject to extra emotional stress from classmates.
2. A child who is unusually small may be underestimated and treated like a younger child.

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3. If there has been a recent change in the growth pattern greater than 1 standard deviation, or if the child's size deviates more than 2 standard deviations from the mean, then the child should be assessed for comorbid illnesses associated with growth delay or with overgrowth.

How should the school be informed about any medications the child is taking?

A note is given to the school indicating the medication the child is taking, its therapeutic usefulness, possible side effects, and dosage schedule. The note should also indicate the primary illness that requires medication, whether it is contagious, and what effect it may have on the child's school performance.

List 8 causes of fatigue in a school-age child.

Late bedtime, getting up too early, an overly long bus ride, skipping meals, lack of time to rest or nap at school when the body is tired, frequent mild respiratory viral illnesses contracted in the school setting, obstructive sleep apnea, and over-involvement in after-school organized activities

List 2 common specific reasons for school physical examinations.

1. The preparticipation sports physical (Ch 13, p. 134)
2. The physical examination required every 3 years for children receiving special educational assistance

List 2 purposes of the every-3-years physical examination.

1. To assess any new or changed physical or developmental limitations, including changes in physical functioning (e.g., vision and hearing)
2. To evaluate new information about a child's chronic illness (e.g., new medications or complications) that affects the child's school performance

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COMMON CLINICAL PROBLEMS

WHEEZING

List 8 common causes of wheezing in infants.

Tracheal malformations, vascular rings, TEF (Ch 19, p. 300), mediastinal masses, aspiration, reflux, CF (Ch 17, p. 228), and infections

In toddlers? (List 5)

Infection (especially RSV and adenovirus; Ch 17, p. 221); asthma; CF; foreign body aspiration (Ch 17, p. 227); tumor

In older children? (List 4)

Infection (especially viral); asthma (Ch 17, p. 223); CF; tumor (especially lymphoma in the mediastinum; Ch 26, p. 416)

FEVER

What body temperature classifies as a fever?

Generally, a rectal temperature of >37.8°C is considered a fever, but some authors use a higher figure (38.0°C–38.2°C). Interpretation of fever may vary with the patient's age.

Does fever equal infection?

No, but the concern about the infection depends on the patient's age and clinical status. Infants with fever should be carefully evaluated for meningitis or septicemia. (See Ch 28, p. 462, for a discussion of meningitis.)

RESPIRATORY DISTRESS

List 8 causes of respiratory distress in children.

1. **Upper airway obstruction,** including **epiglottitis** (Ch 18, p. 239), peritonsillar or retropharyngeal abscess, foreign body in airway (Ch 17, p. 227), edema, vascular malformations (Ch 17, p. 229), intrinsic or extrinsic masses
2. Lower airway obstruction, including foreign body, bronchiolitis, and reactive airway disease

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3. Pneumonia (Ch 17, p. 221)
4. Congestive heart failure or pulmonary edema
5. Trauma
6. Spontaneous pneumothorax (especially thin, adolescent males; Ch 17, p. 230)
7. Metabolic diseases
8. Muscle diseases

ABDOMINAL PAIN

List possible causes to be considered in the differential diagnosis of abdominal pain in children and adolescents.

Viral gastroenteritis, appendicitis, mesenteric lymphadenitis, bacterial enterocolitis, Meckel diverticulitis, inflammatory bowel disease, hernia with incarcerated bowel, food poisoning, intussusception, abdominal adhesions, pneumonia, acute intermittent porphyria, trauma, volvulus, testicular torsion, ovarian torsion, ovarian cyst, tumors. In girls who have begun menstruating, dysmenorrhea and pregnancy must also be considered as a cause for abdominal pain.

DIARRHEA

List 3 causes of acute diarrhea.

Bacterial infections, such as *Salmonella*, *Shigella*, *Escherichia coli*, *Campylobacter*, and *Yersinia*; viral gastroenteritis; food poisoning

List 9 causes of chronic diarrhea.

Fat malabsorption, CF, dietary allergy, lactose intolerance, bacterial infection, celiac disease (gluten intolerance), malnutrition, antibiotic use, inflammatory bowel disease

VOMITING

List 13 causes of vomiting.

Viral gastroenteritis, food poisoning, upper GI obstruction, inborn error of metabolism, CNS tumor, motion sickness, sepsis, paralytic ileus, adhesive obstruction (if child has had a prior operation), incarcerated hernia (Ch 23, p. 357), malrotation, appendicitis, intussusception (Ch 19, p. 272)

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ACNE

What is it?	A skin condition commonly affecting adolescents, consisting of 4 basic types of lesions: open and closed comedones, papules, pustules, and nodular-cystic lesions
List 2 causal factors.	1. During adolescence, **androgens** stimulate the growth of sebaceous glands as well as the production of sebum. Some of these materials are hydrolyzed to free fatty acids, which can cause inflammation. 2. Characteristic abnormal **keratinization** of skin cells at this time also contributes to the lesions.
List 2 features of open comedones ("blackheads").	The orifice of the follicular duct is open and the involved sebum has been oxidized to the black color, usually without a surrounding inflammatory reaction.
List 2 features of closed comedones ("whiteheads").	The follicular duct is occluded, with a surrounding inflammatory reaction.
List 2 ways the development of acne can be minimized.	Cleansing the face 2–3 times daily with a mild soap, and avoiding oil-based skin preparations and makeup
List 3 ways acne is treated.	**Topical agents**—the most common are benzoyl peroxide and retinoic acid. **Systemic antibiotics,** such as tetracycline or erythromycin, may be needed in more severe cases. **13-*cis*-Retinoic acid** is used for the most severe cases of acne (cystic acne or acne conglobata). **Females of childbearing age being treated for acne should avoid pregnancy (consider birth control) because some treatments may have adverse effects on a developing fetus.**

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List 3 potential side effects of retinoic acid preparations.

1. There may be **carcinogenic effects** from the combination of light and retinoids. Therefore, avoidance of sun or, alternatively, the use of sunscreen is recommended for patients using retinoic acid.
2. Retinoic acid also may cause hyperpigmentation in patients with darkly pigmented skin.
3. Retinoic acid and some related compounds are **teratogens.**

INGESTION OF POISONOUS AGENTS

What is the peak age for poison ingestions?

2 years of age; however, teens also are prone to ingesting caustic substances as suicide attempts or gestures.

List 3 categories of commonly ingested household materials.

Cleansers (e.g., sodium hydroxide); batteries (potassium hydroxide); miscellaneous acidic agents (e.g., sulfuric acid)

What should be done if a child ingests a poisonous agent?

The poison control center should be called. Instructions will depend on the agent ingested. The routine use of ipecac is no longer recommended.

List 2 ways that a child who has ingested a caustic agent is evaluated.

Esophagoscopy within 24 hours and **barium swallow** within 48 hours to assess the degree of injury, stricture, and esophageal motility

List 2 components of treatment of ingestion of a caustic agent.

1. Ampicillin and gentamicin (or broad-spectrum antibiotics with similar coverage) with hydration should be started when ingestion is suspected.
2. Esophageal strictures may require dilation, placement of a feeding tube (nasogastric or gastrostomy), or anatomic replacement.

List the 2 best methods of prevention.

"Childproofing" the home and educating children and parents

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DEVELOPMENTAL DELAY

What is it?	Delay in attaining developmental milestones at the appropriate age (Ch 7, p. 58)
Does developmental delay equal mental retardation?	No. There may be reasons for developmental delay that are unrelated to cognitive skills.
List 3 ways for the physician to screen for developmental delays.	Careful history; thorough physical examination; use of a screening test (e.g., the Denver Developmental Screening Test)
What laboratory, radiographic, or other tests are indicated for developmental delay?	This part of the evaluation should be individualized using the history, physical examination, and the developmental evaluation as starting points.

SHORT STATURE

What is it?	Height less than the second percentile for age (**Note:** Definitions may vary.)
List the 2 most common causes.	Normal variation, constitutional delay
List 7 other causes.	Endocrine abnormalities (Ch 24, p. 378), metabolic diseases, genetic syndromes, skeletal dysplasias, chromosome abnormalities, chronic diseases, psychosocial short stature
List 5 ways in which it is evaluated.	History (including family history), review of growth data, physical examination, and appropriate radiographic and laboratory studies as indicated
Why are previous measurements so important?	Growth is a dynamic process; evaluation of change with time gives more information than isolated growth points.

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OBESITY

What causes obesity?	Caloric intake exceeds caloric expenditures.
What is the most common cause of childhood obesity?	Exogenous obesity (excessive intake)
What is the relationship of obesity to height?	Children with exogenous obesity tend to be taller than average. Most endocrine disorders and syndromes in which obesity is seen are associated with stature that is shorter than average.
List 4 syndromes associated with obesity.	Prader-Willi syndrome, Cushing syndrome (Ch 24, p. 390), pseudohypoparathyroidism type I, growth hormone deficiency (Ch 24, p. 380)

FAILURE TO THRIVE

What is it?	Usually, the term refers to failure to gain or maintain weight adequately.
List 7 causes.	GI disorders, immune disorders, chronic diseases (consider CF!), inborn errors of metabolism, inadequate nutritional intake, CNS abnormalities, psychosocial problems
List 6 ways it is evaluated.	Careful history, physical examination, weight measurements, review of growth data, and laboratory and radiographic studies as indicated

ENCOPRESIS (SEE CH 19, P. 290)

What is it?	Fecal incontinence caused by overretention of stool
List 6 causes.	Psychological problems, Hirschsprung disease (Ch 19, p. 293), chronic stool retention, neurologic abnormalities, hypothyroidism, previously unrecognized imperforate anus with perineal fistula

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List 5 ways it is evaluated.	Careful history, physical examination, review of growth data, and laboratory and radiographic studies as indicated. Evaluation should be done to rule out Hirschsprung disease (Ch 19, p. 293).
List 3 components of treatment.	Treatment depends on the etiologic factors. 1. Educating and supporting the parents and child are key. 2. A "clean-out" regimen, followed by a program to maintain regularity (if not caused by Hirschsprung or imperforate anus) 3. Attention to emotional and behavioral issues

ENURESIS

What is it?	Urinary incontinence in child 5 years of age or older
What is the differential diagnosis?	Urologic abnormalities, neurologic abnormalities (including seizures), diabetes mellitus, diabetes insipidus, psychosocial stress
List 5 ways it is evaluated.	Careful history, physical examination, review of growth data, laboratory evaluation (including urinalysis, regardless of suspected cause), and radiographic studies as indicated
What is the treatment?	Depends on the etiologic factors
List 3 components of therapies of nonorganic enuresis.	Medications, alarms, behavioral modification

SCHOOL PHOBIA

What is it?	Fear of school or refusal to attend school
List 3 of the common potential causes.	Real fear of the school environment (e.g., bullies, violence); fear of a teacher; fear of separation from the parent or family

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List 3 components of evaluation when somatic complaints are present.

History; physical examination as indicated; detailed interview with the parents and the child

What is the treatment goal?

The goal is to normalize the child's school experience, which usually involves returning the child to the classroom as soon as possible. Parental education is important, as well as additional counseling if the school phobia is a manifestation of more serious emotional problems.

ATTENTION DEFICIT HYPERACTIVITY DISORDER (ADHD)

What is it?

A disorder characterized by limited capacity for attention, overactivity, and impulsivity. It is distinct from abnormal conduct behavior and specific learning disabilities.

What are the etiologic factors?

These are still not fully known.

What is the prevalence?

About 7% of children (There are studies with higher and lower figures.)

Is it more common in boys or girls?

Boys, by a 5:1 ratio

What is the age of onset?

Usually before 4 years of age

Is family history important?

Yes. ADHD is more common in children who have had family members with ADHD.

List 5 typical characteristics of the history of a child with ADHD.

A history of behavior in specific situations is important, as are birth and neonatal histories. There is often a history of difficult birth, colicky behavior as an infant, sleep difficulties and feeding difficulties as an infant, and excessive temper tantrums as a toddler.

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List 4 typical clinical manifestations.

A child (especially in school) may be uncontrollable, refuse to sit still, intrude on other children, and refuse to follow instructions.

During formal examination, symptoms may be difficult to detect because these children can behave well in significantly structured situations.

On what basis is the diagnosis made?

Usually on a clinical basis

List 7 other causes of difficult behavior for which children with symptoms of ADHD should be tested.

Specific learning disabilities, hearing impairment, petit mal epilepsy, side effects of medication, anxiety, depressive disorders, or poor living situations

List 3 potential components of the treatment.

1. Behavioral and psychosocial therapy with a confirmed structure to the child's environment
2. Stimulants, including methylphenidate (Ritalin), dextroamphetamine, pemoline, and clonidine, are sometimes used in conjunction with these therapies.
3. Tricyclic antidepressants may also be efficacious.

What is the prognosis?

Prognosis appears to be better if a child with this condition does not exhibit aggression. There are concerns that children with ADHD may be more prone to alcoholism, sociopathy, and hysteria in adulthood. Steady gainful employment seems to be helpful.

LIMP

List 10 causes of limp.

Foot problems (e.g., calluses, foreign bodies, warts, shoe problems), sprains, strains (Ch 13, p. 138, re sprains and strains), fractures, dislocated hip, toxic synovitis, osteomyelitis, soft tissue trauma, arthritis (septic, inflammatory), and cancer (e.g., osteosarcoma, leukemia, neuroblastoma; Ch 26, p. 406)

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List 4 ways the limp is evaluated.	History and physical examination, laboratory and radiographic studies as indicated

HEADACHE

Are headaches a sign of dangerous disease?	Rarely
What are the 4 signs that a headache may be serious?	Excruciating pain, stiff neck, accompanying neurologic findings, impairment of consciousness
List 6 characteristics of migraine.	Rapid onset; pain is often hemicranial and behind the eye; visual changes (e.g., seeing flashing lights, black spots); pain is intense and pounding; photophobia; child sleeps and then awakens without headache
Is a family history of migraine common?	Yes
What are the possible treatments?	Sleep is often the best treatment. Other therapies include ergot derivatives, isometheptene, and analgesics. Some patients may require chronic treatment with β-blockers, tricyclic antidepressants, or calcium-channel blockers. Biofeedback or relaxation therapy may be helpful.
List 7 characteristics of tension headaches.	1. Slow onset of pain 2. Bifrontal distribution 3. Pain is not really debilitating 4. Pain is squeezing in quality and may have a throbbing component 5. No visual changes or photophobia 6. Gradual remission of pain 7. May be associated with definable psychological stress
List 3 treatments of tension headaches.	Analgesics, biofeedback therapy, relaxation therapy

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Chapter 13 Pediatric Sports Medicine

GENERAL ISSUES IN SPORTS MEDICINE

What is the role of a primary care physician in sports medicine?

Roles range from being a team physician to providing medical care dealing with an individual player's specific illness. The primary care physician should be comfortable addressing injuries to bone and joints, nutrition, fitness, and health issues as they relate to sports.

What is the difference between "weight lifting" and "weight training"?

"Weight lifting" is the process of pressing and jerking maximum amounts of weight for a one-snatch opportunity. **"Resistance strength training"** ("weight training") involves selecting a weight that can be lifted or moved in a series of repetitions to attain increased strength and flexibility.

What is the role of weight training for prepubertal boys and girls (Tanner stage 1 or 2) (Ch 14, p. 155)?

1. There is disagreement about the effects of weight training by prepubertal children.
2. Weight training will not result in significant increases in muscle mass until testosterone levels increase later in puberty.
3. Specific muscle training under the supervision of a knowledgeable trainer may increase strength and flexibility to some degree. There is no evidence that this correlates with improved athletic performance.
4. Concern exists about abuse of weight training causing damage to growth plates. (**Pain** is a good indicator that muscle, tendons, or ligaments are being abused.)

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At what age should a child be allowed to begin participation in competitive sports? Collision sports?

The American Academy of Pediatrics guidelines suggest:

1. Competitive sports: no child younger than 6 years
2. Collision sports: no child younger than 10 years
3. Obviously, individual children may be ready sooner than this, whereas others may not be ready even on reaching those guidelines for age.

List 2 criteria for allowing a child to participate in a sport.

1. The child states a sincere interest in participating.
2. The physician must feel the child is able to participate safely, based on the physical assessment of the child and the physician's knowledge of the sport and the program the child will be joining.

PREPARTICIPATION SPORTS ASSESSMENT

Is there a standard form or set of requirements for sports PE?

No. Although the American Academy of Pediatrics and American Academy of Family Practice have made recommendations on this issue, state requirements vary. The National Federation of State High Schools and the Compendium on Personal Protective Equipment (PPE) have good templates to consider.

What 5 particular factors are important during a comprehensive sports PE?

In addition to the components of a standard physical examination, a sports PE should focus on:

1. **History**—provides highest yield of factors that might affect an athlete's participation. History of any previous injury, individual or family cardiovascular conditions, and previous concussions and head injury should be obtained.
2. **Assessment of physical maturity**—may reveal factors for disqualifying a girl or boy from participating in a collision sport

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3. **Musculoskeletal examination**—the **most common source** of physical findings that predispose to recurrent injury
4. **Measure of fitness and state of nutrition**—especially important for sports in which weight loss is common (gymnastics and wrestling) or in which obesity might predispose the athlete to heat injury (e.g., football practice in hot weather)
5. **Analysis of history and physical examination and knowledge of the nature of sports**—necessary for making an appropriate recommendation for specific sports participation

List 2 ways in which physical maturity is measured.

1. Tanner stage (sexual maturation rating) of pubertal development (Ch 14, p. 155).
2. Measure handgrip strength with a hand ergometer.

Boys and girls should reach what Tanner stages before participating in vigorous competition?

Boys at less than Tanner stage 4 should not participate in collision sports (e.g., football, lacrosse, ice hockey) with fully mature boys because of increased risk for injury, especially to epiphyseal plates. Physical maturity is much more relevant to injury risk than weight or size.

Girls who reach 15 years of age and are still at Tanner stage 2 or less should receive close scrutiny and evaluation before participation in "weight-conscious" sports, such as gymnastics, dance, or cross-country running.

List 3 areas of fitness that should be determined at the time of the sports PE.

Cardiovascular, pulmonary, and general health and fitness (including nutrition)

List 3 ways cardiovascular fitness is measured.

1. **BP**
2. **Cardiac examination** for murmur and rhythm, especially after exercise with the heart rate increased

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3. **Exercise.** One practical method: the athlete performs a 2-minute jumping-jack task at 1 jump/s. The physician measures resting pulse, pulse immediately after exercise, and pulse after a 1-minute recovery period. A rise in pulse to >95 beats/min during exercise, or a drop to <2/3 of the resting level on recovery would be an indication for slow and cautious advancement to full practice and activity.

List 2 ways to measure general health, fitness, and nutrition.

1. **Height:weight proportion:** use standard growth chart.
2. **Body fat:** the appropriate range in girls and women is 10–25%; in boys and men, 7–20%. Body fat lower than these ranges may indicate **malnutrition** or **eating disorders.** Higher body fat indicates obesity and could be a problem in acclimatization to heat.

In what 2 ways is the body fat percentage determined?

1. Determining body density by water immersion (most accurate)
2. Measuring skin fold with calipers

How is the pulmonary fitness measured?

Some sports medicine specialists recommend a pre-exercise and postexercise FEV_1 test (forced expiratory volume in 1 second) to identify exercise-induced bronchospasm. As much as 10% of the athlete population experiences this condition enough to affect performance.

What laboratory tests should be included in a sports PE?

Routine laboratory tests are not needed for all children; tests should be based on the history or physical examination findings and whether assessment of anemia has been done in recent years. Screening for hemoglobinopathies, including sickle cell trait, is recommended if it has not been previously done.

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List 2 instances in which it is reasonable to include urinalysis in an asymptomatic patient as part of a sports PE.

1. For baseline data in case of future injury to the kidney
2. If the boy or girl has not had a urinalysis in the past 2 years and is receiving no other health care

In what instance is it reasonable to test Hgb and Hct as part of a sports PE in an asymptomatic patient?

If the athlete is not receiving any other health maintenance

What recommendation should be made if a boy or girl has only 1 kidney or has experienced 5 or 6 concussions?

Probably **not to participate in collision sports** (although in the case of 1 kidney, more latitude is typically given). If the family insists on participation, precedents have been set so that courts may allow a child to participate over a physician's recommendation; appropriate protection and caution should be shared.

What are the 8 key ingredients to an excellent sports preparticipation evaluation?

1. Thorough sports-specific and medical and family history
2. Thorough physical and neurologic examination
3. Additional examination:
   - Tanner staging
   - Body fat measurements
   - Pulse—resting, exercise, and recovery
   - FEV_1—preexercise and postexercise
4. Some measurement of iron stores and anemia
5. Performed 2–4 weeks before sports practice begins
6. A quiet and private environment convenient for the athlete, conducive to a good examination and an opportunity for conversation
7. Performed by individual(s) knowledgeable and interested in athletes and in all aspects of sports
8. Summation by a physician and appropriate recommendation and follow-up for the athlete, family, coach, and school

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MUSCULOSKELETAL INJURIES

What are the 2 major types of musculoskeletal injuries?

1. **Traumatic injury:** acute and involves an impact or undesired motion
2. **"Overuse" injury:** may seem acute but usually has an insidious onset over the course of a long time (e.g., stress fracture)

List 7 factors that predispose a sports participant to musculoskeletal injury.

1. Reinjury of a previous injury that had not been totally rehabilitated—**the most common cause of injury**
2. Imbalance or asymmetry in muscle strength or range of motion
3. Overuse of particular joint(s) or muscle(s)
4. Improper warm-up and stretching, which limit the flexibility
5. Improper equipment
6. Improper technique in an athletic skill
7. Poor conditioning

What is the difference between a strain and a sprain?

Sprain: injury to a ligament

Strain: injury to a tendon or muscle

What is the most common soft tissue injury?

Muscle hematoma or contusion (a.k.a. "Charley horse"). The goal is to **minimize the amount of bleeding** to prevent myositis ossificans (muscle calcification).

MANAGEMENT OF INJURIES

ACUTE MANAGEMENT

List 5 steps in the acute treatment of a traumatic injury.

1. Calm the injured patient and move others away.
2. Assess for life-threatening situations such as compromised airway, major bleeding, or neck or vertebral spine injury.
3. Get history of the nature of injury and whether any "pop," "snap," or other acute feeling or sound was noted.

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4. Check for the swelling and ask when it first occurred (if physician was not at the scene of the injury). Immediate swelling is generally caused by bleeding. Swelling after the first hour is usually secondary to inflammation and exudation of tissue fluid.
5. **"PRICEM"** (a variation of the more familiar "RICE")—is always appropriate for any musculoskeletal injury.
   - **Protection:** If appropriate and possible, immobilize the joint above and below the injury until further assessment.
   - **Rest:** Complete rest for a significant injury is usually indicated for a **minimum** of 48 hours.
   - **Ice:** Cold packs applied for 20 minutes every 2–4 hours minimizes swelling from bleeding or inflammation. This can shorten the recovery time and reduce muscle destruction.
   - **Compression:** Pressure (e.g., hand compression, elastic bandage) can control bleeding and swelling and minimize destruction of tissue and rehabilitation time.
   - **Elevation:** When possible, elevating the injured area will enhance resolution of swelling.
   - **Medicine:** Judicious use of pain and anti-inflammatory medication.

List 3 components of "early rehabilitation."

1. Alternate heat and ice: ice for 10–20 minutes, heat for 15 minutes, ice again for 10–20 minutes (2 or 3 times per day)
2. Stretching and passive range of motion exercises to the point of minimum discomfort should be done midway through the ice phase of treatment. Active range of motion exercise can begin when no significant pain occurs.
3. Isometric contractions against resistance can begin. **Always let pain be the limiting guide.**

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ADVANCED LEVEL REHABILITATION

When does "advanced level" rehabilitation begin?

It begins when all the above levels are accomplished without pain.

List 5 components of advanced level rehabilitation.

1. Continue most early rehabilitation activities (use heat to the injured area before exercising and ice after exercising).
2. More intense stretching, especially before and after exercising
3. Exercising should consist of range of motion against resistance (e.g., using weight training principles). Isometrics continue to be good.
4. Cross-training is important to keep noninjured muscles and cardiovascular system fit. It also helps the athlete to feel she or he is active and contributing to self-healing. For example, judicious use of exercise in water or bicycling provides low impact, good aerobic conditioning so long as careful consideration for the injury site is maintained.
5. A protective device for the injured region can be helpful during rehabilitative exercise (e.g., knee or ankle braces; elastic wrap for compression of a muscle injury).

How long does advanced level rehabilitation continue?

Until the injured area can be used in a normal manner without difficulty; this can last from approximately 14 days to 4 weeks after injury.

How soon can an injured athlete return to active participation in sports?

2–8 weeks or more depending on the injury. See the next answer.

What is the best approach to use with athletes during the return-to-action stage?

Explain to an athlete what must happen before she or he can participate rather than give a specific length of time. **Pain and functionality** are the main determining factors.

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How does one approach an overuse injury?

Same as for traumatic injury, except that a reduction in activity or cross-training may be satisfactory instead of total withdrawal from activity.

How does the physician treat a specific injury such as a sprained ankle or a painful knee?

Evaluate the injury as discussed earlier. If there is no underlying fracture or major instability suggesting disruptions of ligaments, tendons, or other supporting structures, then following the progression of management as described earlier is appropriate.

What is the role of "TENS," ultrasound, "whirlpool," physical therapy, occupational therapy, and so forth, in treating these injuries?

For most mild to moderate injuries (the vast majority of those that occur), the above outline of management by a primary care physician is all that is needed.

For more severe injuries, referral to an orthopaedist, a sports medicine facility with an athletic trainer, or other therapist should be done. Referral may be helpful if the athlete does not seem likely to follow a program on his own.

The therapist may choose to use electrical stimulation or other treatment modalities. Data supporting their effectiveness are fairly limited, but they appear harmless and may provide placebo benefit if nothing else.

How are the injuries graded in severity?

Injuries are graded by a somewhat arbitrary method meant to suggest the amount of damage and the length and type of management and rehabilitation necessary.

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List the grades of ankle sprains, with the characteristics of each.

Grade I—Mild pain and slight swelling, stable joint tests, normal range of motion, pain-free weight-bearing

Grade II—Moderate pain and intermediate swelling, stable joint, decreased range of motion by a few degrees, and painful weight-bearing

Grade III—Severe pain, significant swelling that obliterates landmarks, unstable joint, limited range of motion, and inability to bear weight

HEAD INJURIES

How are the concussions graded?

There are many grading methods with loss of consciousness, pain, confusion, and amnesia being important ingredients. There is not a uniformly accepted system, but **any head injury should be considered potentially serious** until proven otherwise. For simplicity in this discussion, we will consider 3 grades of head injury:

1. **Mild concussion**—player is "dazed" without amnesia and no loss of consciousness
2. **Moderately severe concussion**—confusion after the event, retrograde amnesia, or both, plus loss of consciousness for less than 1 minute or even without loss of consciousness
3. **Severe concussion**—signs of moderately severe concussion plus significant loss of consciousness and a prolonged period of confusion

The latest thinking emphasizes more on the course of the concussion than the initial presentation for evaluating severity; no initial symptom (LOC or amnesia) is an accurate prognosticator of the course of recovery of that concussion. New techniques (such as neuropsychological testing) show that even mild concussions with few symptoms can be associated with abnormal cognitive function beyond the time previously thought.

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How is each type of concussion managed?

All concussions, regardless of severity, should be managed in the same manner.

1. The athlete should be removed from participation and evaluated immediately for any life-threatening or significant neurologic abnormalities.
2. The athlete should then be evaluated for signs of abnormalities in mental status, memory, cognitive function, and other classic signs of concussion (dizziness, "fogginess," tinnitus, change in personality, irritability, etc.). Studies suggest that asking the player questions relating to their current activity ("What team are you playing?" "What is the score?") is more relevant to the player's possible confusion than standard orientation questions ("What is your name and address?") in identifying the concussed from the nonconcussed athlete.
3. No high school level athlete or below should be returned to play ("RTP") in any practice or game if even the mildest concussion has been diagnosed. Even if the athlete appears "clear" after 15 minutes, studies show delayed abnormalities can occur in that setting, and RTP is not recommended that day and medical clearance is required before RTP should occur.
4. Once the athlete becomes totally asymptomatic, she or he may be exercised aerobically in a low-impact activity and then rechecked for return of symptoms. This can be gradually increased in intensity and duration until it simulates the level of the specific sport. Use of computerized neuropsychological instruments, assessment tests such as SAC or SCC, may be helpful in providing an objective measure to the athlete's status as they are not always "accurate or truthful" about their symptoms if they are eager to return to play ASAP.

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5. The athlete may then be returned to practice to participate in all drills and activities, except scrimmaging. If no symptoms return and no abnormalities are identified on testing, the athlete may then be returned to full participation.
6. This new "return-to-play" guideline simulates more the RTP after other injuries as in gradual increase in activity, emphasis on functionality, and not the predicted length of time until RTP.
7. Any return of symptoms should return that athlete to a previous lower level of activity with gradual increase again when appropriate.

What is the danger of repeated head injuries?

The **second impact syndrome.** Death can even occur from a relatively minor blow to the head occurring within a few weeks of a previous concussion if brain swelling persists. This is the rationale for not allowing an injured player to return to action for a period of time and until **all** symptoms are gone.

How many concussions are too many?

Concussions can have a cumulative destructive effect. Cognitive deterioration, parkinsonian-type disorder, and "boxer syndrome" have all been attributed to repeated concussions. There are no data to show exactly how many concussions should create major concern.The advent of new testing methods should help physicians give better advice after repeated concussions. The data suggest that every additional concussion increases the likelihood of a subsequent one in that individual athlete. It is not clear whether there is a cause and effect relationship. Some researchers now feel that if baseline cognitive testing results are available and the athlete returns to that level after a concussion, no permanent damage exists and return to play is acceptable regardless of the number of previous concussions. Others are more cautious, but no "magic number" exists.

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What is a "stinger"?

A stinger is a burning or stinging of the arm and hand that can be accompanied by weakness and sometimes paralysis. This occurs from a stretching or compression of the neck, injuring the brachial plexus. Recurrent episodes may lead to permanent weakness.

What are the components of the management program for stingers?

Initial treatment consists of ice, resting the arm in a comfortable position, and anti-inflammatory agents. When the pain is gone and strength returns to normal, the athlete can resume participation. If symptoms persist for more than 24 hours, a more comprehensive evaluation and rehabilitation program should be arranged. Some athletes with long necks are predisposed to stingers, and a neck collar may be somewhat protective.

EYE INJURIES

Which eye injuries should be evaluated by an ophthalmologist?

Injuries with trauma to the globe or the periorbital tissues

List 5 serious sequelae of trauma to the eye.

Hemorrhage, retinal detachment, hyphema, lens dislocation, and orbital floor fracture

How can eye injuries be prevented?

By using protective eye wear—especially individuals with visual impairment in 1 or both eyes

MEDICAL ISSUES RELATED TO SPORTS PARTICIPATION

List 3 skin lesions that may prohibit an athlete from participating in a sport in which physical contact occurs.

Impetigo, tinea, herpes.

A boy or girl should not participate unless the lesions are covered or healing. This is a significant concern in wrestling. Infection with MRSA may also warrant consideration against participation.

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List 3 signs in an athlete with infectious mononucleosis that require a delay in returning to sports participation.

Fatigue, splenomegaly, lymphadenopathy. When splenomegaly is present, no activity that involves exertion or collision should occur for a **minimum of 2–4 weeks after the resolution of splenomegaly.**

What sports should an athlete with a bloodborne infectious disease (hepatitis B, AIDS, HIV) be strongly discouraged from playing?

Sports in which close physical contact and exposure to body fluids and blood occur (e.g., wrestling, boxing) pose the greatest concern.

The risk of this transmittal appears to be low, but theoretically it is not zero.

A physician can recommend that an athlete not participate in these sports if infected. However, the athlete cannot be forbidden from playing under present legal interpretations. **Note:** The physician also operates in the doctor-patient confidentiality relationship.

List 5 universal precautions that relate to sports participation.

1. An athlete should cover wounds whenever possible.
2. If a bleeding wound occurs, the individual's participation stops until the wound stops bleeding and is cleansed and covered securely. Any uniform contaminated with blood should be changed.
3. Skin exposed to blood or other body fluids contaminated with blood should be cleaned as promptly as possible with soap and warm water.
4. Rubber gloves and disposable towels should be used by individuals when cleaning and handling blood and blood-contaminated materials. These items should be placed in a container lined with a plastic bag for disposal.
5. Any surface contaminated with blood should be cleansed with fresh household bleach solution (1 part of bleach to 100 parts of water).

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In what sports should an athlete with asthma not compete?

With current therapeutic options, most young people with asthma should be able to participate in any sport they desire. Sports that require more prolonged and constant action (long-distance running, soccer, and field hockey) are more likely to cause difficulty than sports with short bursts of activity (football, tennis, and basketball).

List 2 ways athletes with asthma can maximize their safety and effectiveness in sports competition.

1. Use of peak flowmeters to monitor asthma status and helping an athlete learn about his or her "refractory period"
2. Preexercise medication (cromolyn, β_2-agonists, leukotriene inhibitors) can be helpful in minimizing bronchospasm.

What is the refractory period?

There is a period shortly after beginning exercise when reactive airway bronchospasm is particularly severe. Once that period is passed, there is a **refractory period** during which the airway seems to be particularly open. This period can last for a fairly extended time. If an athlete learns where his or her "bad" period is, she or he may warm up more vigorously for that prescribed period, so that when the game or practice begins she or he is already at the refractory period.

What is exercise-induced asthma?

Bronchospasm that occurs only with exercise

In what 3 ways is it recognized?

1. Shortness of breath occurs with exercise.
2. Bronchospasm occurs with exercise.
3. Hyperactive airway is identified in exercise portion of physical examination.

As many as 10% of athletes are identified in preparticipation sports PEs as having hyperactive airway.

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List 2 treatments for exercise-induced asthma.

Preexercise use of cromolyn or β_2-agonists

What is exercise-induced anaphylaxis?

Episodes of apparent syncope with characteristics simulating anaphylactic shock at or near the end of participation in an athletic activity. It can be confused with exercise-induced asthma.

List 3 possible causes.

1. Something triggers the release of histamine or some other mediator, often in people without a history of atopic disorders.
2. A combination of heat, stress, and dehydration may trigger the episode.
3. In other instances various foods appear to be the trigger—but only if the food was eaten near the time of the exercise, because the food alone is tolerated well.

List 4 components of management in severe cases.

Adrenalin (epinephrine), steroids, IV fluids, and **antihistamines** and any other supportive measurements appropriate for the situation

What other allergic disorder can create problems for athletes?

Severe generalized systemic reaction to stings from *Hymenoptera* (e.g., bees, yellow jackets, fire ants; see Ch 29, p. 510 for more on allergic reactions to insect stings and bites)

What athletes are at greatest risk from anaphylaxis and severe allergic reactions?

Participants who are outdoors at a distance from trainers and medical care (e.g., cross-country runners)

List 2 measures that athletes at risk for anaphylaxis can take to prevent or treat allergic reaction.

1. Carrying adrenalin (epinephrine) in an easy-to-administer form (such as an "epi-pen")
2. Consultation concerning the appropriateness of venom desensitization

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What are the 2 guidelines for participation in sports by young people with seizures?

1. **Noncollision sports**—Usually youngsters can participate in any noncollision sport including swimming and diving, so long as the seizures are well controlled with medications and the activity is supervised.
2. **Collision sports**—Controversy surrounds the appropriateness of playing a collision sport in which blows to the head typical of these sports could lower the threshold for seizure.

Generally, participation is permitted if control is good and the family and athlete are motivated and compliant.

What sports should a person with diabetes mellitus avoid?

With good control, good knowledge about his or her disease, and appropriate precautions and accommodations, a person with diabetes should be able to participate in any sport.

List 3 precautions that should be observed in athletes with diabetes.

1. The coach, the trainer, and other teammates should be aware of the athlete's disorder.
2. Glucagon and a source of sugar (e.g., juice, candy bar) should be available at all times.
3. The athlete should always carry identification that includes information about his or her diabetic condition.

What can the young athlete do to maintain control of diabetes if he or she chooses to participate in sports?

Learn what adjustments in diet and insulin (with close monitoring) may be necessary to maintain good control with variations in practice and game times.

Are athletes with "sickle cell trait" at increased risk for complications?

Although most athletes with sickle cell trait can participate in vigorous physical activities without complications, there is an increased risk of rhabdomyolysis and sudden death during strenuous exercise. Training regimens should be modified for athletes with sickle cell trait to reduce the risk of these complications.

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NUTRITION

Can an athlete be too obese to participate in sports?

Yes. If an obese athlete (>20% body fat for boys and men, >25% body fat for girls and women) has any other risk factors such as hypertension or poor cardiovascular conditioning, the athlete is at greater risk and should be evaluated fully and monitored closely.

What is the greatest risk for the obese athlete?

If no other risk factors are present, the greatest risk is the problem of **acclimatizing to heat.**

List 4 ways to manage acclimatization to heat.

1. Begin a conditioning program 2–3 weeks before the practice begins.
2. Increase physical activity gradually during a 14-day period.
3. Monitor preexercise and postexercise weight.
4. Take frequent breaks for fluids. (Water is best!)

Can a young person be too thin to participate in sports?

If the participant is physically mature (greater than Tanner stage 3), he or she should not be at greater risk for injury in a collision sport despite a difference in size from the other athletes unless the thinness is related to an eating disorder (see the following text).

What 2 groups may be at increased risk in sports?

1. Boys and young men with <7% body fat
2. Girls and young women with <12% body fat

List 6 factors for which they are at risk.

Decreased endurance, alterations in growth patterns, delayed pubertal development, abnormal or absent menstrual cycles (females), eating disorders, and osteoporosis

List 5 examples of sports in which the greatest risks occur.

Those in which weight loss tends to be encouraged, including wrestling, gymnastics, dance, cheerleading, and crew

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Are there safe ways to lose weight if it is indicated or desirable?

It is acceptable to burn excess fat to lose weight. A rate of **2 pounds a week** is acceptable. Weight loss accomplished by losing fluid (dehydration) or by losing muscle (malnutrition) can be harmful and can lead to poor endurance and reduced strength.

List 6 harmful effects of rapid weight loss.

Dehydration, malnutrition, electrolyte imbalance, negative nitrogen balance, and renal and liver insults

What are the calorie requirements of an athlete?

Enough calories to meet basic metabolic needs plus calories required for activity and exercise. **Prepubertal athletes** require additional calories for growth. Most growing, active athletes need 2,000–3,000 calories per day—and more if they are involved in vigorous activity and endurance sports. No athlete, even those trying to lose weight for wrestling or similar sports, should eat fewer than 1,500 calories per day.

What are the best foods for an athlete to eat?

A daily diet that offers plenty of water, 60% carbohydrates, 25% proteins, and 15% fat

What kind of vitamin supplements should an athlete take?

An athlete who eats a normal diet does not need additional vitamins.

How can an athlete gain weight if he or she desires?

A good caloric intake and appropriate weight training add "good" muscle weight to an athlete that can be a positive contribution to performance and appearance. (This is only true for athletes who have entered puberty.)

Are high-calorie and high-protein supplements safe for the athlete?

No. These may create potentially harmful renal solute loads and liver toxicity. Also, creatine and androstenedione should be avoided. Labeling of contents is not necessarily accurate.

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List 5 dangers of steroids.

Abnormal growth in prepubertal boys and girls; liver, skin, and CNS abnormalities; psychological disorders such as rage and depression; adrenal gland suppression; many long-term adverse effects

List 3 common types of substance abuse among athletes and the effects of each.

1. **Tobacco** can be detrimental to lung function and performance in sports, with no redeeming values. Smokeless tobacco is an even bigger problem in incidence and leads to cancer of the lip and mouth.
2. **Alcohol** can lead to nutritional and hydration problems and provides a large number of calories that may not be desirable.
3. **Marijuana** and **cocaine** have adverse effects on lung function and heart function.

HEAT INJURY

List, in order of severity, the 5 types of heat injury syndromes.

From mild to severe:

Heat fatigue, heat syncope, heat cramps, heat exhaustion, and heat stroke

List the symptoms of the 5 types of heat injury.

1. **Heat fatigue:** weakness, fatigue, light-headedness
2. **Heat syncope:** fainting, frequently at the end of a workout
3. **Heat cramps:** painful cramping of muscles, especially in the legs. Sometimes called "tired leg" or "dead leg" syndrome, heat cramps result in fatigue, weakness, and difficulty running.
4. **Heat exhaustion:** prostration, lethargy, hyperventilation, dizziness, inability to concentrate
5. **Heat stroke:** hyperpyrexia, shock, disorientation, or confused mental status up to level of coma

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List 6 ways the athlete can prevent heat injury.

1. Acclimatize.
2. Weigh before and after practice and replace weight loss by frequent and generous intake of water.
3. Wear lightweight clothing until acclimatized.
4. Use a sling psychrometer to measure "wet bulb temperature," humidity, and temperature, and regulate sports practice according to readings.
5. Be aware that lost sodium, potassium, and other salts and minerals are replaced in a normal diet. An exception may be endurance sports like cross-country, in which sports drinks (**NOT POWER DRINKS**) may be appropriate.
6. Watch for early signs of heat injury and allow generous fluid intake and cooldown.

List 5 components of treatment for heat injury.

1. Stop exercise.
2. Lie in cool place with legs elevated.
3. Drink lots of water if conscious enough to be safe.
4. Remove helmets or other restricting clothing items.
5. In case of heat stroke or severe heat exhaustion, IV fluids and other more aggressive emergency-room level treatments should be sought immediately.

What is the correct dose for salt tablets or potassium supplements?

No salt or potassium supplements are needed, even with leg cramps. Enough of these elements are provided by a normal diet. (See above exception for marathon and long-distance running, in which sports drinks should provide needed anions.)

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What should athletes drink?

Water is the ideal drink to replace lost fluid. Sports drinks can be too hypertonic during activity, and they can delay stomach emptying and suppress the thirst mechanism. Diluted sports drinks (2–3 parts water to 1 part sports drink) are acceptable. (See above for intense activities lasting more than 1 hour.)

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Chapter 14 Adolescent Medicine

PUBERTY AND GROWTH

What is adolescence?	The period between childhood and adulthood (the Latin *adolescere* means "to grow up"); generally from 10 to 21 years (exact limits vary)
What are the 5 tasks of adolescence?	Identity formation, autonomy, separation from family, exploration of vocation, and establishment of internal moral standards
What is puberty?	Biologic maturation (Latin *pubescere* means "to grow hair")
When does puberty occur?	8–13 years of age in girls; 9–14 years of age in boys
When is menarche?	10–15 years of age (average age is 12); ovulation usually occurs within 2 years after menarche
When does sperm production begin?	13–14 years of age
What are Tanner stages?	Stages of external physical (sexual) maturation. Also referred to as "Sexual maturity ratings" (SMRs)
What is Tanner stage 1?	Prepubertal
Stage 2?	Onset of any sign of sexual change; in girls: breast buds, sparse pigmented pubic hair; in boys: enlargement of testes, scrotum, and penis, with sparse pigmented pubic hair

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Stages 3 and 4?	Increasing pubic hair; other findings in girls include increased breast tissue, raised areola (stage 4); other findings in boys include continuing enlargement of genitalia
Stage 5?	Adult secondary sexual characteristics; areola now continuous with breast tissue in females, pubic hair on the inner thighs
What is precocious puberty?	Onset of puberty before 7 years of age in girls (6 years of age in African American girls) and before 9 years of age in boys
At what age is puberty considered delayed?	If there has been no development of secondary sexual characteristics by 13 (females) or 14 (males) years of age (Ch 24, p. 378)
List 3 categories of causes of precocious puberty.	1. Idiopathic (most common) 2. Endocrine abnormalities (see Ch 24) 3. CNS abnormalities
List 7 causes of delayed puberty.	Idiopathic (most common); Turner syndrome (in girls); Klinefelter syndrome (in boys) (Ch 30, p. 515); chronic illness; weight problems; CNS abnormalities, including secondary abnormalities; psychological or psychosocial problems
List 5 important factors in the evaluation of abnormal timing of puberty.	History, physical examination, plotting of longitudinal growth data, bone age, laboratory tests as clinically indicated (Ch 24, p. 375)
What information does one use to assess growth at puberty?	Longitudinal data
What are the normal growth rates?	Prepubertal: about 5 cm/yr During puberty: 9 cm/yr for girls; 10 cm/yr for boys (boys also have a "strength spurt" near the end of puberty)

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List 6 causes of a delayed growth spurt or strength spurt.

Idiopathic, stress, chronic illness, nutritional deficiencies, genetic disorders, endocrine disorders

LEGAL ISSUES

What is emancipation?

Fiscal and physical independence

What is the legal age of emancipation?

Usually 18 years of age; may vary with states (check local statutes)

List 6 types of procedures to which minors can legally consent.

In most states, minors can consent to:
1. Emergency care
2. Diagnosis, treatment, and prevention of STDs
3. Contraception—but not sterilization
4. Diagnosis and management of pregnancy
5. Management of rape or sexual abuse
6. Diagnosis and treatment of mental health problems, including substance abuse

Note: State laws vary and may change regarding these issues.

Who is an emancipated minor? (List 4 categories)

A minor who is currently or was married; a teenage parent; a self-supporting minor living away from home; a minor in the armed forces

BASIC ISSUES OF TEEN HYGIENE

When should a woman have her first pelvic examination?

By 18 years of age; before initiation of sexual intercourse; any time there is a gynecologic problem

List 3 immunizations that are given to adolescents.

1. Second MMR vaccine, varicella vaccine, or both (if not received earlier)
2. DTaP booster
3. Hepatitis B series (if not already received)

Note: Some physicians also suggest meningococcal vaccine, depending on the school setting. This is required by some colleges and universities.

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When is adult dentition present?	By midpuberty
How often is routine dental care needed?	Hygiene every 6 months; checkup yearly
How often are eye examinations needed?	Every 1–2 years during adolescence, more often if indicated

ACNE

See Chapter 12.

MENSTRUATION

List 3 characteristics of a normal menstrual period.	Menstrual flow <8 days; cycle duration of 21–35 days; blood loss of about 50 mL during menstrual flow

OLIGOMENORRHEA

What is oligomenorrhea?	Too little menstrual bleeding; skipping months
List 8 causes.	Anovulatory cycles, stress, pregnancy, weight change (loss or gain), polycystic ovary syndrome, thyroid disease, increased prolactin production, androgen excess
What is the treatment?	Regulate periods with medroxyprogesterone acetate (Depo-Provera) or oral contraceptives, and treat the underlying cause.
What are the 2 most common causes of secondary amenorrhea?	Pregnancy and stress. Other causes, as in oligomenorrhea

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DYSFUNCTIONAL UTERINE BLEEDING

What is dysfunctional uterine bleeding?	Excessive menstrual bleeding; menstrual flow lasting longer than 8 days, cycles that are unusually short (<20 days) or long (>40 days)
What are the causes?	Anovulatory cycles, pregnancy problems (e.g., ectopic, miscarriage), STDs, endocrine causes (similar to oligomenorrhea), diabetes, blood dyscrasias, iron deficiency
List 4 important factors in evaluation.	History, physical and pelvic examination, hematocrit, platelet count
What is the treatment?	Same as for oligomenorrhea

DYSMENORRHEA

What is dysmenorrhea?	Cramping, colicky pain immediately before or during menses
How common is it?	It is the most common cause of school absence for adolescent females.
List 7 associated symptoms.	Headache, irritability, emotional lability, nausea, vomiting, diarrhea, backache
What are the 3 important factors in the evaluation?	History, physical, and pelvic examinations
List 3 treatments.	Nonprescription analgesics, nonsteroidal anti-inflammatory drugs (NSAIDs), and oral contraceptives

SEXUALLY TRANSMITTED DISEASES

See Chapter 28.

How common is the sexual activity among teenagers?	Varies widely with the population—some studies show 50% of teens engage in sexual activity by 16 years of age and 75% by 19 years of age

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How common are STDs?	Present in 25% of sexually active teenagers
List 3 infestations that may be transmitted by close contact.	Pubic lice ("crabs"), body lice, and scabies
List 3 genital infections that are usually not sexually acquired.	*Monilia*, bacterial vaginosis, folliculitis
What is HPV?	Human papilloma virus
What are the clinical findings in HPV infection?	Genital warts; increased risk of cervical cancer
What is the current recommendation for HPV immunization?	A CDC advisory panel has recommended HPV vaccination for girls aged 11–25 years. (**Note:** This recommendation may be modified.)

PREGNANCY AND CONTRACEPTION

How common is teenage pregnancy?	About 750,000/yr in the United States; about 50% result in spontaneous or elective abortion and 50% result in live birth.
List 4 hazards teenagers face with pregnancy.	Dropping out of school, poor development of job skills, short- or long-term welfare dependency, parenting difficulties
What are the hazards for children of teenage parents?	Increased incidence of low birth weight, prematurity, health or psychosocial problems related to poverty or poor parenting skills, ADHD
List 3 components of managing teenage pregnancy.	Teen-oriented obstetric care; promoting the involvement of family and community; long-term follow-up and support after the birth of child

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List 4 ways teenage pregnancy can be prevented.

1. Education
2. Building teens' skills to enhance self-esteem, self-efficacy, and decision making
3. Understanding abstinence and delayed sexual intercourse as appropriate courses
4. Knowledge and availability of birth control

List 10 methods of birth control.

Abstinence, condom (with spermicide), oral contraceptives (birth control pills), diaphragm (with spermicide), rhythm method, progesterone implant (Norplant), female condom, medroxyprogesterone acetate (Depo-Provera), contraceptive ring, spermicide (alone)

EPIDEMIOLOGY OF ACCIDENTAL AND NONACCIDENTAL DEATH

What are the 3 leading causes of death among adolescents?

Accidents (particularly motor vehicle accidents), homicide, suicide

What segment of the adolescent population is at greatest risk for death from homicide?

African American males

Are suicide attempts (or gestures) more common in males or females?

Females

What adolescents are at greatest risk for death from suicide?

White males

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Are suicide attempts considered to be "acting out"?

All attempts or suicide gestures should be taken seriously and patients should be assessed and hospitalized under observation if they are felt to be at risk. Suicide attempt is frequently a sign of depression or other mental illness and these underlying psychiatric issues should be explored and treated.

Is there a relationship between gun availability and successful suicides?

Yes. Guns are the most commonly used method of successful suicide. Although the guns do not "cause" suicidal behavior, their use by a person with suicidal intent is associated with a high chance of death.

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Section IV Pediatric Diseases

Chapter 15 Hematologic Disorders

NUTRITIONAL ANEMIAS

IRON-DEFICIENCY ANEMIA

What is it?	Hemoglobin (Hgb) level below 95% of the normal for age that is caused by the lack of iron; it is the most common anemia.
What are the 2 causes?	Inadequate iron in diet or chronic blood loss (e.g., from peptic ulcer, inflammatory bowel disease, Meckel diverticulum, polyp, hemangioma, repeated heavy bleeding with menses)
What is the physiologic effect?	Decreased production of heme proteins involved in oxygen transport (Hgb), electron transport (cytochromes), and oxidative metabolism (NADH)
What are the common signs and symptoms?	Pallor, fatigue, shortness of breath, pica (ingestion of nonfood substances, such as ice, paper, dirt, or clay; Ch 19, p. 307), spoon-shaped nails (koilonychia), enlarged spleen. If not severe, iron-deficiency anemia is usually suspected from results of routine laboratory screenings.
What are the 4 findings of a CBC in iron-deficiency anemia?	Hypochromic, microcytic RBCs; decreased reticulocytes; decreased mean corpuscular volume (MCV); decreased mean corpuscular hemoglobin (MCH)

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What are the 4 effects on the indices of iron storage and transport?	Decreased iron, ferritin, and transferrin saturation; increased total iron-binding capacity
Why are free erythrocyte protoporphyrins (FEPs) increased?	Low iron concentration limits the production of Hgb. FEPs are heme precursors that accumulate as a result.
List 3 investigations that may be helpful to detect occult blood loss.	Stool guaiac test for occult blood; urinalysis; a careful menstrual history
How may iron-deficiency anemia be diagnosed if other tests and investigations are equivocal?	By the child's response to a trial of iron administration
List 2 treatments.	1. Oral therapy with elemental iron (3 mg/kg per day for 3–4 months); reticulocyte response should be seen in 1–2 weeks. 2. Determine and correct the etiologic factors of iron deficiency.
What are the potential side effects of oral iron administration?	Nausea, abdominal pain, constipation, stained teeth
What is the result if deficiency is not corrected?	Psychomotor and cognitive delays may result.

FOLATE-DEFICIENCY ANEMIA

What is it?	Decreased Hgb caused by folate deficiency. Folate deficiency causes a decrease in folate-dependent enzymes with decrease in 1-carbon transfer reactions.
What are the common causes?	Dietary deficiency is the most common. Others include increased folate demands (hemolytic anemia, sickle cell disease, pregnancy), thalassemias, HIV, malabsorption, certain drugs (e.g., antiepileptics, trimethoprim-sulfa, alcohol), and certain inborn errors of metabolism.

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What are the common signs and symptoms?

Pallor, fatigue, shortness of breath; infants may fail to gain weight and suffer from chronic diarrhea.

List 6 laboratory findings.

1. CBC: decreased Hgb and increased MCV
2. Hypersegmented neutrophils
3. Low reticulocyte count
4. Low serum folate
5. Elevated serum lactate dehydrogenase (LDH)
6. Nucleated RBCs with megaloblastic morphology

What is the treatment?

Oral supplementation with folate. Increased reticulocytes peak at 1 week and Hgb is normal in 6–8 weeks. Duration of therapy is based on the etiologic factors.

VITAMIN B_{12} DEFICIENCY (PERNICIOUS ANEMIA)

What is it?

Decreased Hgb caused by vitamin B_{12} deficiency

What is the physiologic effect?

Deficit of cobalamin, which is a required cofactor in conversion of homocysteine to methionine (via methionine synthetase). Methionine is critical in formulation of tetrahydrofolate.

List 4 causes.

1. Dietary deficiency (e.g., seen in vegans)
2. Decreased absorption in gastrointestinal (GI) tract (e.g., in a child whose terminal ileum has been removed)
3. Defects in B_{12} transport
4. Defects in B_{12} metabolism

List 5 characteristic laboratory findings.

Increased MCV, multilobed neutrophils, decreased serum B_{12}, increased serum methylmalonic acid, increased serum total homocysteine

What are the common signs and symptoms?

Nausea, diarrhea, abdominal pain, glossitis

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What are the neurologic sequelae of prolonged B_{12} deficiency?

Subacute degeneration of spinal cord, decreased vibratory and position sense, pyramidal signs, and peripheral neuropathy; cerebral symptoms and depression may be present.

What are the findings on bone marrow aspiration?

Megaloblastic changes

What is the Schilling test?

The Schilling test uses vitamin B_{12} incorporated with cobalt-57 (^{57}Co) to test for B_{12} absorption. The labeled B_{12} is given orally and is followed by a large IV dose of unlabeled B_{12}. If the labeled B_{12} has been absorbed, it will be displaced by the unlabeled B_{12} and excreted in the urine. If this phase suggests that labeled B_{12} was not absorbed, another oral dose of labeled B_{12} is given simultaneously with intrinsic factor (IF). If labeled B_{12} is then later excreted, the insufficient or poorly functioning IF is the cause for B_{12} deficiency. If labeled B_{12} is still not excreted, a primary malabsorption condition exists.

What is the treatment?

For deficiency without malabsorption or an IF defect, oral vitamin B_{12} is usually sufficient, unless rapid correction is desired, in which case injections are used as initial therapy. Malabsorption or IF defect usually requires vitamin B_{12} injections for life.

What are the indicators of appropriate response to the treatment?

Clinical improvement, increased reticulocytes, decreased MCV, and decreased methylmalonic acid

List 2 complications.

1. Rapid correction of severely anemic patients can be associated with thrombosis, embolism, and hypokalemia.
2. Neurologic symptoms secondary to vitamin B_{12} deficiency usually improve slowly and may not completely resolve.

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HEMOGLOBINOPATHIES

THALASSEMIA

What is it?

The thalassemias are a group of inherited anemias caused by gene mutations that affect the synthesis of Hgb chains. Clinical syndromes vary in severity.

Who is most commonly affected?

α-Thalassemia is more common in people of Asian and African ancestry.

β-Thalassemia is more common in people of Mediterranean and African ancestry.

What is the pathophysiology?

Decreased or absent synthesis of α or β chains leads to increased amounts of rare Hgb compared with normal Hgb. Severe forms of thalassemia lead to hemolysis and ineffective RBC production in the bone marrow.

How many α-globin genes are normally present?

Normally, there are **4** α-globin genes (2 on each homologous chromosome 16). Therefore, there can be different combinations of α-globin gene abnormalities.

List 4 types of α-thalassemia and their characteristics.

Silent carrier. (1/4 genes affected): hematologically normal; electrophoresis normal

Thal trait. (2/4 genes affected): mild anemia, decreased MCV, presence of target cells, electrophoresis normal

Hgb H disease. (3/4 genes affected): moderate hemolytic anemia, decreased MCV, presence of target cells, splenomegaly, electrophoresis reveals Hgb A and H (β_4)

HgB Barts. (4/4 genes affected): results in hydrops fetalis and fetal death, electrophoresis reveals Hgb H and Hgb Barts (γ_4)

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List 4 types of β-thalassemia and their characteristics.

β-Thalassemia is more heterogeneous. Severity is based on the specific mutation in a gene rather than the number of genes affected.

Silent carrier: hematologically normal, electrophoresis normal

Thal trait: mild anemia, decreased MCV and MCH, presence of target cells, electrophoresis reveals increased Hgb A_2 and F

Thal intermedia: severe anemia without transfusion requirement, decreased MCV and MCH, presence of target cells, electrophoresis reveals increased Hgb A_2 and F

Thal major: severe anemia with transfusion requirement, decreased MCV and MCH, presence of target cells, growth retardation, bone deformity, hepatosplenomegaly, electrophoresis reveals increased Hgb A_2 and F

List 3 common diagnostic studies for thalassemia.

CBC; Hgb electrophoresis; measurement of α and β chain biosynthesis

What are the treatments?

Mild syndromes?

Folic acid supplementation, avoidance of oxidant drugs, transfusion if necessary

Severe syndromes?

Folic acid supplementation, transfusion protocol with chelation of iron, splenectomy if hypersplenism develops, bone marrow transplantation

What are the common complications?

Cholelithiasis (Ch 20, p. 311), increased susceptibility to infection, bone marrow hyperplasias with bone deformity, Cooley's facies, "hair-on-end" skull radiograph; liver, endocrine, and cardiac abnormalities associated with iron overload

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SICKLE CELL DISEASE

What is it?

Hemoglobinopathy in which α chains are normal, but β chains are abnormal because valine is substituted for glutamic acid at position 6 of the β chain

Who is most commonly affected?

People of Central African, Mediterranean, and Indian descent, but sickle cell disease can be seen in any population

What is the pathophysiologic effect?

Hgb S forms polymers within red cells, causing sickling of the cells when Hgb is deoxygenated. This sickling causes sludging and obstruction in vessels with subsequent tissue hypoxia.

List 5 common phenotypes associated with hemoglobin S and their characteristics.

1. **Sickle trait:** Hgb AS; not associated with increased morbidity and mortality rates
2. **Sickle disease:** Hgb SS; clinically variable in severity from mild to debilitating
3. **Sickle SC disease:** Hgb SC; mild chronic hemolytic anemia with variability in complications from vaso-occlusion; splenomegaly
4. **Sickle β-thalassemia:** two forms, β^+ (Hgb A is produced) and β° (Hgb A is not produced); sickle β°-thalassemia (i.e., no Hgb A produced) is clinically similar to Hgb SS; splenomegaly
5. **Sickle α-thalassemia:** variable clinical picture

What 3 diagnostic studies are used, and what do they show?

1. CBC: decreased Hgb, normal MCV if not thalassemic
2. Blood smear: sickled forms, target cells, Howell-Jolly bodies
3. Hgb electrophoresis:

Sickle disease: 80–100% Hgb S, 0–20% Hgb F

Sickle SC disease: 50% Hgb S, 50% Hgb C

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Sickle β-thalassemia: 75–100% Hgb S, 0–20% Hgb F, 3–6% Hgb A_2

Sickle α-thalassemia: 80–100% Hgb S, 0–20% Hgb F

List 5 clinical manifestations of sickling hemoglobinopathies.

1. Vaso-occlusion causing pain in bone, hand and foot ("hand-foot syndrome"), abdomen, or chest. Patient may also experience cerebral vascular accident (CVA) or priapism.
2. Splenic sequestration
3. Aplastic crisis
4. Infection caused by decreased opsonins and splenic function. **Infection is the most common cause of death in children with sickle cell disease.** Common organisms include pneumococci, *Haemophilus influenzae*, *Salmonella*, and *Mycoplasma*.
5. Acute chest syndrome

What is acute chest syndrome?

A serious complication of sickle cell disease. It is one of the most common causes of death in these patients.

List 3 causes.

Infection, pulmonary fat emboli, and pulmonary infarction. However, many episodes do not have an identifiable cause.

List 6 symptoms.

The symptoms may include cough, wheezing, fever, chest pain, extremity pain, and dyspnea.

What are the treatments?

Prophylaxis (vaccines against influenza virus, *H. influenzae*, *S. pneumoniae*)

For clinical cases: hospitalization for antibiotics, bronchodilators, supplemental oxygen, transfusions

Bronchoscopy should be considered if there is no clinical response to initial therapy.

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What are the treatments for the following clinical manifestations?

For vaso-occlusive crisis?	Hydration and pain medications
For CVA?	Exchange transfusion with chronic transfusion protocol to keep Hgb S < 30%
For splenic sequestration?	Emergent transfusion with subsequent splenectomy
For aplastic crisis?	Supportive care; transfusion if necessary
For infection?	Appropriate antibiotics, penicillin prophylaxis, vaccines for *S. pneumoniae*, *H. influenzae*, *N. meningitidis*

What are the common progressive complications?

Cardiovascular?	Cardiomegaly and cardiomyopathy secondary to iron overload and chronic anemia
Pulmonary?	Progressive disease with infarcts and infections
Hepatic?	Cholecystitis, hepatitis
Renal?	Hematuria, hyposthenuria, glomerular and tubular fibrosis
Ophthalmologic?	Retinopathy
Skeletal?	Codfish vertebrae, aseptic necrosis

HEMOLYTIC ANEMIAS

GLUCOSE-6-PHOSPHATE DEHYDROGENASE DEFICIENCY

What is glucose-6-phosphate dehydrogenase (G6PD)?

G6PD is a dehydrogenase involved in the pentose phosphate pathway, which is important in NADPH production. It converts glucose-6-phosphate to 6-phosphoglucono-δ-lactone. It maintains NADPH, which in turn maintains reduced glutathione that cleans up free radicals in cells.

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What causes its deficiency?	Mutation in the G6PD gene on the X chromosome. G6PD deficiency is an X-linked recessive disorder.
What is the physiologic effect of G6PD deficiency?	In the RBC that is deficient in G6PD, oxidative stress depletes NADPH and oxidizes glutathione with subsequent oxidative damage to the cell membrane, leading to hemolysis.
What are the common signs and symptoms?	An asymptomatic child with G6PD deficiency usually has normal hematologic variables, but when faced with oxidative stress, the child may have jaundice, hemoglobinuria, splenomegaly, and anemia.
What are some oxidative triggers?	Fava beans, infections, and certain drugs (e.g., sulfas, antimalarials, analgesics)
What diagnostic study is used?	Measurement of G6PD activity in RBCs of reticulocyte-poor blood
What is the treatment?	Usually supportive; with severe hemolysis, transfusion is occasionally necessary.

SPHEROCYTOSIS

What is it?	Autosomal dominant congenital hemolytic anemia with spherical RBCs
What is the pathophysiologic effect?	Loss of membrane surface area as a result of deficiencies in some RBC proteins (e.g., spectrin, ankyrin) leads to spherically shaped, less deformable cells, which become subject to lysis and trapping in the spleen.
List 4 signs and symptoms.	Symptoms (mild or severe) include anemia, hemoglobinuria, jaundice, and splenomegaly. Hemolysis increases with infections.

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List 3 diagnostic studies.

1. CBC: Hgb is decreased.
2. Blood smear, which shows increased spherocytes and reticulocytes
3. Osmotic fragility test

What are the 2 treatments?

1. **Supportive** usually, if the clinical course is mild
2. **Splenectomy,** with or without cholecystectomy, if severe anemia, recurrent significant hemolysis, biliary colic, or cholecystitis exists. Splenectomy is usually delayed until after age 6 if possible.

List 3 common complications.

Aplastic crisis; biliary colic or cholecystitis; dependence on blood transfusions to maintain acceptable RBC levels

APLASTIC ANEMIA

What is it?

Pancytopenia secondary to decreased production of blood cells because of destruction of stem cells in bone marrow or because of abnormal bone marrow environment; may be inherited or acquired

List 2 classifications and their characteristics.

1. Severe: granulocyte count < 500, platelet count < 20,000, reticulocyte count < 1% after correction for Hgb; hypocellular bone marrow on biopsy
2. Mild or moderate: mild to moderate cytopenia; normal or increased bone marrow cellularity

What are some causes?

Acquired? (List 5)

Drugs, radiation, viral infections, preleukemia, paroxysmal nocturnal hemoglobinuria

Inherited? (List 4)

Fanconi anemia (see p. 180), congenital dyskeratosis congenita, Shwachman-Diamond syndrome, myelodysplasia

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List 4 signs and symptoms.

Fatigue, pallor, increased bleeding, and infections

List 5 diagnostic studies.

1. CBC: shows pancytopenia and decreased reticulocytes
2. Bone marrow biopsy: shows hypocellularity

The following studies may also be considered:

3. Viral titers
4. Screening for paroxysmal nocturnal hemoglobinuria
5. Chromosome breakage studies for Fanconi anemia

What are the 4 methods of treatment?

1. Bone marrow transplantation—if a related matched donor is available, transplantation is the primary therapy.
2. Immunosuppressive therapy (e.g., antithymocyte globulin, cyclosporine, steroids)
3. Hematopoietic growth factors
4. Supportive care with antibiotics and transfusion therapy

What is the prognosis?

Prognosis is poor without bone marrow transplantation. **Pretransplantation transfusions should be avoided as much as possible.**

GAUCHER DISEASE

What is it?

Inherited storage disease with deficiency of the enzyme **glucocerebroside β-glucosidase**

How is it inherited?

As an autosomal recessive disorder

What is the physiologic effect?

Accumulation of glucocerebroside in reticuloendothelial (Gaucher) cells

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List 3 types and their characteristics.

1. Chronic nonneuronopathic (adult form): The clinical course is variable and may include anemia, thrombocytopenia with marrow infiltration, bleeding tendency, hepatosplenomegaly, bone pain, and aseptic necrosis of bones.
2. Acute neuronopathic (infantile form): Severe, with presentation in infancy; CNS infiltration with neural defects and hepatosplenomegaly; death usually occurs by 2 years of age.
3. Subacute neuronopathic (juvenile form): Neurologic defects occur later in the course of disease and may increase after splenectomy.

In what ethnic group is the adult form commonly found?

Ashkenazi Jews

List 4 results that diagnostic studies show.

1. Bone marrow biopsy shows Gaucher cells.
2. Decreased lysosomal β-glucocerebrosidase in leukocytes or cultured skin fibroblasts
3. Increased acid phosphatase
4. Increased angiotensin-converting enzyme

List 3 treatments for nonneuronopathic Gaucher disease.

1. Enzyme replacement therapy
2. Splenectomy if hypersplenism exists, but this may increase other symptoms (now only rarely performed)
3. Bone marrow transplantation

POLYCYTHEMIA

What is it?

RBC count, Hgb level, and total RBC volume all exceed the upper limits of normal. In postpubertal children, it is distinguished by Hgb $>$ 16 g/dL and a total RBC mass $>$ 35 mL/kg.

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What is the appropriate term for high Hgb level with a concurrent decrease in plasma volume (e.g., as occurs in acute dehydration and burns)?	Hemoconcentration
What is polycythemia rubra vera?	This is a **primary myeloproliferative polycythemia.**
List 3 diagnostic criteria of polycythemia rubra vera.	Increased total RBC volume, Hgb, and hematocrit; arterial oxygen saturation $\geq$ 92%; splenomegaly
List 5 laboratory findings.	Thrombocytosis, leukocytosis, increased leukocyte alkaline phosphatase, increased vitamin B_{12} or unsaturated B_{12}-binding capacity
List 2 treatments.	Phlebotomy and chemotherapy
List 2 long-term risks.	Myelofibrosis and acute leukemia
What is the prognosis?	Poor
What is secondary polycythemia?	Polycythemia as a result of other inciting causes
List 6 of these causes.	1. Hypoxia 2. Hemoglobinopathies 3. Neonatal conditions, such as twin-twin transfusion or maternal hemorrhage, being the infant of a diabetic mother, intrauterine growth retardation, neonatal thyroid toxicosis, adrenal hypoplasia, trisomy 21 4. Benign and malignant tumors that secrete erythropoietin 5. Excess presence of anabolic steroids caused by either adrenal disease or excessive administration of anabolic steroids 6. Familial

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GRANULOCYTE DISORDERS

What is leukocyte adhesion deficiency?

It is a deficiency of a β_2-integrin. The condition is autosomal recessive and results in the deficiency of leukocyte adhesion to offending agents.

List 3 clinical manifestations.

Leukocytosis, delayed separation of the umbilical cord, and bacterial infections

What is Chédiak-Higashi syndrome?

An autosomal recessive disorder affecting granule-bearing cells. Granulocytes and melanocytes are characteristically affected.

List 3 ways in which granulocytes are affected.

Defects in chemotaxis, degranulation, and bactericidal activity

List 3 clinical manifestations.

Oculocutaneous albinism; large neutrophil granules; and recurrent bacterial infections

What is chronic granulomatous disease (CGD)?

A genetically heterogeneous condition that results in a defect in "respiratory burst" in leukocytes

What is the respiratory burst?

It is a reaction catalyzed by NADPH oxidase that forms hydrogen peroxide and hydroxyl radicals, which are thought to play a key role in killing microbes.

What is the clinical manifestation of CGD?

Recurrent bacterial and fungal infections

List 3 treatments.

There is no cure. Trimethoprim-sulfamethoxazole prophylaxis may limit infections. γ-Interferon or bone marrow transplantation may help some patients.

What is the definition of neutropenia?

Absolute neutrophil count (ANC) $< 1{,}000$

What is cyclic neutropenia?

The neutrophil count cycles between a normal and low ANC.

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What is autoimmune neutropenia?

A condition resulting from the presence of antineutrophil antibodies. Usually found in infants with no predisposing cause. May also be seen after transplacental transfer of maternal IgG (neonatal alloimmune neutropenia) or in neonates whose mothers have an autoimmune disease (neonatal maternal autoimmune neutropenia)

Can infection induce neutropenia?

Yes

List 4 types of infections that most commonly cause neutropenia.

Viral infections are the primary cause. Bacterial, mycobacterial, and rickettsial infections may also cause neutropenia.

What is Kostmann syndrome?

This is a rare autosomal recessive condition associated with neutropenia at birth.

What is the cause of this disease?

Unknown

What is the clinical manifestation?

Severe, often fatal, infections

What is Schwachman-Diamond syndrome?

An autosomal recessive condition characterized by neutropenia and pancreatic insufficiency. Chemotaxis is also defective in these neutrophils.

List 5 clinical manifestations.

Pancreatic insufficiency, potential growth failure, dry skin, eczema, and ichthyosiform lesions

For which malignancy are these patients at risk?

Leukemia

List 5 treatment options for neutropenia conditions.

1. Judicious use of antibiotics to either treat or prevent serious infection
2. Steroids may be used in autoimmune neutropenia.
3. Granulocyte colony-stimulating factor (G-CSF) may be used in some neutropenic conditions.

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4. γ-Interferon may be useful in CGD.
5. Bone marrow transplantation may be used in Chédiak-Higashi syndrome, CGD, Wiskott-Aldrich syndrome, and leukocyte adhesion deficiency.

PLATELET DISORDERS

CONGENITAL PLATELET DISORDERS

What is Wiskott-Aldrich syndrome?

Thrombocytopenia, purpura, eczema, and an increased susceptibility to infection as a result of impaired humoral immune responses and chemotaxis of neutrophils. This syndrome is an X-linked recessive trait.

What causes the thrombocytopenia?

Uncertain—probably an intrinsic platelet abnormality or defective formation or release of platelets. However, the number of megakaryocytes is normal.

List 3 treatment options.

Splenectomy improves platelet count (postsplenectomy sepsis is a risk); administration of transfer factor; bone marrow transplantation

List 3 long-term risks of Wiskott-Aldrich syndrome.

Infections; bleeding; malignancy (about 12% of patients develop malignancies, including leukemic lymphoreticular malignancies)

What is TAR syndrome?

Thrombocytopenia associated with **a**plasia of the **r**adii. There may also be cardiac and renal anomalies. The thumbs are usually normal.

What is the clinical manifestation of thrombocytopenia?

Hemorrhage, which may be evident even in the first days of life (e.g., during circumcision)

List 3 laboratory findings.

Thrombocytopenia; normal Hgb; and possibly leukocytosis

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List 2 findings of bone marrow aspirate.

Megakaryocyte count is normal; nuclear morphology may be abnormal.

What is Fanconi anemia?

A hypoplastic or aplastic anemia characterized by pancytopenia with associated skeletal, solid organ, and skin abnormalities

What is the cause?

Autosomal recessive inherited condition

What are its clinical manifestations?

Usually pancytopenia beginning at 3–4 years of age, which can lead to bleeding and infection. Other manifestations include hyperpigmentation, skeletal abnormalities (including absent or hypoplastic thumbs), short stature, and other anomalies. Skeletal findings may be subtle in some patients.

List 4 major risks of Fanconi syndrome.

Hematologic malignancy, infections, bleeding, solid organ (especially liver) failure

What are the bone marrow findings?

Aplasia (similar to that seen in acquired aplastic anemia). The bone marrow may be normal if there is no pancytopenia.

What are the treatment options?

Steroids and androgens (relapse occurs in 50% of patients); G-CSF; bone marrow transplantation

What is the prognosis?

Poor. The median survival age is about 20 years.

What is Kasabach-Merritt syndrome?

A condition of platelet trapping and consumptive coagulopathy associated with congenital hemangioma, usually of the liver

What is the pathophysiology?

Trapping and destruction of platelets within the extensive vascular bed of the hemangioma

What are the peripheral blood smear findings?

Thrombocytopenia with RBC fragments

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What are the bone marrow findings?

Normal megakaryocytes

What is the clinical manifestation?

Spontaneous hemorrhage

List 5 treatment options.

1. Administration of steroids
2. γ-Interferon
3. Occlusion of hepatic artery (liver hemangioma)
4. Resection or compression of hemangioma, although this might result in uncontrollable hemorrhage
5. Radiation to the hemangioma

What is hemolytic-uremic syndrome (HUS)?

A clinical syndrome of microangiopathic hemolytic anemia, thrombocytopenia, and acute renal failure (Ch 19, p. 285, for more on HUS)

What is thrombotic thrombocytopenic purpura?

A condition characterized by thrombocytopenia and hemolytic anemia

List 2 of the major clinical manifestations.

1. Hemorrhage
2. Neurologic sequelae—may include aphasia, blindness, and convulsions as a result of embolism and thrombosis of small blood vessels of the brain

List 3 treatment options.

1. Plasmapheresis and plasma infusions (effective in 60–70% of cases)
2. Steroids
3. Splenectomy if condition is refractory to the above therapies

List 5 common drugs that may cause drug-induced thrombocytopenias in children.

Carbamazepine (Tegretol), phenytoin, sulfonamides, trimethoprim-sulfamethoxazole (Bactrim), chloramphenicol

IDIOPATHIC THROMBOCYTOPENIC PURPURA

What is it?

Development of platelet antibodies with subsequent destruction of platelets

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What is the peak age for ITP?

2–4 years

In whom is it most commonly seen?

Previously healthy children, often after viral illness. It can be associated with autoimmune disease and HIV.

List 4 characteristic laboratory and diagnostic findings.

1. Platelet count < 50,000/mm^3
2. Sparse, large platelets on blood smear
3. Bone marrow aspirate shows an increased number of megakaryocytes, usually of immature forms
4. Antiplatelet immunoglobulins are present.

List 3 treatment options for severe ITP.

Steroids; IV immunoglobulin; possibly, splenectomy (if ITP is chronic). Platelet transfusions are usually not helpful but may be used in conjunction with other therapy in episodes of serious bleeding or in surgery. If ITP is not severe (platelet count > 20,000), observation is prudent.

List 3 complications of ITP.

Severe GI hemorrhage; severe CNS hemorrhage; hematuria

What is the prognosis?

Majority of childhood ITP is benign and self-limited; 10–15% of patients develop chronic ITP.

COAGULATION DEFECTS

HEMOPHILIA A AND B

What is hemophilia A, or classic hemophilia?

Factor VIII deficiency

What is hemophilia B, or Christmas disease?

Factor IX deficiency

What is the pathophysiology of these conditions?

Both are X-linked recessive disorders with decreased production of factor VIII or IX, respectively. There is a moderately high spontaneous mutation rate.

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How is the severity of hemophilia A classified?	By the percentage of factor VIII present: severe, <1%; moderate, 1–5%; mild, >5%
What is the physiologic result?	Inability to generate normal fibrin
What are the common signs and symptoms?	1. Bleeding, including neonatal bleeding (especially with circumcision) or ICH; oral, muscular, or joint bleeding 2. Easy bruising and bleeding with mild trauma
List 3 components of the diagnosis.	1. Family history 2. Prolonged partial thromboplastin time (PTT); bleeding time is usually normal, except in very severe cases 3. Decreased level of factor VIII (hemophilia A) or IX (hemophilia B)
What is the treatment?	Replacement therapy with the deficient factor: recombinant factor VIII and recombinant factor IX are now available; DDAVP may be useful in patients with mild hemophilia.
How much does 1 unit/kg of factor VIII raise the patient's plasma factor VIII?	1 unit/kg of factor VIII will give a 2% rise in plasma factor VIII.
How much factor is appropriate for:	
Mild to moderate hemorrhage?	Achieve a factor level of **30–40%.**
Major surgery or a life-threatening bleeding episode?	Achieve **100%** and maintain for **7–14 days.**
Oral bleeding?	Antifibrinolytics (e.g., aminocaproic acid) may also be used.

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List 4 complications of these deficiencies.

1. Joint damage from repeated episodes of joint bleeding
2. Serious hemorrhage
3. Development of factor inhibitors (usually seen in factor VIII deficiency)
4. Infections (e.g., HIV, hepatitis) from factor replacement

VON WILLEBRAND DISEASE

What is it?

A disorder of von Willebrand factor (vWF) protein production; several variants are known, based on laboratory tests and platelet count. In the most severe form, there is an undetectable level of vWF and a decreased level of factor VIII. Most cases are caused by mutation of a single copy of the gene. Therefore, it can be inherited as an autosomal dominant trait. However, some patients are homozygous, so autosomal recessive inheritance is sometimes seen.

What is the physiologic result?

Inability of platelets to adhere to damaged endothelium

How common is von Willebrand disease?

It is **probably the most common inherited bleeding disorder.** The actual prevalence is difficult to determine because of its clinical variability.

List 3 signs and symptoms.

1. Easy bruising and bleeding with or without trauma
2. History of recurrent epistaxis
3. History of recurrent menorrhagia

Clinical severity may vary. Some affected persons may be asymptomatic.

List 5 components of the diagnosis.

1. Bleeding time is prolonged.
2. PTT may be increased.
3. A decrease in von Willebrand antigen, ristocetin cofactor, and factor VIII levels
4. Platelet count may be decreased in certain variants.
5. Normal-to-abnormal vWF multimers

Testing often has to be repeated to assure diagnosis.

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List 3 treatments.

1. DDAVP can be used to increase the vWF in some types of the disease.
2. Cryoprecipitate or certain factor VIII concentrates (e.g., Humate-P) can be used in DDAVP failure, in a severe bleeding episode, or in major surgery.
3. Patients with oral bleeding may also benefit from antifibrinolytics (e.g., α-aminocaproic acid).

DISSEMINATED INTRAVASCULAR COAGULATION

What is it?

Consumptive coagulopathy that activates the plasma coagulation system and depletes clotting and antithrombotic factors as well as platelets

What is the physiologic effect?

Cycle of intravascular thrombosis and fibrinolysis, particularly in small vessels

What are some common etiologic factors?

Sepsis, malignancy (especially promyelocytic leukemia), obstetric complications, extensive tissue damage from trauma, burns, hypoxia, snakebites

List 3 signs and symptoms.

Bleeding or clotting; embolic signs; oozing from vascular access or phlebotomy sites

What are the diagnostic findings?

Decreased platelet count, prolonged prothrombin time (PT) and PTT, decreased fibrinogen, increased fibrin split products

What are the 2 approaches to treatment?

1. Successful treatment is possible only with correction of underlying etiologic factors.
2. Symptomatic treatment includes transfusion with platelets, fresh-frozen plasma (FFP), and cryoprecipitate. Heparin may be used at times if thrombosis is a prevalent symptom.

What are the significant complications?

Severe bleeding or thrombosis in the GI, pulmonary, and CNS systems

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HEPARIN-INDUCED THROMBOCYTOPENIA (HIT)

What is it?	The development of thrombocytopenia from the administration of heparin
Mechanism?	The immune system forms antibodies (usually IgG) to platelets bound to the protein platelet factor 4 (PF4). Blood clots form resulting in a fall in platelet count.
Common presenting sign?	Most common sign is a fall in platelet count.
Symptoms?	Most patients are asymptomatic. However, symptoms may be of a general systemic nature or are the result of arterial and venous thrombosis. They include fever, chills, hypertension, tachycardia, chest pain, tachypnea, stroke, myocardial infarction, limb ischemia, and pulmonary embolism.
How is the diagnosis made?	1. Low platelet count 2. ELISA test for circulating antibodies (may be nonspecific) 3. Serotonin release assay is used in patients with a positive ELISA.
What other studies may be important?	Doppler ultrasound to check for deep venous thrombosis of legs
Treatment?	Alternative anticoagulation (e.g., lepirudin, argatroban). **Warfarin is contraindicated as it may cause "warfarin necrosis" (a type of skin necrosis).**

NEONATAL ALLOIMMUNE THROMBOCYTOPENIA

What is it?	Severe thrombocytopenia in infants secondary to having different platelet antigens from the mother, with subsequent platelet destruction by maternal antiplatelet antibodies—similar to Rh sensitization in blood groups

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Is there a high risk of bleeding?

Yes. It can sometimes occur prenatally.

How is it diagnosed?

Platelet typing of mother and father

List 3 treatments.

1. Transfusion with irradiated maternal platelets
2. Steroids prenatally or postnatally
3. Possible IV immunoglobulin

What are "hypercoagulable states"?

Conditions that predispose to blood clotting

Give 7 examples.

Examples include DIC (see p. 185), protein C deficiency, protein S deficiency, factor V Leiden, antithrombin III deficiency, homocystinuria (attributable to cystathionine synthase deficiency), and homocystinemia (attributable to methylene tetrahydrofolate reductase deficiency).

How are these diagnosed?

DIC should be excluded (by platelet count, D-dimer assay, peripheral smear, and a search for associated conditions). There is no simple screening test for the genetic causes of hypercoagulability. These require specific tests.

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Chapter 16 Pediatric Cardiology

INTRODUCTION TO PEDIATRIC CARDIOLOGY

GENERAL CONSIDERATIONS

What is the incidence of CHD?

Almost 1% of children are born with CHD.

What is cyanosis?

Blue discoloration of the skin and mucous membranes; not to be confused with acrocyanosis (blue coloration of the hands and feet, a normal finding in newborns)

How is cyanosis affected by the hemoglobin level?

Patients with a normal hemoglobin level will appear cyanotic at a saturation of 75%. Anemic patients will not appear cyanotic until an oxygen saturation of 50%, whereas polycythemic patients will appear cyanotic at a saturation of 85%.

What is clubbing?

The appearance of the fingers and toes of being bulbous and rounded, with a loss of the angle between 2 distal interphalangeal joints

What are the primary diagnostic tests used in pediatric cardiology?

1. Electrocardiography
2. Echocardiography
3. Cardiac catheterization

What is echocardiography?

Cardiac ultrasound; besides history and physical exam, this is the main diagnostic tool used by pediatric cardiologits.

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What is congestive heart failure?

A syndrome of decreased systemic cardiac output; this term may be used in patients with severe left ventricular dysfunction due to a dilated cardiomyopathy, and also for infants with a large VSD, a large left-to-right shunt, and excessive pulmonary blood flow but normal left ventricular systolic function

What is a shunt?

Any abnormal mixing of blood, whether congenital or created surgically. A left-to-right shunt occurs when the blood from the left side of the heart enters the right side, for example, small VSD in a child, and a right-to-left shunt is the converse, for example, VSD shunting in Eisenmenger syndrome.

What is a Blalock-Taussig (BT) shunt?

Originally, it was a connection made by end-to-side anastomosis of the subclavian artery to the PA; a modified BT shunt consists of an artificial conduit sutured from the innominate artery to a branch PA.

What is pulmonary banding?

A palliative procedure whereby the surgeon places a ligature around the main PA to reduce the pulmonary blood flow

What is a Damus-Kaye-Stansel (DKS) operation?

The anastomosis of the main PA and ascending aorta; this is done to treat severe subaortic stenosis while permitting perfusion of the coronary arteries; a source of pulmonary blood flow must be provided, for example, a BT shunt.

What is the Norwood procedure?

This operation treats hypoplastic left heart syndrome; it consists of a DKS operation, atrial septectomy, and placement of a shunt to provide pulmonary blood flow (either a BT shunt or an RV-to-PA [Sano] conduit).

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What is the Rastelli procedure?

For lesions with pulmonary atresia or severe pulmonary stenosis (PS) and a VSD, for example, TOF/pulmonary atresia, D-transposition of the great arteries (D-TGA)/VSD/pulmonary stenosis; the LV is baffled across the VSD to the aorta and a conduit is placed from the RV to the branch PAs; if a native PA is present, it is ligated.

What is the Glenn procedure?

The connection of the SVC to the PA; this can be bilateral or right or left; it is bidirectional; the "classic" Glenn is the right SVC to the right PA, which is disconnected from the left PA; commonly done as the "2nd stage" of single ventricle palliation, often at 6 months of age

What is the Fontan procedure?

The connection of the inferior vena cava to the PA; usually done with a Dacron tube; can be done as a "lateral tunnel" within the RA; can be "fenestrated" with a connection from the tube to the atrium; done as the "3rd stage" of single ventricle palliation, commonly at 3 years of age

CONGENITAL HEART DEFECTS

ATRIAL SEPTAL DEFECT

What is it?

A hole in the septum between the RA and the LA (5–10% of all CHD)

What are the 3 types?

Secundum ASD (most common); primum ASD (usually associated with atrioventricular septal defect [AVSD]); superior or inferior sinus venosus ASD

What is the physiologic result?

Left-to-right shunt with overload of the RA and RV

List 2 factors that determine the volume of the shunt.

1. The size of the defect
2. The compliance of the RV

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What are the symptoms?	Usually asymptomatic in children; it can cause right heart failure and/or pulmonary hypertension in the third to fifth decades of life.
List 2 physical signs or symptoms.	Murmur from increased blood flow through the pulmonary valve; fixed split of S_2
List 3 diagnostic studies.	ECG (may show right ventricular hypertrophy); echocardiography; cardiac catheterization if complicated defect or interventional closure planned
What is the treatment?	Device closure done in the cardiac catheterization laboratory for secundum defects, or surgical closure
What are the main complications of the repair?	Arrhythmias, pericardial effusion

VENTRICULAR SEPTAL DEFECT

What is it?	A hole in the interventricular septum (20% of all CHD)
List the 4 types.	1. Muscular 2. Perimembranous 3. Supracristal 4. Inlet (see atrioventricular septal defect)
What is the physiologic result?	Left-to-right shunt, which increases the volume of the blood going to the lungs
List the 2 factors that determine the volume of the shunt.	Size of the hole and resistance in the lung vasculature
What are the signs or symptoms of a small VSD?	No symptoms, with a loud holosystolic murmur
List typical signs or symptoms of moderate or large VSD?	Tachypnea, poor feeding, failure to thrive, pulmonary infections, with a soft holosystolic murmur and diastolic rumble

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What is the treatment for a small VSD?	Possibly none needed—at least 50% of VSDs close spontaneously
For a moderate or large VSD?	May require diuretics and a special feeding regimen until surgical repair; child will usually be allowed to grow as much as possible since the defect could become smaller. Requires patch closure, usually done when the child is under 12 months of age; some muscular defects can be closed by device placement in the catheterization laboratory.
List 3 common complications of surgical repair.	Residual VSD; aortic insufficiency (AI); complete heart block

COMPLETE AVSD

What is it?	A type of CHD resulting from the failure of the central portion of the heart to develop, resulting in an inlet VSD, a primum ASD, and a common atrioventricular valve
What are the 2 other names for this defect?	1. Atrioventicular canal defect 2. Endocardial cushion defect
With what condition is an AVSD commonly associated?	Down syndrome (trisomy 21)
What is the physiologic result of complete AVSD?	Left-to-right shunt at the AVSD, with pulmonary congestion and elevation of PA pressure
What are the 3 symptoms of a complete AVSD?	Failure to thrive; CHF, caused by pulmonary overcirculation; cyanosis may be present at birth until pulmonary vascular resistance drops
What will the ECG show?	Superior axis (negative deflection in aVF)

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What is the treatment?	Medical management with diuretics, high-calorie formula/fortified breast milk until an operation is done. Surgical treatment: patch closure of the primum ASD and inlet VSD, and division of the common AV valve into separate left-sided and right-sided AV valves
What are the most common complications of repair of a complete AVSD?	Mitral insufficiency, mitral stenosis, complete heart block

PARTIAL AVSD

What is it?	Primum ASD; cleft mitral valve; no VSD
What is the physiologic result?	Left-to-right shunt
What are the potential signs and symptoms?	Murmur from mitral regurgitation (MR) or pulmonary flow murmur
What will the electrocardiogram show?	Left axis deviation
What is the primary diagnostic tool?	Echocardiography
What is the treatment?	Surgical patch closure of the primum ASD and closure of the mitral cleft
What is a possible complication?	Residual MR

TRANSITIONAL AVSD

What is it?	In-between form of partial and complete AVSD; there is a VSD but it is small and restrictive.
What is the physiologic result?	Left-to-right shunt

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What is the treatment?	Patch closure of the ASD and VSD, closure of the mitral cleft
What are the possible complications?	MR, complete heart block

PATENT DUCTUS ARTERIOSUS

What is it?	Persistent patency of the ductus arteriosus (5–10% of all CHD)
What group of infants is at high risk for PDA?	Premature infants—as many as 80% of infants of 28 weeks' gestation have a PDA.
What is the physiologic result?	Left-to-right shunt from the aorta into the PA and back to the LA
What are the symptoms?	Premature infants may have respiratory distress, pulmonary hemorrhage, and apnea. Usually asymptomatic in older children
What are the physical exam signs?	Premature infants can have bounding pulses, widened pulse pressure, a ventricular heave, a systolic murmur; older children can have a continuous, "machinerylike" murmur at the left clavicle.
What is the treatment?	In a premature infant, first-line therapy is a prostaglandin inhibitor such as indomethacin. Second-line treatment is surgical duct ligation. In older children, the PDA can be occluded in the catheterization laboratory with vascular coils or plugs.
What is a complication of repair?	Residual shunt

COARCTATION OF THE AORTA

What is it?	A narrowed area of the aortic arch at the level of the ductus arteriosus or the ligamentum arteriosum (8% of all CHD)

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How do children present?

Newborns can present in cardiogenic shock; older children can present with hypertension, a murmur, or absence of femoral pulses.

What can be found on the chest x-ray?

1. The "3" sign of dilated ascending aorta, coarctation segment, and dilated descending aorta
2. **Rib notching** from dilated intercostal vessels

Which medication is used in a newborn with a ductal-dependent coarctation?

Prostaglandin

What is the most commonly used surgical treatment for coarctation?

Extended end-to-end anastomosis via thoracotomy

How can coarctation be treated in the catheterization lab?

1. Balloon angioplasty
2. Stent placement

What are the complications of repair?

Early: Residual obstruction; hypertension; chylothorax; recurrent laryngeal nerve damage

Late: Recoarctation, aneurysm

List 3 commonly associated cardiac anomalies.

Bicuspid aortic valve (50%); VSD; mitral valve stenosis

AORTIC STENOSIS

What is it?

Obstruction to flow across the aortic valve; is due to a hypoplastic annulus, thickened and abnormal leaflets, or both (5% of all CHD)

What is the physiologic result?

Obstruction causes abnormally increased left ventricular myocardial oxygen demand, which can lead to exercise intolerance, chest pain, syncope, sudden cardiac death, or heart failure.

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What are some physical signs?

A harsh murmur heard best at the right upper sternal border with radiation to the neck; a thrill; an ejection click

What will the ECG show?

Left ventricular hypertrophy with strain, but it can be normal

What will the echocardiogram show?

A thickened, doming aortic valve, possibly with a small annulus; Doppler echo will show an increased velocity of blood flow across the valve; there may be left ventricular hypertrophy or dysfunction.

What is the role of cardiac catheterization?

The pressure gradient across the valve can be directly measured and cardiac output can be determined; if the gradient is severe, balloon angioplasty can be performed.

What are the options for surgical repair?

1. Valvuloplasty
2. Valve replacement (homograft, mechanical valve, porcine valve)
3. Autograft valve (Ross procedure)

What are the possible complications?

Residual stenosis, valve insufficiency, left ventricular dysfunction, complete heart block

PULMONARY STENOSIS

What is it?

Obstruction of the blood flow through the pulmonary valve due to valve thickening and/or annular hypoplasia (5% of all CHD)

What is the physiologic result?

High right ventricular pressure; cyanosis can occur if there is an ASD and decreased right ventricular compliance results in right-to-left shunting through the ASD.

What are the typical signs?

1. Mild-to-moderate stenosis: potentially no symptoms. Systolic murmur heard best at the left upper sternal border, ejection click
2. Severe stenosis: cyanosis, reduced exercise capacity, loud murmur

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What is the treatment?	Mild-to-moderate stenosis: usually, no treatment is indicated. Severe valve stenosis: balloon valvuloplasty in the catheterization lab, or surgical valvuloplasty
What are the complications of treatment?	Pulmonary valve insufficiency, residual obstruction

TGA WITH INTACT VENTRICULAR SEPTUM

What is it?	The aorta arises from the RV and the PA arises from the LV. It represents 10% of CHD (second in incidence to VSD).
Is it more common in boys or girls?	Boys by 4:1 ratio
What is the physiologic result?	**Cyanosis;** oxygenated blood remains in pulmonary circulation and deoxygenated blood in the systemic circulation (**parallel** rather than normal **series** circuit). Blood can mix through a PDA and/or ASD.
How does this condition present?	Cyanosis immediately after birth, respiratory distress
What is the classic finding on the chest x-ray?	Heart classically described as an "egg on a string"
What are the 2 reasons the infant might require cardiac catheterization?	1. For balloon atrial septostomy (Rashkind procedure) to enlarge the ASD and allow better oxygenation 2. For coronary angiography
What is the surgical procedure?	The arterial switch operation (Jatene procedure)
What are the potential complications?	Poor perfusion of the reimplanted coronary arteries, left ventricular dysfunction, AI, supra aortic or pulmonary stenosis

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TETRALOGY OF FALLOT

What 4 elements comprise the tetralogy?	VSD; pulmonary stenosis (subvalvular and valvular); overriding aorta; right ventricular hypertrophy
TOF represents what percentage of all CHD?	5%
What is the physiologic result?	VSD allows interventricular shunting, usually right-to-left. The amount of right-to-left shunting depends on the degree of pulmonary outflow obstruction.
What is the source of the murmur in TOF?	It is primarily due to the pulmonary stenosis; flow across the VSD is usually low velocity and thus not audible.
What is the classic radiographic finding?	Boot-shaped heart
What is a key symptom?	Cyanosis: worsens with activity or may be spontaneous (i.e., "Tet spell")
How is a Tet spell managed?	1. Sedative medication; classically, morphine 2. Knee-chest positioning 3. Oxygen 4. Intravenous medication to raise systemic blood pressure without inotropy or chronotropy, for example, phenylephrine or vasopressin 5. If no reversal of spell occurs, then emergency surgery
What is the treatment?	Patch closure of the VSD, relief of right ventricular outflow tract and pulmonary valve obstruction, often with a transannular patch
List 4 complications of surgical repair.	Residual VSD, residual right ventricular outflow tract obstruction, pulmonary valve insufficiency, arrhythmias (particularly ventricular ectopy)

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What is the outcome?	>90% of patients with definitive repair survive well into adulthood (slightly fewer than the average population). Working capacity, maximum heart rate, and cardiac output are generally less than those for the average person. Pulmonary valve replacement is commonly needed in adulthood.

PULMONARY ATRESIA WITH INTACT VENTRICULAR SEPTUM WITH NORMAL RV SIZE

What is it?	Imperforate pulmonary valve (1% of all CHD)
What are the physiologic results?	Severe cyanosis as newborn; all affected infants have right-to-left shunt at the atrial level; pulmonary blood flow is supplied by the PDA.
List 2 signs or symptoms.	Cyanosis; circulatory collapse if the PDA closes
What is the treatment?	Medical: prostaglandin infusion Surgical options: 1. Pulmonary valvotomy 2. Transannular patch
List 3 complications of repair.	Severe pulmonary insufficiency; residual pulmonary stenosis; right ventricular dysfunction
What are the outcomes?	Similar to TOF

PULMONARY ATRESIA WITH INTACT VENTRICULAR SEPTUM AND HYPOPLASTIC RV

What is it?	Underdeveloped RV and an imperforate pulmonary valve (1% of all CHDs). Coronary abnormalities are common.
What are the physiologic results?	Severe cyanosis as newborn; all affected infants have right-to-left shunt at the atrial level; pulmonary blood flow is supplied by the PDA.

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List 2 signs or symptoms.

Cyanosis; circulatory collapse if the PDA closes

What is the role of cardiac catheterization?

1. Evaluate coronary anomalies
2. Balloon atrial septostomy if atrial communication is too small
3. Stent placement in the PDA

What is the treatment?

Medical: prostaglandin infusion

Catheter: Stent placement in the PDA

Surgical options:
1. BT shunt
2. Transplant

List 3 complications of repair.

Palliative shunts may clot or develop stenosis; myocardial ischemia

What are the outcomes?

Moderate mortality rate for all procedures; long-term survival rates are improving.

PERSISTENT TRUNCUS ARTERIOSUS

What is it?

A single great artery arises from the heart (the truncal artery) which becomes the aorta; the branch PAs arise from the truncal artery in varying patterns; there is always a large VSD (1% of CHD).

What genetic syndrome is seen in 30% of patients with truncus arteriosus?

DiGeorge syndrome

Are patients with truncus arteriosus cyanotic?

Usually, no. They have unrestricted pulmonary blood flow, so despite complete mixing of systemic and pulmonary venous blood, oxygen saturations are often normal at birth.

List 4 signs or symptoms.

Tachypnea, tachycardia, holosystolic murmur with loud S_2; diastolic murmur may also be present.

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List 3 radiographic findings.

Cardiomegaly; increased pulmonary vascular markings; right aortic arch (in one-third of patients)

What is the treatment?

Medical: management of CHF with diuretics

Surgical: establishing continuity from the RV to the branch PAs with conduit, VSD closure (similar to Rastelli procedure)

What are the potential complications?

Conduit stenosis or insufficiency, branch PA stenosis, truncal valve insufficiency or stenosis

What are the outcomes?

Surgically untreated patients die at <1 year from the development of pulmonary vascular obstructive disease.

Surgical therapy now achieves early survival rates of >90%; long-term survival is also improving. Multiple surgical procedures are expected.

TOTAL ANOMALOUS PULMONARY VENOUS RETURN

What is it?

None of the pulmonary veins drain to the LA (1% of all CHD).

List 3 areas into which the anomalous pulmonary veins may drain.

1. **Supracardiac:** connect to a vertical vein or SVC
2. **Cardiac:** connect to the coronary sinus or RA
3. **Infracardiac:** connect to veins below the diaphragm, often the portal vein; obstruction of pulmonary venous return is most common in this group.

What lesion is always associated with TAPVR?

An ASD, which allows blood to enter the left side of the heart

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What is the physiologic result of TAPVR?

Unobstructed flow: comparable to ASD with left-to-right shunt

Obstructed flow: decreased filling of the LA and LV, cyanosis, and decreased cardiac output. Severe pulmonary congestion

What are the signs or symptoms?

Unobstructed flow: tachypnea, murmur, failure to thrive

Obstructed flow: cyanosis and circulatory collapse

What are the characteristic radiographic findings?

Cardiomegaly with increased vascular markings; classic "**snowman**" shape of the heart due to prominent vertical vein, large SVC, and enlarged RA

What is the treatment?

Operation to connect pulmonary venous confluence to the LA and close the ASD

List 2 complications of repair.

Continued obstruction of pulmonary venous return to the LA; persistent pulmonary hypertension

What is the outcome?

Infants with severe pulmonary venous obstruction have the worst outcome and face a 30–35% early and late mortality rate after surgery. If there is no obstruction, outcomes are excellent.

PARTIAL ANOMALOUS PULMONARY VENOUS RETURN

What is it?

A type of CHD in which at least 1, but not all, of the pulmonary veins drain directly or indirectly to the right heart

List 3 areas into which the anomalous pulmonary veins commonly drain.

1. The innominate vein
2. The SVC
3. The RA (often associated with sinus venosus ASD)

What is the physiologic result of PAPVR?

A left-to-right shunt

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What are the signs or symptoms?	Pulmonary flow murmur (similar to ASD), failure to thrive
What is the name of the syndrome in which there is right lung hypoplasia and anomalous return of at least part of the right lung to the IVC-RA junction?	Scimitar syndrome
What is the treatment of PAPVR?	Surgical rerouting of the anomalous vein to the LA
List 2 complications of repair.	Obstruction of the rerouted veins, arrhythmia
What is the outcome?	Usually, there is normal life expectancy.

HYPOPLASTIC LEFT HEART SYNDROME

What is it?	Underdevelopment of the left heart (i.e., the mitral valve, LV, aortic valve, and ascending aorta with arch hypoplasia and coarctation). It represents 3–4% of CHD.
What is the physiologic result?	Almost uniformly fatal within first weeks of life without intervention—systemic circulation depends on left-to-right flow at the ASD and right-to-left flow at the ductus arteriosus.
What are the common signs or symptoms?	Mild cyanosis, sometimes within hours of birth; circulatory collapse with poor perfusion if the ductus closes; soft systolic murmur
What is the medical treatment?	Prostaglandin infusion until surgery
What are the surgical alternatives?	1. **Norwood procedure** (see the preceding text) 2. Heart transplantation

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What are the potential complications of surgical treatment?	Cyanosis, CHF, and reduced exercise tolerance; complications of transplantation include infection, rejection, coronary artery disease, and malignancy.
What are the outcomes?	The mortality rate from the Norwood procedure is 5–30%. For transplanted patients, repeat transplant is expected within 15 years.

TRICUSPID ATRESIA

What is it?	Agenesis of the tricuspid valve (1% of all CHDs)
Why is an ASD obligatory?	To allow systemic venous blood to enter the LA
What are the commonly associated problems?	VSD, TGA, pulmonary stenosis
What is the physiology of a patient with TA and TGA?	The PA arises from the LV; there is usually no pulmonary stenosis in this case, so pulmonary blood flow is unrestricted, causing normal oxygen saturations and CHF; must also rule out coarctation
What is the treatment in the newborn period?	PA banding, or DKS and shunt
What is the physiology of a patient with TA and normal great artery relationship?	Pulmonary stenosis or even atresia is common; thus, the patient will be cyanotic.
What is the treatment in the newborn period?	1. None, if oxygen levels are acceptable 2. Prostaglandin to maintain ductal patency if required for adequate pulmonary blood flow 3. BT shunt 4. Stent placement in the PDA
What is the treatment outside of the newborn period for TA patients with or without TGA?	Glenn operation followed by a Fontan operation

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List 4 complications of surgical repair.

Shunts may form clots or develop stenosis; pleural and pericardial effusions; supraventricular arrhythmias; left ventricular dysfunction as a late outcome

What is the outcome?

After the Fontan procedure: cyanosis is resolved; exercise capacity is less than that of the average person. The 10-year survival rate is about 85%.

ACQUIRED HEART DISEASE

ENDOCARDITIS

What is it?

An inflammatory process, usually caused by bacterial infection, of the endocardium and/or heart valves

What is the name for infection of the endothelium of blood vessels?

Endarteritis

Who is at risk for endocarditis?

Patients with:
1. CHD
2. Prior endocarditis
3. Prosthetic heart valves

List the 2 most common causative agents.

Streptococcus viridans and *Staphylococcus aureus*, accounting for about 80% of cases

What are the potential signs?

Fever, petechiae, new or changing heart murmur; Roth spots (retinal hemorrhages), Janeway lesions (painless hemorrhagic areas on palms and soles), and Osler's nodes (painful nodules on the fingers) are rare in children.

What is the most important diagnostic test?

Multiple positive blood cultures

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What is the role of echocardiography?

Echocardiography can prove the presence of an intracardiac mass or abscess. Transthoracic echo is usually adequate in children. A normal echo does not rule out endocarditis.

List 2 modes of treatment.

1. **Antibiotic treatment** against the identified pathogen (intravenous, generally continuing for at least 4 weeks)
2. **Surgical valve replacement** for intractable heart failure related to severe valve insufficiency or for an infection that cannot be cleared with antibiotics

What are the outcomes?

Streptococcal endocarditis generally has a good outcome. Staphylococcal and fungal endocarditis have high morbidity and mortality rates.

What is thought to be the best way to prevent endocarditis?

Good dental hygiene

What is the role of bacterial endocarditis prophylaxis?

To prevent bacteremia due to dental procedures in patients at highest risk for complications of endocarditis

ACUTE RHEUMATIC FEVER

What is it?

Systemic inflammatory illness caused by the body's response to a pharyngeal group A streptococcal infection

What are the 5 major Jones criteria?

1. Polyarthritis
2. Carditis
3. Chorea
4. Erythema marginatum
5. Subcutaneous nodules

What are some manifestations of carditis on physical exam?

1. Tachycardia out of proportion to fever
2. Heart murmur
3. A pericardial friction rub
4. Gallop rhythm

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What are the possible manifestations of carditis on electrocardiogram?

1. Sinus tachycardia
2. Diffuse ST segment elevation
3. Prolonged PR interval (1st-degree heart block)

What might be found on the echocardiogram?

1. Pericardial effusion
2. MR
3. AI

List 3 components of initial treatment.

1. Treatment for streptococcal pharyngitis (primary prevention)
2. Treatment for inflammation with aspirin for 4–8 weeks or steroids for 2–3 weeks
3. Treatment for CHF, if present

What are the outcomes?

Generally very good; rarely, severe carditis may cause severe valvular dysfunction that requires valve replacement.

MYOCARDITIS

What is it?

An infection of the heart muscle

What are the most common causes?

80% of cases are viral; most commonly, coxsackievirus, influenza virus, and echovirus

Bacteria are the second most common cause.

List 4 signs or symptoms

Fever, tachycardia, ventricular arrhythmias, fulminant heart failure

What can the electrocardiogram show?

Variable; diffuse low voltages, frequent premature ventricular contractions

What will the echocardiogram show?

Poor left ventricular function, dilated LV, MR, pericardial effusion

What is the treatment?

Largely, supportive care to treat heart failure; immune modulators (IVIG, glucocorticoids) are often used.

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What are the outcomes? Some cases completely resolve, with a return of normal left ventricular function; some lead to chronic dilated cardiomyopathy; severe cases can lead to need for ECMO, transplantation, or death.

KAWASAKI DISEASE

What is it? A systemic inflammatory illness of unknown cause occurring in children with a characteristic set of features

How many days of fever are required to make the diagnosis? 5

What are the diagnostic criteria?

1. Conjunctivitis
2. Oral erythema
3. Cervical lymphadenopathy
4. Rash
5. Swelling of the hands and feet

What is the most dreaded complication? Development of coronary artery aneurysms, which can lead to myocardial infarction and death

What is the role of echocardiography? Evaluate for coronary artery aneurysms; also, might reveal myocardial dysfunction, MR, or pericardial effusion

What 2 drugs are used as standard therapy?

1. IVIG
2. Aspirin

What are the outcomes? Without IVIG, 25% of affected children will develop coronary aneurysms; with IVIG, the number is 5%.

CARDIOMYOPATHIES

Hypertrophic Cardiomyopathy

What is it? Cardiac disease characterized by a markedly thickened left ventricular wall

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What are some causative factors?

Altered cardiac myosin or other ultrastructural proteins. Certain gene defects are associated with severe disease and sudden death. Some storage diseases can present with hypertrophic cardiomyopathy.

What is the physiologic result?

Impaired filling of the LV as a result of a thick (stiff) LV; the hypertrophy can cause significant left ventricular outflow tract obstruction.

List 4 signs or symptoms.

1. Sudden death—may be the first indication of the disease
2. Murmur if there is significant outflow obstruction (should be louder when upright vs. supine)
3. Ventricular arrhythmias
4. Decreased capacity for exercise

What might the echocardiogram show?

1. Severe hypertrophy of the left ventricular myocardium, usually of the interventricular septum
2. Systolic anterior motion of the mitral valve
3. Left ventricular outflow tract obstruction
4. MR
5. Left atrial enlargement
6. Hyperdynamic left ventricular systolic function

What is the medical treatment?

1. β-Blockers
2. Placement of an intravenous defibrillator
3. Alcohol septal ablation

What is the surgical treatment?

1. Resection of muscle in the left ventricular outflow tract
2. Mitral valve replacement

What is the outcome?

Variable

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Dilated Cardiomyopathy

What is it?	A disease caused by a dilated, poorly contractile LV
What causes dilated cardiomyopathy?	Many cases are due to sequelae of myocarditis, but metabolic disorders, mitochondrial abnormalities, or inherited or spontaneous mutations in myocardial genes have been identified in some patients.
What is the physiologic result?	Inadequate cardiac output
What are the expected symptoms?	Excessive fatigue, syncope
What are the common signs on physical exam?	Growth failure, tachycardia, gallop rhythm, murmur due to mitral regurgitation
List 2 radiographic findings.	Cardiomegaly; pulmonary edema
What will the echocardiogram show?	Severe left ventricular enlargement and dysfunction; MR; pulmonary hypertension
What are the common medical treatments?	Diuretics β-Blockers Anticoagulants Intravenous inotropes, such as milrinone or dobutamine Digoxin
What is the surgical treatment?	Orthotopic transplantation
What is the outcome?	Rarely, dilated cardiomyopathy can spontaneously resolve; chronic heart failure can be managed on an outpatient basis; need for transplantation is variable.

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ARRHYTHMIAS

What is an arrhythmia? Any rhythm other than regular sinus rhythm

What is sinus arrhythmia? A normal but exaggerated increase in heart rate with inspiration and decrease with expiration seen in children

Common Atrial Arrhythmias

What is sinus tachycardia? Sinus rate greater than normal for patient's age

What is sinus bradycardia? Sinus rate less than normal for patient's age

What are premature atrial contractions? Electrical activity from a site in the atrium occurring earlier than the sinus node that depolarizes the atrium and can then depolarize the ventricles; this is a common finding in normal newborn infants.

What is the physical exam finding? An irregular heart rhythm

Is treatment needed? No

What is a wandering atrial pacemaker? The site within the atrium acting as the pacemaker "wanders" from site to site. The P-wave morphology and PR interval change; QRS complex is normal.

Is treatment necessary? No; it is a normal variant.

What is supraventricular tachycardia? Any tachycardia that begins with a source above the ventricles, that is, the atria or the atrioventricular node. It is usually narrow complex. The rate is generally >200 beats/min.

What is atrioventricular reentry tachycardia (AVRT)? A type of SVT caused by an accessory pathway between the atria and ventricles

What is the eponym applied to a subtype of AVRT? Wolf-Parkinson-White syndrome

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Is AVRT likely to resolve without therapy?

The younger the patient, the more likely it is to resolve and not require long-term treatment

How can the acute episodes of AVRT be treated?

1. Vagal maneuvers
2. Adenosine
3. Electrical cardioversion

How can AVRT be cured?

Ablation of the accessory pathway in the electrophysiology catheter laboratory

Ventricular Arrhythmias

What are the premature ventricular contractions?

Heart beats originating from the ventricular myocardium that are seen on the electrocardiogram as wide QRS beats; they can occur in 5% of normal children.

What is ventricular tachycardia?

A dangerous heart rhythm in which the heart rhythm is driven by a focus in the ventricular myocardium

What is the differential diagnosis?

Electrolyte disturbance, drug toxicity (e.g., digoxin), myocarditis, and myocardial ischemia

LONG QT SYNDROME

What is it?

A genetic disorder causing dysfunction of the ion channels in the heart leading to abnormal repolarization of the myocardium

What are the findings on electrocardiogram?

1. Prolonged QT interval (corrected for heart rate [QTc])
2. Abnormally shaped T waves
3. Bradycardia

What else can cause a prolongation of the QT interval?

1. Medications
2. Electrolyte abnormalities

What is the consequence of long QT syndrome?

Ventricular fibrillation **(Torsade de pointe),** which can cause syncope and sudden cardiac death

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What is the treatment?

1. β-Blockers
2. Defibrillator

PULMONARY HYPERTENSION

What is it?

Abnormally elevated blood pressure in the PAs

What causes it?

1. Heart disease causing elevated pulmonary venous pressure (mitral stenosis, dilated cardiomyopathy)
2. CHD causing left-to-right shunts
3. Thromboembolic disease
4. Connective tissue disease
5. Lung disease
6. Genetic disorders causing abnormal function of the PA endothelium

What is the physiologic result?

Right ventricular failure

What are the potential symptoms?

Syncope, chest pain, poor exercise tolerance, hemoptysis

What are the possible physical exam findings?

Loud S_2, murmur of tricuspid regurgitation, distended neck veins, hepatomegaly

What might the echocardiogram show?

1. Elevated PA pressure (estimated on the basis of tricuspid regurgitation velocity)
2. Tricuspid regurgitation
3. Dilated, thick RV
4. Pericardial effusion

What is the role of cardiac catheterization?

1. Directly measure PA pressure and pulmonary vascular resistance
2. Test whether pulmonary vasodilators are effective

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What is the treatment?

1. Treat the underlying cause, if known
2. Oxygen
3. Pulmonary vasodilators, for example, sildenafil, bosentan
4. Anticoagulation
5. Lung transplantation

What are the outcomes?

92% 5-year survival for children receiving prostacyclin

EISENMENGER SYNDROME

What is it?

The development of right-to-left shunting in patients born with cardiac defects that initially had left-to-right shunting; for example, VSD, ASD, or PDA

What is the physiologic result?

High pulmonary vascular resistance elevates the PA pressure, and deoxygenated blood enters the systemic circulation through any existing communication.

List 3 signs or symptoms.

Cyanosis; poor exercise tolerance; risk of sudden death

What might be seen on the chest x-ray?

Decreased pulmonary vascular markings; heart size usually normal

What will the echocardiogram show?

1. Right-to-left shunting at the anatomic defect
2. Dilated, thickened RV
3. Pericardial effusion (possible)

What is the treatment?

1. Oxygen
2. Pulmonary vasodilator medications
3. Diuretics

What is the outcome?

Prognosis is poor. Pregnancy is contraindicated.

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Chapter 17 Respiratory and Thoracic Disorders

CONGENITAL DIAPHRAGMATIC HERNIA (CDH)

What is it?

A congenital defect in the diaphragm as a result of failure of the pleuroperitoneal canal to close at 8 weeks' gestation

What is a Bochdalek hernia and what are its main characteristics?

By far the most common CDH (85–90%). It is a **posterolateral defect;** 15% have an intact sac.

What is a Morgagni hernia and what are its main characteristics?

An **anterior, parasternal defect.** It is usually smaller than a Bochdalek hernia, tends to have an intact sac, and does not have the pulmonary and systemic ramifications of a Bochdalek hernia.

What is the incidence of CDHs?

About 1 in 4,000 live births

What percentage of Bochdalek hernias are on the left?

85%

List 5 anatomic ramifications of Bochdalek hernia.

1. The abdominal contents herniate into the chest.
2. The lung on the involved side is small and hypoplastic, but the lung on the opposite side also has hypoplasia.
3. There are fewer branches of pulmonary arteries, and they are hypermuscular.
4. The abdominal cavity may be smaller than normal.
5. There is malrotation of the bowel (see Ch 19, p. 294).

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Of Morgagni hernia? (List 2)

The viscera is typically in the hernia. Chest structures are not significantly affected.

What are the physiologic ramifications of a Bochdalek hernia? (List 2)

Bochdalek CDH was once believed to be a surgical emergency. It is now understood that **pulmonary vascular hyperreactivity** (and therefore pulmonary hypertension) and **pulmonary hypoplasia** are the major complications, and therefore, Bochdalek CDH is a medical emergency when it presents at birth.

Of Morgagni hernia?

Usually none, unless the hernia is very large

What are the signs and symptoms of a Bochdalek hernia?

Respiratory distress—usually immediately at birth; the infant's **chest appears expanded,** with **scaphoid abdomen.** Occasionally, a child survives with Bochdalek CDH undetected in the perinatal stage (and therefore bypasses the medical emergency stage) and presents with respiratory symptoms (typically not life-threatening) or, more commonly **gastrointestinal (GI) symptoms** days to weeks later due to the bowel herniated in the chest.

Signs of Morgagni hernia?

May be asymptomatic—or child may have mild respiratory symptoms or GI difficulties. Often, Morgagni hernia is an incidental finding on a chest x-ray.

List 2 prenatal ultrasound findings in Bochdalek hernia.

A multicystic appearance in the involved chest; there may be polyhydramnios.

What are the 3 perinatal radiographic findings in a Bochdalek hernia?

1. Chest radiograph shows viscera in chest.
2. Nasogastric tube, if present, is often seen curling into the involved chest field if the stomach is herniated into the chest.
3. The heart is shifted away from the herniated side.

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Why are the radiographic findings in a Morgagni hernia more subtle?

Because the defect is anterior

What are the initial elements of management of an infant with a Bochdalek hernia?

Intubation with appropriate respiratory support; judicious IV fluids; placement of orogastric or nasogastric tube for decompression of the stomach. Surgical intervention to repair the hernia is **not** emergently required.

What are the major ventilation goals?

Attempt to normalize P_{CO_2}, P_{O_2}, and pH as soon as possible as tolerated by the infant. In the past, hyperoxygenation and hyperventilation were aggressively pursued with subsequent injury to the lung tissue. Now, "permissive hypercapnia" (a.k.a. "gentle ventilation") is pursued with less aggressive ventilator settings.

What variables are aimed for with "permissive hypercapnia"?

Goals are generally to keep the pH above 7.20–7.25, $P_{CO_2} < 55$–60 mm Hg, and preductal O_2 saturations above 85–90%. Oscillatory ventilation and nitric oxide have probably been helpful. However, these infants can be extremely tenuous.

What agent is commonly used to help reduce pulmonary hypertension?

Inhaled nitric oxide is the most commonly used agent because it may act selectively on the pulmonary vasculature. It has not been proven to increase survival.

What may be necessary if conventional therapies fail?

ECMO

What is ECMO?

Extra**c**orporeal **m**embrane **o**xygenation. Essentially, it is a lung bypass machine.

What is the primary goal of ECMO?

To allow the lung to grow and the pulmonary hypertension to subside

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When should surgical repair be performed?

Some measure of respiratory and hemodynamic stability is desired before surgery is performed. This may entail days, and in some cases, may only be obtained by placing the infant on ECMO. Repair may be undertaken before, during, or after ECMO (if ECMO is needed).

List the typical steps in surgical correction of CDH.

1. Subcostal (or occasionally, chest) incision
2. Reduction of the viscera into the abdomen, making sure there is no volvulus
3. Closure of the diaphragm (may require a prosthetic patch)
4. Closure of the abdomen; if the abdomen cannot be closed primarily, a temporary prosthetic may be placed with final closure accomplished 1–2 weeks later.

Repair has been accomplished using minimally invasive techniques as well.

What is the treatment for a Morgagni hernia?

Treatment is **surgical closure** of the defect with routine supportive preoperative and postoperative care. This surgery can be done through a transverse substernal incision or by using laparoscopic technique.

What is the outcome for Bochdalek hernia?

Overall survival is about 75–85%. Some infants may have long-term respiratory deficiency, GI reflux, or CNS sequelae from hypoxia or complications from ECMO.

What percentage of infants with CDH who are placed on ECMO survive?

Approximately 50–60%

What is the prognosis for Morgagni hernias?

Excellent

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EMPHYSEMA

What is it?

An enlargement of the airspace distal to the terminal bronchioles caused by either dilatation or destruction of the surrounding walls

List 4 potential mechanisms for emphysema.

1. **Congenital** (usually restricted to 1 lobe)
2. **Hyperexpansion of distal airspace** to fill the space left by loss of adjacent lung volume from resection or atelectasis ("compensatory emphysema")
3. **Obstruction of gas egress** by foreign body, mass (e.g., tumor, adenopathy), mucosal edema (e.g., asthma), or vascular ring
4. **Destruction of airspace walls,** typically due to the presence of proteases in excess of proteinase-inhibitor activity either because of a deficiency in inhibitor concentration or activity (as in α_1-antitrypsin deficiency) or because of an excess of protease concentration or activity (as in CF, bronchopulmonary dysplasia, or cigarette smoking)

List 4 signs and symptoms.

They vary with underlying etiologic factors. A child may be **asymptomatic** (e.g., compensatory emphysema after lobectomy). Alternatively, symptoms may include **cough, dyspnea, decreased breath sounds,** or an **inspiratory phase lag** over the involved region.

How is it diagnosed?

By radiograph; delineation of the underlying cause may require other measures. Obstructive emphysema can be distinguished from the other forms by obtaining images (plain or fluoroscopic) during expiration, because the increase in lung volume will persist. Decubitus positioning may be used for this purpose.

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What does a radiograph show?

Areas of **hyperlucency** and **decreased lung (vascular) markings.** There may be associated contiguous areas of opacity (either primary or compressive atelectasis) or mediastinal shift.

What is the treatment?

1. Congenital: remove the involved lobe
2. Obstruction: remove the foreign body or relieve obstruction
3. All other causes: good pulmonary toilet with management of underlying condition

LOBAR EMPHYSEMA

What is it?

Overdistension of a histologically normal lung lobe

What are the causes?

It is thought to be caused by poorly developed cartilage of the involved bronchus, creating a "ball-valve" effect. It may also be acquired.

What are the 2 symptoms?

Mild to moderate tachypnea; failure to thrive

In what 2 ways is it diagnosed?

Chest radiograph; CT scan. Bronchoscopy may be helpful in assessing the integrity of the pertinent bronchial branch.

What is the treatment?

If the bronchus is patent, pulmonary toilet may allow temporization until the bronchus strengthens and allows appropriate air passage. When this is not the case, lobectomy is required.

LUNG CYSTS

What are they?

Simple cysts of the lung that most commonly reflect injury to the lung

List 3 common causes of injury to the lung.

Trauma; mechanical ventilation (particularly in premature infants); disease processes (e.g., infection or cystic fibrosis, p. 228)

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Treatment?

Most cysts may be observed. Occasionally, large ones should be resected.

PNEUMONIA OR PNEUMONITIS

What is it?

An inflammatory (pneumonitis) or infectious (pneumonia) process involving the distal airspace. The term is also applied to processes involving the lung interstitium ("interstitial" pneumonia or pneumonitis). It should be distinguished from processes involving the trachea (tracheitis), bronchi (bronchitis), and distal airways (bronchiolitis).

What is the incidence?

Varies with age. Risk is roughly 5% per year in the preschool age group and is increased in institutional settings (e.g., dorms, the military).

What are some common signs and symptoms?

They vary with age and etiologic organism:

Commonly: cough, fever, and chills, but child may also have chest pain, vomiting, diarrhea, or abdominal pain (can mimic **gallbladder disease** or **appendicitis!**)

On physical examination: tachypnea, evidence of increased work of breathing (e.g., nasal flaring, retractions)

What do percussion, auscultation, and oximetry show?

Percussion may demonstrate an area of dullness, either from consolidation or associated pleural effusion. **Auscultation** may reveal areas of decreased breath sounds and inspiratory crackles or rales. However, auscultation may reveal normal breath sounds, especially in small infants. **Oximetry** usually reveals mild to severe oxygen desaturation, depending on the severity of the process.

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How is it diagnosed?

Diagnosis can be made clinically, although radiograph should be used for confirmation in immunocompromised and severely ill children and children with a history of repeated episodes.

List 4 ways to determine the etiologic agent.

By **blood** or **sputum culture,** although in mild cases, in an otherwise healthy child, this is probably unnecessary. Identification is more urgent in the immunocompromised child, thus **bronchoscopy** or **biopsy** for diagnosis may be warranted.

What are the etiologic agents?

Viral and bacterial pathogens and other agents such as fungi. They vary with the child's age and immune status. In all groups, however, **viral pathogens are most common. Geography** or **exposure** may dictate consideration of agents such as fungi (coccidiomycosis, blastomycosis, histoplasmosis) or *Mycobacterium tuberculosis.* If aspiration pneumonia is a possibility, anaerobes should be considered.

What is the most common viral pathogen?

RSV

List the typical bacterial pathogens in the following age groups:

Newborns (list 3)

Group B streptococcus; gram-negative bacilli; *Chlamydia*

1 month–6 years (list 2)

Streptococcus pneumoniae; *Haemophilus influenzae* (*H. influenzae* is becoming less common with increasing use of vaccine.)

Children older than 6 years and adolescents (list 4)

Mycoplasma species; *Streptococcus pyogenes*; *Staphylococcus aureus*; *S. pneumoniae*

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List 3 categories (with examples) of typical agents in hospitalized or immunocompromised children.

1. Gram-negative rod bacteria (e.g., *Pseudomonas*, *Klebsiella*, *E. coli*, *Serratia*)
2. Fungi (*Candida*; others may rarely occur)
3. Other nonbacterial agents (*Pneumocystis*, CMV, Epstein-Barr virus)

List 2 components of treatment for most children.

Most otherwise healthy children can be treated as outpatients with:

1. Oral antibiotics (e.g., amoxicillin-clavulanate, erythromycin, cephalosporin)
2. Antipyretics

Generally, cough suppressants are avoided, but they may be acceptable at bedtime to facilitate sleep.

For severely ill children?

Severely ill children (i.e., those with high fever, dehydration, intractable cough, hypoxemia) may need to be admitted to a hospital for IV antibiotics and supportive therapy (e.g., IV fluids, oxygen, chest physiotherapy). **Any immunocompromised child should be hospitalized.**

List 5 complications.

Pleural effusion, empyema, pulmonary abscess, respiratory failure, bronchiectasis (more common with recurrent episodes but may occur acutely and be reversible)

ASTHMA

What is the definition of asthma?

A chronic disease of the airways characterized by intermittent respiratory symptoms and persistent inflammation. These symptoms are secondary to narrowing of the large and small airways from inflammation and smooth muscle spasm. The inflammation also causes airway hyperresponsiveness to a variety of stimuli.

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What is "reactive airway disease (RAD)"?

RAD is a vague term that is often substituted for asthma. However, the nature of RAD is nonspecific and literally means the capacity of the airways to react to a host of stimuli, whereas the term "asthma" incorporates a disorder, though heterogeneous, which incorporates both functional and inflammatory components.

What is the prevalence of asthma?

It is the most common chronic disease in childhood. Its prevalence in the United States is 5–7%. African American children are more frequently affected than Caucasian children and have a higher rate of hospitalization and death from asthma. The total prevalence in industrialized countries has greatly increased during the past several decades.

What is the current theory of the pathophysiology of asthma?

Asthma involves multiple inflammatory mediators, such as histamine, IgE, cytokines, and leukotrienes, and cell types, including mast cells, activated lymphocytes, eosinophils, and neutrophils. Chronic inflammation can result in basement membrane thickening, airway remodeling, smooth muscle hypertrophy, and mucous plugging.

What are the risk factors for developing asthma?

Family history, atopy (eczema, allergic rhinitis), smoking in the family, male sex. There are likely other environmental exposures including early-life viral infections and airborne pollutants, which modify or even cause asthma in childhood.

List triggers that evoke asthma symptoms.

Viral infections (primarily human rhinovirus), allergens (dust mites, mold, dander, pollen, cockroaches), irritants (smoke, pollution, fumes, odors), cold air, exercise

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What are the symptoms of asthma?

Daytime and nighttime coughing, wheezing, chest tightness, dyspnea, tachypnea, exercise intolerance

What are the physical examination findings during an asthma exacerbation?

End-expiratory wheezing, inspiratory and expiratory wheezing (severe), increased work of breathing with use of accessory muscles, increased inspiratory to expiratory ratio (e.g., 1:3), pulsus paradoxus (reduction of systolic BP by 10 mm Hg during inspiration), tachycardia, absent breath sounds with cyanosis (severe), impaired sensorium

List 4 typical findings on chest radiograph.

1. Lung hyperinflation with flattening of the diaphragms
2. Increased AP diameter
3. Increased perihilar markings owing to inflammation
4. Atelectasis

How is asthma severity categorized?

Mild intermittent

Symptoms < 2 times a week and night symptoms < 2 times a month

Mild persistent

Symptoms > 2 times a week, but < 1 time a day, and night symptoms > 2 times a month

Moderate persistent

Symptoms daily with night symptoms > 1 time a week

Severe

Symptoms continually with frequent night symptoms

What is the rule of 2's?

If a patient has symptoms > **2** times a week or nighttime symptoms > **2** times a month, he has **persistent,** not intermittent asthma.

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What is the differential diagnosis of wheezing (in each of the following categories):

Upper airway disease (including the larynx)? Vocal cord dysfunction, allergic rhinitis, laryngeal edema/anaphylaxis, laryngomalacia, partially lodged foreign object, sinusitis, retropharyngeal abscess, epiglottitis

Large airway obstruction? Foreign body, tumor, or lymph node compressing airway, vascular rings, laryngeal webs, tracheomalacia, tracheitis, tracheal stenosis, cystic lesion compressing airway, tracheal involvement in Crohn disease, chronic aspiration syndromes

Small airway obstruction? Viral bronchiolitis, CF, chronic lung disease, or bronchopulmonary dysplasia, bronchiolitis obliterans organizing pneumonia (BOOP), pan-bronchiolitis syndrome associated with pseudomonas infection, organizing pneumonias, drug-induced (chemotherapy), chronic aspiration syndromes, immotile cilia syndromes

Nonpulmonary? Heart disease, GERD, immunodeficiency, sickle cell vasculopathy, pulmonary artery hypertension, posttransplant bronchiolitis syndrome

What are the 5 asthma treatment goals?

1. Prevent symptoms
2. Maintain normal activity levels
3. Prevent recurrent exacerbations
4. Minimize the use of β_2-adrenergic agonists to <1 time per day
5. Avoid adverse effects from medication

What is the treatment for:

Mild intermittent asthma? β_2-Adrenergic agonists as needed

Mild persistent asthma? Low-dose inhaled corticosteroids; alternative is leukotriene receptor antagonists (LRAs).

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Moderate persistent asthma?

Low- to medium-dose inhaled corticosteroids plus long-acting β_2-adrenergic agonists; alternative is the addition of theophylline or LRA.

Severe persistent asthma?

High-dose inhaled corticosteroids plus long-acting β_2-adrenergic agonists plus systemic steroids if necessary; some patients require daily systemic steroids and/or anti-IgE treatment (omalizumab).

Why is using a spacer with an MDI (metered-dose inhaler) important?

It allows for better administration of medication to the airways instead of being deposited on the mouth or tongue.

Do inhaled corticosteroids affect growth?

Growth velocity may slow during the first year, but there is no difference in predicted adult height.

What is the treatment of an acute asthma exacerbation?

Oxygen, oral or IV corticosteroids, inhaled β_2-adrenergic agonists, inhaled anticholinergics (when used in the ER, they decrease hospital admission). For severe exacerbations: IV magnesium, terbutaline, epinephrine

List 7 signs of theophylline overdose.

Early signs are insomnia, headache, nausea, and vomiting; high theophylline levels may cause seizure, coma, and death.

List 4 other treatment modalities to minimize asthma symptoms besides medications.

Allergen exposure reduction, patient and family education, immunotherapy, immunizations

AIRWAY FOREIGN BODY

Where do aspirated foreign bodies typically lodge?

1. Usually below the carina
2. In toddlers, foreign bodies lodge with equal incidence in either mainstem.
3. In older children, they lodge more frequently in the right mainstem.

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List 4 symptoms.

Coughing, gagging, choking, and wheezing. An asymptomatic interlude may follow.

List 2 radiographic findings.

1. Either hyperinflation or hypoinflation of affected lung may occur.
2. Foreign body may be visible if radiopaque, but not all are (especially peanuts and popcorn kernels, which are 2 of the most commonly aspirated foreign bodies in small children)

What is the treatment?

Removal of the object via rigid bronchoscopy because the patient's airway can be controlled during the procedure.

List 5 potential sequelae if the foreign body is not removed.

Pneumonitis or pneumonia; abscess; bronchiectasis; pulmonary hemorrhage; erosion and perforation of the enclosing structure

CYSTIC FIBROSIS (CF)

What causes CF?

A defect in the CF transmembrane regulator protein

How is CF transmitted?

Autosomal recessive trait; about 1 in 20 Caucasians carry the gene.

How common is CF?

In the Caucasian population, about 1 in 2,000. It is also found in other populations.

What is the pathophysiology?

An imbalance in Na and Cl creates a thick, sticky mucus that is difficult to clear. Abnormal, thickened secretions occur in a variety of organs, causing inspissation and mucus buildup. Pancreatic insufficiency is present.

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List 4 ways patients with CF may present.

1. Meconium ileus in the neonate
2. Recurrent bronchitis, pneumonia, or both
3. Malabsorption, with failure to thrive
4. Male infertility

What percentage of infants with CF have meconium ileus?

10% of CF infants have meconium ileus; 99% of full-term infants with meconium ileus have CF.

List 2 ways CF is diagnosed.

Usually by an elevated sweat chloride concentration. DNA testing for specific mutations may also be used.

What is the most common CF gene mutation?

ΔF508, although there are over 1,000 other mutations. The relative frequency of specific mutations varies with ethnicity.

List 6 components of treatment.

Nutritional support, pancreatic enzyme supplementation, chest physical therapy, antibiotics, bronchodilators. Lung transplantation is now being attempted.

What is the outcome?

With appropriate therapy, many patients live into adulthood. The end-stage event is usually respiratory failure.

TRACHEOMALACIA

What is it?

Suboptimal integrity of the tracheal wall and cartilage rings that leads to partial collapse of the trachea on inspiration

What group of children gets this condition?

Neonates; it reflects incomplete maturation of the tracheal structures.

List 2 associated conditions.

It can be a primary condition or exacerbated by other conditions, such as esophageal atresia or vascular rings.

List 2 signs and symptoms.

Inspiratory stridor; in severe cases, the infant may have **"dying spells"**—periods of **prolonged apnea** resulting in **cyanosis** and requiring stimulation for resolution.

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List 2 ways it is diagnosed. Fluoroscopic examination; bronchoscopy

List 4 treatments.

1. If condition is primary, it is usually self-resolving.
2. If associated with exacerbating process, then that process needs to be corrected (e.g., division of vascular ring).
3. Occasionally, aortopexy is done to enhance opening of the trachea.
4. Tracheostomy is occasionally needed for airway support until the trachea matures.

PNEUMOTHORAX

What is it? Separation of the visceral pleura from the parietal pleura, resulting in the presence of air in the pleural space

List 3 most common causes in infants. Barotrauma; RDS (previously called "hyaline membrane disease"; Ch 10, p. 94); bronchopulmonary dysplasia

In older children? (List 6) Trauma, rupture of apical bleb, CF, severe coughing, asthma; it may also be idiopathic.

What are the symptoms? Mild to severe respiratory distress, and sometimes chest pain. A small pneumothorax may be asymptomatic.

What is a tension pneumothorax? Air collection in the pleural space under pressure, creating a shift in the mediastinum, compression of the opposite lung, compromise of venous return to the heart, and hemodynamic compromise. **THIS IS A LIFE-THREATENING CONDITION!**

How is pneumothorax diagnosed? **Chest radiograph.** Occasionally, the supine trauma victim will have pneumothorax discovered during a CT scan, because the air is anterior in this situation.

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What is the treatment?

A small, asymptomatic pneumothorax may be allowed to resolve spontaneously. Otherwise, chest tube placement is required until the pneumothorax resolves and an air leak, if present, seals.

List 2 ways recurrent pneumothorax is treated.

1. Instillation of a sclerosing agent, such as talc or tetracycline, via a chest tube or thoracoscopy
2. Resection of an apical bleb or other site of parenchymal leak via thoracoscopy or thoracotomy

How is tension pneumothorax treated?

Immediate placement of a chest tube. If a tube is not available, a large-bore angiocatheter or needle should be placed in the anterior **second intercostal space in the midclavicular line** for decompression.

CHYLOTHORAX

What is it?

An accumulation of lymph fluid (chyle) in the thorax. It can be **congenital** or **acquired.**

List 2 common congenital causes.

Abnormalities of the thoracic duct; birth trauma

List 6 common causes of acquired chylothorax.

Trauma; operative injury (especially during cardiothoracic procedures); neoplasm; thrombosis of the subclavian vein or the superior vena cava; lymphangiomatosis; severe coughing

What are the symptoms?

Respiratory insufficiency or distress if collection is large enough

How is it diagnosed?

Chylothorax appears as a radiopaque fluid collection on the chest radiograph. Diagnosis is confirmed by thoracentesis and analysis of the fluid.

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List 5 typical characteristics of chyle.

Appearance is milky or straw colored; lymphocyte predominance; protein content ≥ 5 g/dL; fat content ≥ 400 mg/dL; triglyceride level ≥ 110 mg/dL

List 3 components of the initial treatment.

Thoracentesis; low-fat diet or parenteral nutrition; chest tube drainage or repeated thoracentesis as necessary

List 3 surgical options that may be used for refractory cases.

1. Right thoracotomy with ligation of thoracic duct
2. Thoracoscopy of the affected side with clipping or suturing of the leaking site, application of fibrin sealant, or both
3. Placement of pleuroperitoneal shunt. The shunt is removed when the leak subsides.

PECTUS DEFORMITY

PECTUS EXCAVATUM

What is pectus excavatum?

Also known as "funnel chest," this condition manifests as a significant depression of the sternum.

What is the cause?

Believed to be an abnormality in growth of the cartilage connecting the sternum to the ribs

List 4 significant anatomic and physiologic effects.

1. The heart is shifted to the left.
2. In severe deformities, the heart or the lungs are compressed.
3. Children may manifest symptoms of asthma or dyspnea on exertion. However, many children are asymptomatic.
4. Some children may experience chest pains.

List 6 common associated conditions.

Scoliosis, Marfan syndrome, clubfoot, syndactyly, Klippel-Feil syndrome, mitral valve prolapse

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What are the 3 indications for surgery?

1. Significant respiratory insufficiency
2. Significantly abnormal appearance; some children with pectus deformity may be ridiculed by their peers and be self-conscious to a degree that significantly affects their self-image and their activities.
3. Ongoing chest pain with no other cause

What is the Haller Index?

The Haller Index is calculated by dividing the transverse diameter of the chest by the AP diameter (from inside the sternum to the anterior edge of the vertebral body) using a CT image of the thorax.

What is its significance?

A high Haller Index predicts cardiopulmonary functional compromise from a pectus excavatum. A level > 2.5 is considered significant and a level > 3.2 is considered severe.

List 2 methods of repair.

1. In the traditional repair (Ravitch procedure), the abnormal cartilages are removed, whereas the surrounding perichondrium is preserved. The sternum is then elevated by any of a number of different methods and secured. Often, a metal strut is placed substernally for support and is removed 3–6 months later. While the sternum is supported, new cartilage grows back within the perichondrium in the appropriate position.
2. The Nuss procedure is a minimally invasive procedure in which a metal strut is placed under the sternum with thoracoscopic guidance. The strut forces the chest into a normal configuration but must stay in place for about 3 years.

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What is the outcome?

The cosmetic and the physiologic results of the traditional repair are very good. Patients return to full activity after 3–6 months. There are reports of some incidences of compromise of chest growth in repairs done in very young children. Therefore, this procedure may be best done in patients who have experienced a good portion of their chest growth.

The Nuss procedure has been favorably received by many and results have been promising. It has a higher incidence of strut displacement, and a longer postoperative requirement for pain management. Long-term results are not as well established as for the traditional repair but are promising.

PECTUS CARINATUM

What is pectus carinatum?

A condition in which the sternum protrudes. It is also a result of abnormal growth of costal cartilages.

List 7 associated conditions.

Congenital heart disease, marfanoid habitus, scoliosis, kyphosis, muscular defects, skeletal defects, asthma

What are the options for repair?

1. The most common method is similar to the traditional repair of pectus excavatum, except with **depression** and stabilization of the sternum.
2. There has been some reported success with external braces that compress the sternum. These must be worn on a regular basis for months to a few years.
3. A repair using an adaptation of the Nuss Procedure may also hold promise.

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ESOPHAGEAL DUPLICATION CYST

What is it?	Congenital cyst arising from an abnormality in foregut development
What is the location?	Mediastinum; it may share a common wall with the esophagus.
What is the histology?	Squamous epithelial lining, but may have ciliated mucosa with some cartilage in the wall
What are the 2 ways a patient may present?	Respiratory distress; the condition may also be found incidentally on a radiograph where it appears as a solid mediastinal mass.
List 2 ways it is diagnosed.	Chest radiograph, CT scan (may be first suspected on prenatal ultrasound)
What is the treatment?	**Surgical excision** via thoracotomy or thoracoscopy. If there is a common wall with the esophagus, cyst mucosa should be stripped from the common wall.

BRONCHOGENIC CYST

What is it?	Congenital cyst arising from cells that become isolated during bronchial development
What are the 2 locations?	1. Central: near the hilum or mediastinum; usually solitary 2. Peripheral: may be multiple
What are the 2 ways the patient may present?	1. Respiratory distress 2. The condition may be found incidentally on radiograph.

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What are the radiographic findings of central and peripheral cysts?

Central: solid-appearing mass, or cystic lesion with air-fluid level

Peripheral: multiloculated appearance that may be confused with congenital cystic adenomatoid malformation (see CCAM, p. 237 or even congenital diaphragmatic hernia (see CDH, p. 215)

List 2 ways it is diagnosed.

Chest radiograph, CT (may be first suspected on prenatal ultrasound)

What is the treatment for each type of cyst?

Central: surgical excision of cyst via thoracotomy or thoracoscopy

Peripheral: wedge resection or resection of involved lung lobe via thoracotomy or thoracoscopy

PULMONARY SEQUESTRATION

What is it?

Mass of abnormal lung tissue receiving an abnormal (i.e., systemic) blood supply, with no communication with the tracheobronchial tree

List the 2 types and the 3 characteristic features of each.

1. **Intralobar** (90%): lies within the lobe of a lung; arterial supply is systemic; venous drainage may be systemic or pulmonary
2. **Extralobar** (10%): has its own pleura; arterial supply and venous drainage may be systemic or pulmonary; may have immature parenchyma or an associated congenital cystic adenomatoid malformation

What are the symptoms?

Child is usually asymptomatic at birth. Serial bouts of pneumonia may follow after 1–2 years.

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List 2 ways it is diagnosed.

Chest radiograph, CT (may be first suspected on prenatal ultrasound)

What is the treatment?

Surgical excision via thoracotomy or thoracoscopy

List 3 associated anomalies.

Congenital heart defects (Ch 16); congenital cystic adenomatoid malformation (CCAM; see the following text); arteriovenous malformation with shunting

CONGENITAL CYSTIC ADENOMATOID MALFORMATION (CCAM)

What is CCAM?

Congenital cystic changes of the lung. These lesions may also now be referred to as "congenital pulmonary airway malformations" (CPAMs).

What are the 3 types and their characteristics?

Type I: Large, irregular cysts

Type II: Smaller, more closely arranged cysts

Type III: Dense, small cysts; may resemble fetal lung

What is the histology?

Cuboidal and low columnar epithelium; few mucogenic cells

By what mechanism do the cysts arise?

Excessive proliferation of bronchioles at the expense of alveoli

List 3 symptoms.

1. Respiratory distress in infants if involved area is large (usually type II or III; rarely, fetal hydrops may occur)
2. Older children or adults may present with infection.
3. Many children are asymptomatic and the lesion is found on prenatal ultrasound or is an incidental finding on the chest radiograph.

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List 2 ways it is diagnosed.	Chest radiograph, CT (may be first suspected by prenatal ultrasound)
Do CCAMs sometimes resolve spontaneously?	Probably not. However, a CCAM seen on prenatal ultrasound may diminish in size to where it is not visible on postnatal chest x-ray. More sophisticated imaging should be undertaken at some point to determine the status of the lesion.
What is the treatment?	Excision of the affected lobe or lobes
What may CCAM be associated with in older children and adults?	Lung cancer

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Chapter 18 Head and Neck

EPIGLOTTITIS

What is it?	Rapidly progressive bacterial infection causing acute inflammation and edema of the epiglottis and adjacent structures (aryepiglottic folds, arytenoids); also known as "supraglottitis"
Why is it important?	It is **life-threatening!** Affected children may have sudden and complete airway obstruction.
What is the usual age at presentation?	2–7 years of age; infants, older children, and adults can also be affected.
Is there a seasonal incidence?	No
Why has there been a decrease in the incidence of childhood epiglottitis?	The decrease in the incidence of childhood epiglottitis has been due to routine infant vaccination with Hib protein-polysaccharide conjugate vaccine
What are the causative agents?	***Haemophilus influenzae* type b** was the primary cause in the pre-vaccine era and can still occur today despite immunization. Other causes include *H. influenzae* type A, *H. parainfluenzae*, *Streptococcus pneumoniae*, *Staphylococcus aureus*, and β-hemolytic streptococcus. Also consider *Candida albicans* in the immunocompromised patient.

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What is the classic presentation?

3 D's: Drooling, Dysphagia, Distress. A previously well child with the sudden onset of sore throat, high fever, drooling, dysphagia, irritability, or lethargy. The child appears toxic with inspiratory stridor and respiratory distress. Progresses rapidly. The lack of viral prodrome helps differentiate it from croup.

What is the tripod position?

A sitting position in which the arms are extended in front of the body supporting the trunk; the neck is hyperextended with the chin protruding. Children with epiglottitis adopt this position because it maximizes the size of the supraglottic airway.

What is the differential diagnosis?

1. Infection: bacterial tracheitis, peritonsillar abscess, retropharyngeal abscess, diphtheria
2. Foreign body aspiration
3. Anaphylaxis
4. Trauma: burns, thermal injury, blunt trauma, caustic ingestion

What must be done first in evaluation and management?

Protection of the airway is the primary priority.

List the key steps in the management of epiglottitis.

1. Keeping the patient **calm** and with the parents
2. Administer **100% O_2,** may use blow-by with child in parent's lap to avoid further agitation
3. Assembling at bedside: **CPR equipment,** including resuscitation bag and mask, intubation equipment, and instruments for emergency **cricothyroidotomy**
4. Calling senior **pediatrics, anesthesia, and surgical (pediatric surgery or otolaryngology) staff** to bedside

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5. Taking patient (accompanied by parents) to the **operating room** for induction of anesthesia, placement of IVs, direct laryngoscopy, intubation, and blood and epiglottis cultures. Equipment and expertise for an emergency tracheostomy should be present.
6. Admission to the **intensive care unit.** Not every child with epiglottitis will have the classic signs and symptoms. **It is better to initiate a "false" epiglottitis drill than to miss this disease.**

What key laboratory and diagnostic studies are ordered?

Only after the patient has a secure airway:

1. **Blood culture,** only 15% will be positive
2. **WBC count,** which may be moderately elevated
3. **Lateral neck radiograph,** which shows a thickened epiglottis ("thumb sign") and a distended hypopharynx

What should the physician NOT do when evaluating the child's condition?

1. Do not agitate the child. Avoid trying to visualize the pharynx and epiglottis with a tongue blade as well as drawing labs.
2. Do not make the child lie supine.
3. Do not leave the child unaccompanied.
4. No laboratory procedures, needle sticks, or radiographs should be performed before steps are taken to protect the airway.

All of these things can lead to airway obstruction and cardiopulmonary arrest.

How is the diagnosis confirmed?

Diagnosis is confirmed by seeing an edematous cherry-red epiglottis on endoscopy.

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What are the main components of treatment after protection of the airway and diagnosis are established?

1. **Maintain adequate (usually artificial) airway** until inflammation and edema resolve—often 36–72 hours
2. **Parenteral antibiotics,** directed against *H. influenzae*, *S. aureus*, *pneumococci*, and group A streptococci. A third-generation cephalosporin for 7–10 days is 1 option.

What role do steroids play?

There is no evidence to support the use of steroids to decrease airway edema in epiglottitis.

What additional steps should be taken if *H. influenzae* is the causative agent?

Rifampin prophylaxis should be given to close contacts who are at risk.

How can we help prevent acute epiglottitis?

Make an effort to maintain high immunization rates.

CROUP

What is croup?

Viral infection of the upper and lower respiratory tract that causes **subglottic inflammation** (also called "laryngotracheobronchitis")

What are the 2 classic features of croup?

Stridor (a high-pitched sound heard on inspiration) and **barking cough**

What is the usual age at presentation?

3 months to 3 years of age; peak incidence at 2 years of age

What is the epidemiology of croup?

Affects boys slightly more often than girls. Peak occurrence is in fall and winter.

What are the primary causative agents of croup?

Parainfluenza virus (especially type 1), respiratory syncytial virus, adenovirus, influenza virus, *Mycoplasma pneumoniae*, and measles virus

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Describe the typical history and course for croup.

Illness begins with several days of mild upper respiratory symptoms. This is followed by hoarseness, a nonproductive **"barking" cough** (sometimes described as "seal-like"), and **stridor.** Most cases are mild but can progress to significant respiratory distress (tachypnea, nasal flaring, retractions) and hypoxia. Symptoms often worsen at night.

What is the differential diagnosis for croup?

See Epiglottitis, p. 239

It is important to consider other causes for stridor and respiratory distress that require specific therapy, **especially foreign body aspiration.**

How is the diagnosis made?

Clinically. **The physician should try not to agitate the child, particularly if the symptoms are severe.**

What diagnostic studies are used?

Perform studies only if patient is not in respiratory distress.

1. Radiograph of the anterior-posterior neck may show a "pencil tip" or **"steeple sign"** of the subglottic trachea. Do not use a radiograph to make management decisions in a patient with an unstable airway.
2. Laboratory studies (e.g., CBC) usually not helpful

Why do some children improve spontaneously?

Because of natural fluctuations in the disease

List 2 treatments for mild cases.

1. Humidification
2. Exposure to cold night air is thought to help, but this is largely anecdotal.

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What are some treatments for severe cases of croup?

For more severe symptoms requiring hospitalization:

1. **Airway support,** including O_2, pulse oximetry, and intubation if necessary. Use a smaller ET tube than predicted for the child's size as there is airway swelling. Clinical assessment and close observation are of paramount importance.
2. **Humidification,** via cool mist. Avoid croup tents as they make observation of the patient difficult.
3. **Racemic** epinephrine—may cause rapid improvement in symptoms. If used, watch for **rebound phenomenon**—symptoms may abruptly return when the effect of epinephrine wears off, usually within 2 hours.
4. **Corticosteroids (most often dexamethasone due to long half-life)**—may help in mild to severe croup; may be used in the outpatient setting if patients demonstrate maintained improvement 2–3 hours after treatment.

Do most children with croup need hospitalization?

No. Most cases are mild and symptoms typically resolve within a few days.

What is spasmodic croup?

A benign condition with recurrent episodes of stridor and barking cough. It may be triggered by viral illness but is not a direct result of the infection. It typically is of short duration, resolves spontaneously, and is rarely associated with severe respiratory distress.

PIERRE ROBIN SEQUENCE

What is it?

Congenital micrognathia/retrognathia, with associated cleft of the soft palate and a relative glossoptosis (the tongue is not actually larger than normal). It may be isolated or part of a larger syndrome.

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What are the 2 general symptoms?

1. Airway obstruction causing respiratory distress, worse when the infant is supine
2. Feeding difficulties (especially with palate and tongue abnormalities)

What are the treatments for the 2 components of the malformation?

1. **Micrognathia:** Prone positioning while sleeping and feeding. A nasopharyngeal tube may be helpful and tracheostomy is required in some cases. Rarely, suturing the tip of the tongue to the lower lip may support a patent airway, though this is controversial. Distraction osteogenesis, a method of enlarging the mandible, is an emerging technique.
2. **Palate abnormalities:** Surgical intervention is necessary and is usually done around 10–18 months of age. A tracheostomy may be needed until repairs are completed. If feeding is difficult, a gastrostomy tube is needed.

CHOANAL ATRESIA

What is choanal atresia?

Congenital persistence of a bony membrane across the nasopharyngeal passage

How does choanal atresia typically present?

Marked respiratory difficulty at birth that improves with crying (because when not crying, infants are obligate nose-breathers)

How is choanal atresia diagnosed?

Examiner's inability to pass a suction catheter into the pharynx via the nasal passages. The diagnosis may be confirmed by contrast nasopharyngography.

What are the 2 treatment components for choanal atresia?

1. Initial treatment: maintenance of the oral airway until the infant can breath on his or her own
2. Resection of the bony septum and placement of stents until the passage epithelializes

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VOCAL CORD PARALYSIS

What is it?	Paralysis of 1 or both cords, which may be either **congenital** or **acquired**
What are the 4 common causes of acquired vocal cord paralysis?	Birth trauma (thought to be due to stretching of the recurrent laryngeal nerve); direct injury to the recurrent laryngeal nerve during thoracic or cardiac surgery (most commonly ligation of a patent ductus arteriosus); CNS tumors; infections
What are the potential symptoms?	**Bilateral:** inspiratory and expiratory **stridor** or frank respiratory distress **Unilateral:** symptoms may be minimal; dysphonia and feeding difficulties are most common
What is the method of diagnosis?	Bedside fiberoptic laryngoscopy
What is the treatment?	Many cases of vocal cord paralysis resolve spontaneously over weeks to years, depending on the etiology. Tracheostomy may be needed to alleviate severe respiratory symptoms.

LARYNGEAL WEB

What is laryngeal web?	Congenital abnormality of the glottic region resulting in a weblike lesion
What is the range of symptoms?	Symptoms range from mild inspiratory-expiratory stridor and dysphonia to frank respiratory distress.
How are laryngeal webs treated?	1. A thin web may be lysed with cautery or a laser. 2. A thicker web may require more extensive reconstruction and may necessitate tracheostomy.

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LARYNGEAL CLEFT

What is it?	Failure of fusion of the posterior larynx. It may extend to the trachea.
What are the 2 major symptoms?	Hoarseness and aspiration
How is it diagnosed?	Contrast swallow may show the presence of the cleft. Endoscopy defines the anatomy.
How is it treated?	Very mild clefts may only require thickening of feeds. Repair may be accomplished using endoscopic assistance for less severe clefts, and direct surgical repair for larger clefts.

SUBGLOTTIC STENOSIS

What is it?	Narrowing of the subglottic region, which may be either **congenital** or **acquired**
How is it acquired?	May be a sequela of a previously placed endotracheal tube, especially if it was inappropriately large for the airway or the child was intubated for a prolonged period of time
What are the grades of subglottic stenosis using the Myer-Cotton system?	Grade 1: No obstruction to 50% obstruction Grade 2: 51–70% obstruction Grade 3: 71–99% obstruction Grade 4: No detectable lumen
What are the treatments for congenital and acquired subglottic stenosis?	Depends on severity. Mild cases may only require supportive care with the infant outgrowing the condition. Dilation or laser therapy may also be useful. More severe cases will require reconstruction (typically cricoid split or laryngotracheoplasty). A tracheostomy may be required.

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What is laryngeal atresia? Complete nonformation of the laryngeal area, which is **incompatible with life** unless there is a large tracheoesophageal fistula at birth.

BRANCHIAL CLEFT REMNANTS

What are they? Remnants of branchial arches that are embryologic sources of head and neck structures

List 4 forms they may take. Cysts, sinuses, fistulae, and cartilaginous remnants

What are the names and locations of the commonly found remnants?

First branchial remnant: lies anterior to the ear and may extend to the eustachian tube

Second branchial remnant: begins in the midneck, anterior to the sternocleidomastoid muscle, and may extend up through the carotid bifurcation to the pharynx

Third branchial remnant: located superior to the medial portion of the clavicle. Deep involvement of third branchial remnants is rare, but if present, extends lateral to the carotid bifurcation, up toward the pharynx.

What are the typical features of presentation? A draining area, dimple, or mass at 1 of the 3 branchial remnant sites. Infection may be the first presenting sign.

What is the treatment for branchial cleft remnants? Surgical excision; more than 1 incision may be needed for extensive lesions.

THYROGLOSSAL DUCT REMNANT

What is it? Remnant of the embryologic path that the thyroid takes from the foramen cecum to its final position

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What 2 forms may it take?

Cyst (75%); sinus (25%)

List key features of presentation.

1. The child usually has an asymptomatic mass in the anterior midline of the neck.
2. The mass may be an infected, draining site.
3. The mass moves upward with swallowing.

What is the treatment?

Surgical excision with a Sistrunk procedure: the cyst or sinus is excised widely along its tract to the base of the tongue. Excision includes the middle third of the hyoid bone.

Are thyroid function tests necessary?

Yes, if thyroid tissue is found in the excised tissue

Why?

Thyroid tissue in the excised cyst or sinus may represent the only thyroid tissue the child has, necessitating thyroid hormone replacement.

What may thyroglossal duct cysts be a risk for later in life?

Papillary thyroid carcinoma

TORTICOLLIS

What is torticollis?

An intense spasm of the sternocleidomastoid muscle

What is the cause?

Uncertain. If present at birth, it may be due to intrauterine positioning. Other causes are birth trauma and infection.

What are the symptoms?

Typically recognized in infants 2–8 weeks old. The infant tends to keep her or his head tilted toward the side of the spasm with the face turned away from the side of the spasm. The baby resists movement in the opposite direction.

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What are the typical physical findings?

A mass is noted in the midportion of the sternocleidomastoid muscle; the rest of the muscle is very tight.

What is the treatment?

The goal is to stretch the sternocleidomastoid muscle and relieve the spasm. This is done in 3 ways:

1. Stimulate the infant to turn his or her face toward the side of the affected muscle. This can often be done during feeding.
2. Turn the infant's face passively toward the side of the affected muscle.
3. **Gently** massage the spasm area. (If massage is too vigorous, bradycardia may result from carotid body stimulation.)

These steps are repeated on a routine basis during daily feeding and care activities until torticollis resolves.

What is the outcome?

Torticollis usually resolves in 2–6 weeks with the therapy noted earlier. Only rarely is surgical intervention required.

PERITONSILLAR ABSCESS

Where is it?

Adjacent to the palatine tonsil. This is the most common abscess of the head/neck region.

How does it present?

Fever, sore throat that does not improve with antibiotics, trismus, and "hot potato" voice

Key physical exam findings?

Erythematous pharynx with **asymmetric** swelling, **uvula deviation,** and area of **fluctuance** next to the tonsil

What are the causative bacteria?

Usually polymicrobial with both aerobic and anaerobic organisms. Predominantly *Streptococcus pyogenes*, *S. aureus*, and respiratory anaerobes

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How is it treated?

Needle aspiration, incision and drainage or tonsillectomy, followed by antibiotics. Augmentin is the drug of choice since most causative organisms are β-lactamase producers.

CRANIOSYNOSTOSIS

What is it?

Premature fusion of 1 or more of the cranial sutures, frequently resulting in an abnormal head shape

Which way is growth restricted?

Perpendicular to the affected suture

How can it be differentiated from positional plagiocephaly?

1. If one-sided occipital flattening is due to closure of the lambdoid suture, there will be frontal bossing on the opposite side (leading to a "trapezoid" shape).
2. If the flattening is due to positioning only, there will be frontal bossing on the same side (leading to a "parallelogram" shape).

What may be occurring if more than 1 suture is affected?

Involvement of more than 1 suture usually suggests a broader craniofacial syndrome. A genetics consult is recommended.

When is surgery indicated?

When there is increased intracranial pressure, progressive abnormality of the face or cranium, concern about compromise of visual function, or for significant cosmetic reasons

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Chapter 19 Gastrointestinal Disorders

SHORT-GUT (SHORT-BOWEL) SYNDROME

What is it?	Nutrient malabsorption and excessive intestinal fluid and electrolyte losses after massive small intestine loss or resection
List the 3 most common causes of extensive small bowel loss in children.	Malrotation with midgut volvulus; small intestine atresia(s); NEC (see p. 254)
How much small intestine does an infant have?	A full-term infant has ~250 cm of small intestine (an adult has 600–800 cm). Total intestinal length in a full-term infant is ~300 cm. The diameter increases from 1.5 cm during infancy to 3.5 cm in adulthood.
How much intestine does a child need to lose before developing short-gut syndrome?	There is no absolute amount of loss that defines short-gut syndrome. As much as 75% of the small intestine may be lost without serious long-term problems if the duodenum, terminal ileum, and ileocecal valve are spared. In general, infants with greater than 45–60 cm of residual small bowel should be able to survive without continued use of total parenteral nutrition. In contrast, the loss of 25% of the small intestine coupled with the loss of the terminal ileum and the ileocecal valve may cause significant difficulties.
What is the primary symptom?	Diarrhea
List 4 clinical results.	Growth failure, protein-calorie malnutrition, recurrent dehydration, and a variety of nutritional deficits

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What are other major risks?

Liver injury from cholestasis as a result of prolonged total parenteral nutrition. Increased risk of bacteremia and septicemia while receiving IV nutrition. Dysregulation of intestinal motility.

Does it matter which part of the bowel is lost?

Yes. Loss of much of the jejunum may cause few long-term symptoms because the ileum "adapts." The jejunum is less adaptable and unable to develop the specialized functions of any lost distal small intestine.

What conditions are caused by the loss of the terminal ileum or ileocecal valve?

Usually, **vitamin B_{12} deficiency** and **bile salt malabsorption.** Loss of the ileocecal valve may lead to bacterial contamination of the small bowel and poor regulation of flow of intestinal contents.

Describe the 2 phases of the treatment.

1. **Acute phase** (usually lasts several weeks after surgery): The child often has massive secretory diarrhea. Attention must be paid to fluid and electrolyte status. H_2 receptor antagonists or proton pump inhibitors may decrease intestinal secretion. Parenteral nutrition should begin as soon as possible to prevent catabolism.
2. **Chronic phase:** Goal is to support normal growth and development while maximizing intestinal adaptation. Maximal adaptation may take 6–12 months after surgery. Calories are provided totally or in part from parenteral nutrition during this period.

Is enteral nutrition beneficial?

Yes. It stimulates bowel growth and adaptation and may reduce the cholestasis associated with parenteral nutrition. Initial feeding may be an elemental or polymeric formula administered as a constant infusion through an NG tube or a gastrostomy tube.

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How are attempts at enteral feeding regulated?

The volume, concentration, and content of the formula are adjusted in response to the stool volume and clinical symptoms, such as feeding residuals, vomiting, and abdominal bloating.

What is the prognosis?

Outlook for long-term survival is good. Children who have >40 cm of small intestine and an ileocecal valve are usually ultimately capable of enteral nutrition alone.

NECROTIZING ENTEROCOLITIS

What is it?

An acute fulminating inflammatory disease of the intestine associated with focal or diffuse ulceration and necrosis of the small bowel, colon, and, rarely, the stomach

What are the common complications of NEC?

NEC is the most common cause of gastrointestinal (GI) perforation and acquired short-gut syndrome among hospitalized premature infants.

Which infants are most susceptible to NEC?

NEC is predominantly a disease of premature infants. The overall incidence is 3–5% of all neonatal intensive care unit admissions. Full-term infants rarely acquire NEC. If they do, it tends to involve the colon.

What is the pathogenesis of NEC?

The pathogenesis is multifactorial. It likely represents a final common pathway of the response of immature intestine to injury rather than a distinct disease.

List 5 implicated factors in NEC.

1. Ischemia-reperfusion injury of the intestine
2. Enteral alimentation
3. Infectious and inflammatory agents
4. An immature immune system
5. Immature intestinal mucosa

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What are the early GI signs and symptoms?

Early GI signs and symptoms are nonspecific and may include vomiting, delayed gastric emptying, increased gastric residual volume, hematemesis, bright red blood from the rectum, diminished or absent bowel sounds, abdominal distension with or without tenderness, and diarrhea.

What are the non-GI symptoms?

The infant may also exhibit a number of nonspecific non-GI symptoms consistent with bacterial sepsis, including apnea, respiratory distress, bradycardia, lethargy, temperature instability, cyanosis, mottling, systemic acidosis, and hyperglycemia or hypoglycemia.

List 5 complications that may appear as the disease progresses.

The infant may develop septicemia, DIC, hypotension, ascites, and intestinal perforation with peritonitis.

How is NEC diagnosed?

Primarily clinically. Confirmation can be provided by the radiographic presence of pneumatosis intestinalis (i.e., accumulation of gas within the intestinal wall), portal venous gas, or pneumoperitoneum.

What is the differential diagnosis?

Sepsis with ileus

Malrotation with midgut volvulus (see p. 294)

Pseudomembranous colitis (see p. 263)

Hirschsprung disease (see p. 293)

Gastric stress ulcer

Hemorrhagic disease of the newborn

Swallowed maternal blood

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What are the components of treatment?

1. Enteral feedings should be discontinued, and NG suction and IV fluids started.
2. Broad-spectrum parenteral antibiotics

Abdominal radiographs should be performed every 6 hours during the acute phase to detect perforation and to monitor the condition of the intestine.

Do most cases resolve with medical treatment?

Yes

When can enteral feeding be reintroduced?

After 7–10 days of medical therapy in uncomplicated cases

What are the 2 absolute indications for surgery?

Intestinal perforation or clinical deterioration unresponsive to medical therapy

What does surgery usually involve?

Resection of the perforated or necrotic bowel, an end stoma, and mucous fistula. The bowel may be reconnected when the infant is fully recovered.

Alternatively, if perforation is being addressed, placement of an abdominal drain may ameliorate systemic symptoms and allow resuscitation until a definitive procedure can be performed. Occasionally, the intestine will heal with drainage and laparotomy will not be needed.

What are the long-term complications of NEC?

1. **Short-gut syndrome:** if significant bowel resection
2. **Bowel stricture:** 18–25% of cases; mostly left colon. Infants treated medically with chronic or recurrent GI symptoms should undergo an upper GI series with follow-through and/or contrast (not barium!) enema, to look for strictures. Infants who are undergoing intestinal reconnection after surgery for NEC should have the remaining intestine assessed radiographically and intraoperatively for strictures.

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What is spontaneous intestinal perforation (SIP)?

This is a condition in which an isolated intestinal perforation occurs without any particular prodrome of symptoms. It is now recognized as an entity distinct from NEC.

Which infants are prone to this?

This occurs primarily in very low-birth-weight (VLBW) infants (<1,000 g).

What is the cause?

It is believed to be a vascular event. VLBW infants who have had umbilical artery catheters and who have received indomethacin may be at higher risk.

How is it recognized?

Typically by a sudden distension of the abdomen with free air on abdominal radiograph

How is it treated?

Bowel rest, broad-spectrum antibiotics, and surgical intervention

What are the surgical options?

1. Laparotomy with formation of a stoma and mucous fistula (some have reported performing primary repair)
2. Placement of a drain through a small incision. This may facilitate sealing of the perforation, or allow temporization until laparotomy is done if needed.

ULCERATIVE COLITIS

What is it?

An inflammatory bowel disease that involves rectal and colonic mucosa. Typically, the rectum is involved first, and the disease progresses proximally in a contiguous manner. Severe fulminant colitis usually involves the entire colon as "pancolitis."

What are the characteristics of the mucosa?

Crypt abscesses form, leading to mucosal ulcerations, pseudopolyps, and ultimately denuding of the mucosa. Inflammation is typically limited to the mucosal surface. With fulminant toxic megacolon, all layers of the colon (mucosa, submucosal, muscularis) may be involved.

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What are the etiologic factors?

Uncertain, but an immune-mediated process with a genetic predisposition is currently the most popular theory. Possible environmental trigger(s) remain elusive. About 15% of patients have a family member with inflammatory bowel disease.

What are the possible associated conditions?

Ankylosing spondylitis, uveitis, growth retardation, anemia, osteoporosis, nephrolithiasis, arthralgia, skin lesions (e.g., erythema nodosum, pyoderma gangrenosum), liver and biliary tract lesions (e.g., sclerosing cholangitis, fatty infiltration of liver), and aphthous stomatitis

What is the age of onset?

Usually adolescence or the third decade, occasionally earlier

What are the signs and symptoms?

The most common symptoms are crampy abdominal pain and persistent bloody diarrhea. In milder cases, signs and symptoms can be insidious—crampy abdominal pain can progress to diarrhea containing blood or pus. If the condition is unchecked, a toxic colitis (toxic megacolon) can ensue. Occasionally, this is the initial presentation.

List 2 components of diagnosis.

1. The diagnosis is largely one of exclusion and is confirmed by endoscopy and biopsy of colonic mucosa.
2. As many as 85% of children with ulcerative colitis have significant circulating titers of perinuclear antineutrophil cytoplasmic antibodies (p-ANCA).

What is toxic megacolon?

A fulminant presentation characterized by colonic dilatation, poor motility, and probable bacterial overgrowth. Patients are severely ill and may appear septic. Supportive therapy is needed with IV fluids, broad-spectrum antibiotics, and bowel rest. If medical therapy does not improve the condition, colectomy will need to be performed emergently.

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How is ulcerative colitis medically treated?

1. Initial treatment usually includes 5-aminosalicylic acid (5-ASA) derivatives given orally, such as sulfasalazine (Azulfidine), mesalamine (Asacol, Pentasa, Lialda), olsalazine (Dipentum), or balsalazide (Colazal), or mesalamine given rectally as suppositories (Canasa) or as enemas (Rowasa).
2. Corticosteroids may be given orally, rectally, or intravenously. Because of their side effects, corticosteroids should not be used chronically.
3. Other effective immunosuppressive agents are azathioprine, 6-mercaptopurine, methotrexate, cyclosporine, tacrolimus, and mycophenolate.
4. Anti–tumor necrosis factor antibody infusions (infliximab or Remicade) or injections (adalimumab or Humira) may be helpful.

Although broad-spectrum antibiotics are often used, there is little evidence to indicate they are effective. Antidiarrheal medicines should be used with care, because toxic megacolon may result. **Medical therapy is not curative.**

What is the surgical treatment?

Many patients need surgery. **The "curative" procedure is a total proctocolectomy with ileoanal pull-through or permanent end ileostomy. However, even after surgery, patients may be at risk for extraintestinal manifestations such as sclerosing cholangitis or ankylosing spondylitis.**

What are the outcomes?

Surgical removal of the colon and rectum is "curative" in that the primary target organ is removed. The pull-through procedure causes an increased number of bowel movements and may also result in urgency and occasional incontinence. But with appropriate training and medical support, patients may experience as few as 4–8 bowel movements daily often with resolution of incontinence.

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How is the large bowel at risk if not removed?

Initial reports estimated colon cancer risk to be 3–5% in the first decade of the disease and as high as 20% in each subsequent decade; however, subsequent population-based studies suggest the risk is probably one-tenth of this.

CROHN DISEASE

What is it?

An inflammatory bowel disease that may be transmural and marked by bowel wall thickening and scarring, ulcerations of the mucosa, and "skip lesions" (i.e., lesions separated by normal portions of bowel). Characteristic granulomas are identified in only 60% of patients.

What are the etiologic factors?

Uncertain, but an immune-mediated process with a genetic predisposition is currently the most popular theory. Possible environmental trigger(s) remain elusive. A number of mutations in genes regulating the immune response have been identified more commonly in individuals affected by Crohn disease. As many as 10% of affected individuals carry a mutation in the ***NOD2/CARD15* gene** that codes for an intracellular toll-like receptor. In active Crohn disease, the immune response appears to be directed at normal intestinal flora. This may explain why approximately 60% of affected patients have significant circulating titers of antibodies directed at the ubiquitous yeast *saccharomyces boulardii* **(ASCA).** About 10% of patients have a family member with inflammatory bowel disease. Among identical twins, if 1 twin is affected, the second twin has a 50% chance of developing disease within 10 years of the first twin being diagnosed.

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What is the epidemiology?

Incidence in boys and girls is the same. In the United States and the United Kingdom, the incidence among whites and blacks is now roughly the same. The overall incidence increased significantly during the past 60 years but appears to have leveled off over the past 10 years. Crohn disease is much more common in the developed world than in the developing world.

What age groups usually present with Crohn disease?

Adolescents and young adults

List typical signs and symptoms.

It can be insidious at onset. Typical symptoms include weight loss, abdominal pain, diarrhea, and fever. A perianal ulcer or abscess may be the initial manifestation. There may be rectal bleeding, but this is less frequent than in ulcerative colitis.

What extraintestinal manifestations may occur?

Growth failure—or growth deceleration—is common in children and often precedes any obvious GI symptoms. Other findings can include digital clubbing, delays in sexual maturation, skin lesions (most often erythema nodosum or pyoderma gangrenosum), asymmetric large joint arthritis, liver disease (sclerosing cholangitis), uveitis, and chronic hypochromic microcytic anemia.

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What are some options for medical therapy?

1. 5-ASA derivatives given orally, such as sulfasalazine (Azulfidine), mesalamine (Asacol, Pentasa, Lialda), olsalazine (Dipentum), or balsalazide (Colazal), or mesalamine given rectally as suppositories (Canasa) or as enemas (Rowasa)
2. Corticosteroids given orally, rectally, or intravenously. Because of their side effects, corticosteroids should not be used chronically.
3. Other effective immunosuppressive agents are azathioprine, 6-mercaptopurine, methotrexate, cyclosporine, tacrolimus, and mycophenolate. Anti–tumor necrosis factor antibody infusions (infliximab or Remicade) or injections (adalimumab or Humira) may be helpful.
4. Metronidazole may alleviate perianal or fistulous disease.
5. Ciprofloxacin

List 7 indications for surgery.

1. Failure of the medical therapy to treat symptoms adequately
2. Intestinal obstruction
3. Abdominal abscess
4. Enteric fistulae or fistulae to the genitourinary tract
5. Perirectal fistula or abscess
6. Perforation of bowel (rare)
7. Significant intestinal bleeding (rare)

What is the surgical strategy?

Surgery is used to treat a specific problem and usually involves resection of a symptomatic section of bowel or drainage of perirectal abscesses. **Surgery is a palliative, not a curative, procedure. Unfortunately, a large number of patients with Crohn disease will require surgery at some point during their lifetime.**

Can Crohn disease be cured?

There is currently no known cure.

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ANTIBIOTIC-RELATED COLITIS (PSEUDOMEMBRANOUS COLITIS)

What is antibiotic-related colitis?

A condition in which the normal intestinal flora is altered because of the use of antibiotic medicine

What is the primary symptom?

Watery diarrhea. Stools may be bloody in severe cases.

What is the most commonly identified pathogen?

Clostridium difficile

List 3 ways in which the condition is identified.

1. Identification of a pathologic organism by stool culture
2. Identification of the toxin of a pathologic organism (primarily *C. difficile*) in the stool
3. Sheets of WBCs are seen in the stool.

What is the treatment?

Discontinuing the antibiotic allows recovery of normal stool flora and function. If *C. difficile* is identified, metronidazole (by either mouth or IV) may be used for a cure. Other therapies include oral vancomycin, oral bacitracin, and oral bile salt binding resins such as cholestyramine or colestipol. There is emerging evidence that probiotics (*Lactobacillus rhamnosus* GG and *Saccharomyces boulardii*) may be effective.

CELIAC DISEASE (GLUTEN ENTEROPATHY/ CELIAC SPRUE)

What is it?

An acquired form of intestinal injury. In susceptible hosts, the ingestion of gluten (e.g., wheat gluten and other similar cereal proteins, particularly rye and barley) causes immunologically mediated damage to the small intestinal mucosa. Oats typically do not contain gluten; however, they may be contaminated with other grains during processing.

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How common is it?

Although it used to be thought celiac disease was quite rare, studies performed in blood donors suggest as many as 1% of the population of the United States may be affected by celiac disease. Obviously, the vast majority of affected individuals remain undiagnosed. This observation is often termed **"The Celiac Iceberg."** This analogy is used because the small percentage of overtly symptomatic patients represent the "tip of the iceberg" of a much larger population of people with celiac disease.

Who gets it?

Celiac disease is most common among whites, and it is much less common among people of African or Asian descent. Nearly all affected individuals are either **HLA DQ2 or DQ8 positive.**

At what age do patients typically present?

With classic disease, children typically develop symptoms between 6 and 18 months of age.

What is the significance of the age of onset?

It correlates with the introduction of wheat, rye, barley, and/or oats into the child's diet.

What are some common signs and symptoms?

Diarrhea, a protuberant abdomen, wasted extremities, decreased height and weight

What are other possible signs and symptoms?

Intermittent vomiting, irritability, abdominal pain, unexplained iron-deficiency anemia, unexplained osteopenia, dental enamel hypoplasia, peripheral edema, long eyelashes, digital clubbing, and rectal prolapse

Some children may present with isolated growth failure and an absence of GI symptoms.

Rarely, the presenting symptom is a vesicular skin eruption called "dermatitis herpetiformis."

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What is the differential diagnosis?

Other causes of intestinal malabsorption: CF, milk protein enteropathy, chronic giardiasis, Shwachman-Diamond syndrome, isolated pancreatic enzyme deficiencies, intestinal lymphangiectasia, abetalipoproteinemia, and chronic infections associated with immunodeficiency disorders

What are the 5 associated laboratory findings?

Most laboratory abnormalities in celiac disease are caused by chronic malabsorption. Iron-deficiency anemia; hypoproteinemia; deficiencies of fat-soluble vitamins (prothrombin time may be prolonged because of vitamin K deficiency); abnormal 72-hour fecal fat excretion and D-xylose absorption; IgA deficiency

List 3 ways it is diagnosed.

1. **The definitive test is a biopsy of the small intestine (typically duodenum)** revealing villous atrophy with hyperplasia of the crypts, abnormal surface epithelium, and inflammatory cells in the submucosa.
2. Among children who are NOT IgA-deficient, significant titers of tissue transglutaminase antibodies or endomysial antibodies are quite sensitive and very specific.
3. Full clinical remission after complete withdrawal of gluten from the diet

What is the treatment?

The cornerstone of the therapy is a **strict gluten-free diet,** in which wheat, rye, barley, and oats are excluded from the diet and substituted with rice and corn. Parents must read all food labels, because wheat byproducts are added to many foods. In severe cases, a **short course of corticosteroids** may help. It is also important to consider that gluten may be present in various medication preparations.

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How long do patients have to stay on a gluten-free diet?

Celiac disease is a lifelong disorder.

What are the long-term risks to affected individuals who continue to eat gluten?

Osteoporosis, iron-deficiency anemia, development of refractory celiac disease that no longer responds to gluten-free diet, and small-bowel lymphoma. There may be an increased risk of other autoimmune diseases including autoimmune diabetes mellitus, thyroiditis, and chronic active hepatitis.

GASTROESOPHAGEAL REFLUX IN INFANTS AND CHILDREN

What is it?

The effortless passage of gastric contents into the esophagus. This occurs frequently in most infants and children. In infants, GER may be heard ("wet burp") and/or seen ("spit up"). The volume of the refluxate can vary from a small "spit up," to what appears to be an entire feeding. In most infants, episodes of GER are mostly painless.

What are the primary symptoms of GER?

The most common symptom of GER in infants is painless and effortless regurgitation. Some infants may cry briefly prior to, during, and/or after episodes of reflux. Older children may complain of heartburn or water brash (a sour taste in their mouth). Some parents misinterpret regurgitation due to GER as true "vomiting."

Why does GER occur?

In most cases, GER is the result of transient spontaneous relaxation of the lower esophageal sphincter (LES) and/or an increase in intra-abdominal pressure sufficient to overcome LES pressure.

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Why are infants more prone to GER than older children?

1. Relatively large feeding volumes relative to their body size
2. Being fed a liquid diet
3. Spending a large amount of time in the supine and semisupine position
4. They have a relatively short and small esophagus.
5. They have relatively weak abdominal wall musculature that allows minimal abdominal pressure to overcome LES pressure.

In older children, diet (carbonated and caffeinated drinks, fatty foods), lifestyle (sedentary, obese), and constipation may all contribute to GER.

What is the difference between GER and GERD?

GER is a normal physiologic process. **Gastroesophageal reflux disease (GERD)** represents a smaller subset of patients that may have significant clinical symptoms or physical findings secondary to reflux of stomach contents into the esophagus.

List 5 symptoms concerning for infantile GERD.

1. Weight loss or poor weight gain
2. Refusing to eat
3. Severe irritability after eating
4. Hematemesis and/or hematochezia
5. Abnormal neck posturing (sometimes called **"Sandifer Syndrome"**)

How may GER be evaluated?

1. **History and physical examination:** in most cases can diagnose GER
2. **Upper GI series:** has very poor sensitivity and specificity for diagnosing GER but can be very useful in excluding anatomic abnormalities that might cause vomiting

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3. **pH probe:** may be helpful in correlating symptoms with acid in the esophagus. However, the frequency and duration of acid reflux as measured by a pH probe do not correlate well with the severity of symptoms in infants nor do they predict outcome.
4. **Modified barium swallow** (sometimes called a "radiographic assessment of feeding") is usually performed by a qualified speech pathologist in the radiology suite and may help identify abnormalities of oromotor function or swallowing mechanics.
5. **Endoscopic evaluation and biopsies:** may detect evidence of esophagitis and may help differentiate between reflux disease and other items on the differential diagnosis. The overwhelming majority of infants and children with GER will have normal endoscopies.

How is GER treated in infants?

1. **Educating and reassuring the family** are often all that are needed for simple, uncomplicated reflux.
2. **Time:** The majority of infants with physiologic reflux will "outgrow" their reflux by 15 months of life and most infants will experience improvement between 6 months and 1 year.
3. **Feeding adjustments:** Thickening the feedings with rice cereal or other simple starches has been shown to decrease the frequency and volume of regurgitation. Smaller, more frequent feeds, upright positioning, and frequent burping may help.

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Acid inhibitors (H_2 receptor antagonists and proton pump inhibitors): Although frequently prescribed, there is no evidence that these medications decrease the frequency or volume of regurgitation, and little evidence that they decrease fussiness or irritability, which is often attributed to GER.

In older children?

In older children, diet and lifestyle changes (decreasing fatty foods, reducing fatty bedtime snacks, treating constipation, reducing carbonated beverages, weight loss) may be all that is needed with intermittent, limited antacid therapy.

How is GERD treated?

Dietary and lifestyle changes are often recommended in conjunction with medical therapy.

In infants:

1. Feed an extensively hydrolyzed formula for 2 weeks and see if this lessens symptoms. Switching among various cows' milk formulas and soy formulas is rarely helpful.
2. Decrease acid production using either an H_2 receptor antagonist (ranitidine or famotidine) or a proton pump inhibitor (lansoprazole or omeprazole) to keep gastric (and therefore esophageal) pH above 4. These therapies treat the symptoms of GERD without treating the underlying dysmotility. Antacids (calcium carbonate) and topical agents (sucralfate) can also be used to decrease symptoms.
3. Motility agents, Metoclopramide (Reglan) and Erythromycin (a weak motilin receptor agonist) are often used to promote gastric emptying, but few studies have demonstrated clinical efficacy.

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Is there a role for surgery for GERD in infants and children?

Despite maximum medical therapy, a small subset of children will need surgery because they continue to suffer from GERD that may lead to respiratory insufficiency from chronic aspiration, inability to feed adequate calories, anemia, esophageal stricture formation and rarely, metaplastic changes of the esophagus **(Barrett's esophagus).** This may be especially true for children with underlying neurologic disease and/or profound developmental disabilities.

What operation is most commonly performed?

Nissen fundoplication. The fundus of the stomach is wrapped around the distal esophagus. This operation is performed laparoscopically or open as dictated by the patient's condition and anatomy. Fundoplication is a very effective means of eliminating reflux, with published success rates generally exceeding 95%.

Does GERD cause asthma?

This remains a very controversial topic. Although there is a small subset of children with asthma who may experience worsening of their respiratory symptoms due to GER, the majority of children and adults who suffer from asthma do not experience any improvement in their respiratory symptoms with aggressive treatment of GER.

PEPTIC ULCER DISEASE

What is it?

The disruption of the gastric or duodenal mucosal barrier by a combination of pepsin and gastric acid

Do children get ulcers?

Yes

What is the incidence?

Overall incidence in children is unknown. It is estimated that 3–4 in 10,000 pediatric inpatients have PUD.

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Do babies make enough acid to develop ulcers?

Yes—by 48 hours of life, most infants have a gastric pH between 1 and 3, and by 3 years of age, gastric acid secretion approximates adult values.

What are the common signs and symptoms?

The most common symptom is **abdominal pain,** often most severe at night. Children older than 6–7 years generally complain of classic epigastric pain. Younger children usually complain of generalized or periumbilical pain, and occasionally right lower quadrant pain. Less common symptoms include **vomiting, hematemesis, or melena.**

Is perforated ulcer common in children?

No, it is extremely rare.

What is the differential diagnosis?

Functional abdominal pain, irritable bowel syndrome, gastroesophageal reflux, cholelithiasis, pancreatitis, urinary tract infection or obstruction, lower lobe pneumonia, Crohn disease, ovarian cysts, appendicitis, and constipation

What is the most reliable method of diagnosis?

Flexible fiberoptic endoscopy. The sensitivity and specificity of double-contrast radiographic studies are only 60–70%.

List 3 treatment options.

Antacids to neutralize acid; H_2 receptor antagonists or proton pump inhibitors to inhibit acid secretion; sucralfate or bismuth compounds to provide a protective mucosal barrier. These therapies may be used individually or in combination.

How effective is medical treatment?

The most frequently prescribed therapy is a 6- to 8-week course of an H_2 receptor antagonist; it is effective in 85–95% of cases. There is emerging evidence that long-term use (>8 weeks) of proton pump inhibitors may not infer more benefit, may increase the risk of GI and respiratory infections in children and adults, and may increase the risk of osteoporosis in adults.

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What is the role of *Helicobacter pylori* in childhood PUD?

Unclear—about 15% of children undergoing endoscopy have evidence of *H. pylori* infection, but rates vary from 5% to 75%. Children with documented *H. pylori* infection should probably be treated to eradicate the organism.

INTUSSUSCEPTION

What is it?

A segment of intestine with its associated mesentery (the intussusceptum) telescopes into an adjacent segment of intestine (the intussuscipiens; see Fig. 19–1).

What is the typical age at presentation?

More than 50% of recognized cases occur between 3 and 12 months of age, and more than 75% occur before 2 years of age.

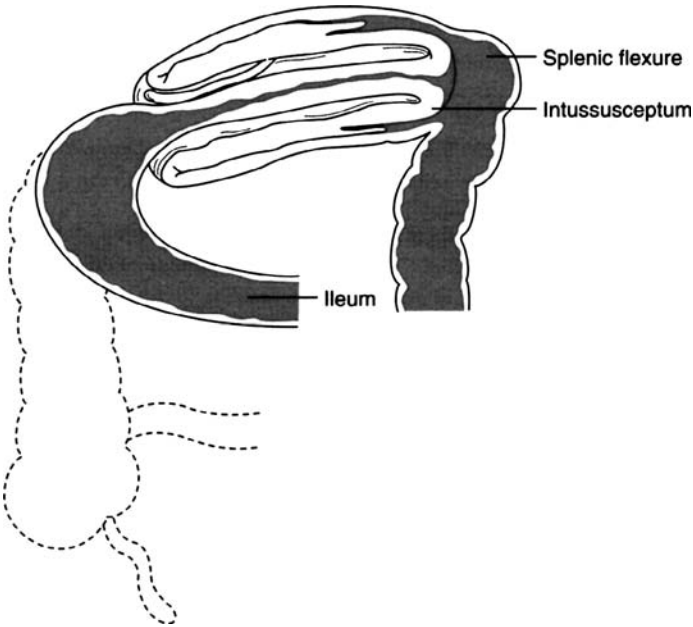

Figure 19–1. Development of intussusception.

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What segments of intestine are most commonly involved?

Most cases are ileocolic, with the intussusception starting immediately proximal to the ileocecal valve. Colocolic, ileoileal, and ileoileocolic intussusceptions are much less common.

What are the causes?

More than 90% of cases are idiopathic, without an identifiable "anatomic lead point." It is believed that most commonly, a viral illness causes hypertrophy of Peyer's patches in the terminal ileum, which serve as a lead point for the intussusception.

What is the most commonly identified lead point?

Meckel diverticulum (see p. 308)

List 6 disorders that seem to predispose a child to intussusception.

CF, Henoch-Schönlein purpura (HSP), Meckel diverticulum, juvenile inflammatory polyps, *Ascaris lumbricoides* (roundworm) infestation, and rotavirus infection

What are the signs and symptoms?

Sudden onset of episodic crampy abdominal pain, often with vomiting. Between episodes of pain, the child may be asymptomatic. Passage of stool and flatus is diminished. Passage of dark blood per rectum ("currant-jelly stools") suggests venous congestion and mucosal sloughing but is often not present at the initial presentation. The intussusception may be palpable on abdominal examination. As the symptoms progress, the child may become lethargic or somnolent.

How is it diagnosed?

The presentation is variable, and clinicians must maintain a high index of suspicion. Laboratory findings may be nonspecific or may suggest dehydration. Abdominal radiographs may be nonspecific or may suggest an abdominal mass, intestinal obstruction, or, rarely, free air. Ultrasound may be helpful in identifying intussusception. The diagnosis is usually established with an air or water-soluble contrast enema, which is usually also therapeutic.

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What is the treatment?

Diagnosis and treatment of an intussusception is by either **an air or a water-soluble contrast enema.** Pressure from the enema serves to reduce the intussusception. Imaging of the reduction is done with fluoroscopy (ultrasound in some centers). The child should receive IV hydration before the enema. Evidence of peritonitis or a general toxic state may be contraindications to the enema. A **surgeon should be immediately available** when an enema is performed in case the enema is unsuccessful or perforation occurs.

How often is an enema successful?

In about 80% of cases. The success rate is reduced if symptoms have been present > 48 hours.

What is the risk of perforation?

Approximately 1%

What is the incidence of recurrence after an enema?

Approximately 10% (usually within 48 hours of the initial episode)

What are the 2 indications for surgery?

Failure of reduction by air or contrast enema; signs or symptoms suggesting peritonitis or intestinal perforation, either at presentation or as a result of the enema

What is involved in surgery?

The intussusception is usually approached and reduced through an open incision, although laparoscopy may be used in some cases. An incidental appendectomy is typically done. Occasionally, the intussuscepted section must be resected because the bowel cannot be reduced during surgery, or because the intussuscepted portion is necrotic.

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ANORECTAL MALFORMATIONS (IMPERFORATE ANUS)

What is imperforate anus?

A spectrum of anomalies caused by an arrest of anorectal development during the cloacal stage between 4 and 12 weeks' gestation

How does it manifest in boys?

There is no true anal opening and, in most cases (90%), there is a fistula between the rectum and the urinary tract (urethra, the prostatic urethra, or the bladder). Fistula to the perineum is less common.

How does it manifest in girls?

There is no opening in the anal region. About 80% of patients have a fistula into the perineum, vaginal fourchette, or vagina.

How are high, intermediate, and low anomalies defined?

1. High: rectal atresia is above the levator sling.
2. Intermediate: atresia occurs at the level of the levator sling.
3. Low: atresia is below the levator sling.

On radiographs, the levator sling is determined by identifying the pubococcygeal line (upper border of the levator sling) and the line between the ischial tuberosities (lower border).

List 2 reasons the level of the anomaly is important.

1. The higher the anomaly, the less well formed are the sphincter mechanisms and the neurogenic innervation of these mechanisms.
2. High anomalies are also more likely to be associated with rectourinary fistulas in boys and high vaginal fistulas in girls.

Are boys or girls more prone to high anomalies?

Boys

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What other types of anomalies are common when imperforate anus is recognized?

Duodenal atresia, esophageal atresia, vertebral anomalies, renal anomalies, Down syndrome, congenital heart disease, anomalies of the limbs. Imperforate anus represents one manifestation of the VACTERL association (see p. 301).

How is this condition initially managed?

In boys?

Initial end colostomy with distal stoma (mucous fistula) formation is typically needed if there is no perineal fistula. The extent of the anomaly is then assessed with a contrast study through the mucous fistula into the atretic rectum. Voiding cystourethrogram (VCUG) is also needed to test for a possible rectourinary fistula.

Other studies assess for the commonly associated anomalies and the status of the spinal cord. These include limb and vertebral radiographs, abdominal ultrasound, echocardiography and MRI of the spinal cord and pelvis.

In girls?

If there is an external fistula to the perineum or vaginal fourchette, it can be dilated for the passage of stool until definitive repair is done. If the fistula opens into the vagina, a diverting colostomy should be considered. Assessment for other associated anomalies is needed as described earlier.

How is definitive treatment undertaken?

Usually, the posterior sagittal anoplasty (popularized by Peña) is used to reconstruct the anus. In boys, laparoscopic approach is used in some centers and is particularly useful when there is a fistula from the rectum to the bladder.

At what age is repair undertaken?

Most commonly, repair of imperforate anus takes place between 2 and 6 months of age. However, repairs are sometimes done in the neonatal period as a 1-stage procedure, that is, without a diverting colostomy.

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What are the outcomes?

Satisfactory continence is usually obtainable in infants with low-lying lesions. Continence may only be possible in about 50–75% of patients in intermediate and high lesions.

What are cloacal malformations?

A condition in girls that represents a common opening of the vagina and rectum, or the urethra, vagina, and rectum

What is the treatment?

Assessment is performed in a manner similar to that for imperforate anus. Colostomy is typically done initially. Ultimately, all 3 pathways must be reconstructed, usually from the posterior approach.

HENOCH-SCHÖNLEIN PURPURA

What is it?

HSP is a systemic vasculitis syndrome affecting small vessels.

At what age is HSP most common?

4–6 years of age, but can occur from 6 months to adulthood

Are boys or girls more commonly affected?

Boys slightly more than girls

Are there temporal or geographic predilections to HSP?

Increased incidence in the **spring** and **fall.** Most cases are sporadic, but temporal and geographic clusters occur.

What is the pathognomonic physical finding in HSP?

Virtually all patients have a characteristic skin rash: **palpable purpuric lesions** measuring 2–10 mm in diameter. Patients may also have coalescent ecchymoses and pinpoint petechiae. Typically, the purpura is concentrated on the buttocks and lower extremities, but it is not confined to those areas. New lesions appear in crops, and then fade over several days. In one-third of patients, other symptoms precede the onset of the rash by several days, making it difficult to establish the diagnosis.

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What is a common musculoskeletal manifestation?

Painful joint swelling (ankles, knees, and the dorsum of hands and feet) in approximately 80% of patients. The arthralgia is self-limited and usually abates with bed rest. Joint symptoms may precede the onset of rash in 20% of patients.

What are common GI manifestations?

Colicky abdominal pain, often accompanied by **vomiting,** in 50–75% of patients. Occult or gross **GI bleeding** is present in 40% of patients. **Intussusception** is reported in 1–5% of patients. Bowel infarction, perforation, and massive GI bleeding are rare but life-threatening complications. GI involvement before the appearance of the rash may mimic appendicitis or inflammatory bowel disease.

What is a common renal manifestation?

Nephritis in 50% of patients. Unlike joint or GI involvement, nephritis almost never precedes the onset of rash. Nephritis may be delayed for a number of weeks after the appearance of other symptoms, but usually becomes apparent within 3 months. It is manifested by microscopic or gross hematuria. Proteinuria is present in two-thirds of patients and hypertension in 25% of patients.

What are the less common complications?

Because HSP is a systemic vasculitis, any organ system may be affected. Other complications include intracranial bleeding, seizures, hemiparesis, coma, pancreatitis, hydrops of the gallbladder, orchitis, pulmonary hemorrhage, ocular involvement, and carditis.

How long does HSP last?

Average duration is **2–4 weeks.**

What is the risk of recurrence?

One-third of patients will have 1 or more recurrences.

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Who is most likely to have a recurrence?

Patients with nephritis

What is the usual nature of recurrence?

Usually occurs within 3 months of the original episode. Recurrences tend to mimic the original episode but are usually milder and of shorter duration.

List 2 characteristic histologic features of HSP.

1. Biopsies of purpuric lesions show polymorphonuclear leukocyte infiltration in and around dermal vessels (leukocytoclastic vasculitis).
2. Immunofluorescent studies demonstrate granular deposits of IgA in the walls of vessels.

What characterizes the histologic changes in the kidney?

They range from minimal change to focal or diffuse mesangial proliferation. The characteristic finding on immunofluorescence is diffuse mesangial IgA deposits.

How is HSP diagnosed?

On clinical grounds—there is no diagnostic laboratory test for HSP. Laboratory studies help to exclude other conditions that resemble HSP.

What laboratory studies should be done?

CBC and platelet count; urinalysis; and serum creatinine

List 4 characteristic serologic findings.

50% of patients have **increased serum IgA.** Antinuclear antibody (ANA), rheumatoid factor (RF), and antineutrophil cytoplasmic antibodies (ANCA) are negative.

What are the etiologic factors?

Unknown—the epidemiology of HSP suggests infectious etiologic factors, and a variety of organisms have been implicated. HSP may represent an unusual immune response to a variety of infectious or environmental insults. It is clear that IgA plays a pivotal role in the immunopathogenesis of HSP.

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What is the pathophysiology of HSP?	**Patients with HSP often have IgA-containing circulating immune complexes.** Deposition of these immune complexes in vessel walls results in inflammatory vasculitis and accounts for the histologic and clinical features of HSP.
What is the treatment?	The mainstay of therapy is supportive care.
Why must hypertension be treated?	To prevent intracranial bleeding
Which drugs should be avoided?	Salicylates and other drugs that interfere with platelet function should be avoided in patients with active GI bleeding.
Are steroids helpful?	Corticosteroids may alleviate joint and abdominal pain, but there is no evidence that corticosteroids affect the cutaneous purpura or hasten the resolution of the disease. Corticosteroids have no benefit in treating established nephritis.
What is the prognosis?	The prognosis is excellent for most patients.
Which manifestation is most prone to chronic problems?	Nephritis
What percentage of patients develop ESRD?	<5%
What patients are at the highest risk for developing renal failure?	Patients with gross hematuria, massive proteinuria, and hypertension during the acute phase of the illness
What follow-up may be useful in the patient who has recovered from HSP?	For a few months, weekly or biweekly evaluation of BP and urine for proteinuria may be useful to assess for the evidence of kidney damage.

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INTESTINAL TRANSPORT DEFECTS

What are they?

Isolated abnormalities of the absorption or secretion (i.e., transport) of specific ions or nutrients across the intestinal mucosa

What is the most common transport defect?

There are several defects that occur commonly. CF is the most common and is related to abnormal secretion of chloride across many different tissues, including the intestinal mucosa.

What is the general presentation?

Usually **chronic diarrhea.** However, symptoms of each defect are best explained by understanding which ion or nutrient cannot be absorbed or transported.

What is congenital chloridorrhea?

The chloride-bicarbonate exchange mechanism in the ileum and colon is dysfunctional, causing excessive chloride secretion with resultant secretory diarrhea and hypokalemic metabolic alkalosis.

What is congenital glucose-galactose malabsorption?

The active transport of glucose and galactose across the intestinal mucosa is defective; thus, the ingestion of any glucose or galactose causes osmotic diarrhea. Affected infants have severe diarrhea with profound growth failure.

What is X-linked hypophosphatemic rickets?

The renal and intestinal phosphate transport protein is defective, causing inadequate intestinal phosphate absorption, excessive urinary phosphate loss, and severe rickets.

What is the treatment?

There are no specific treatments for most transport defects. Instead, the patient's symptoms and the metabolic abnormalities caused by the specific transport defect are treated.

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APPENDICITIS

What is it?

Inflammation or infection, or both, of the appendix caused by occlusion of its lumen

What are some causes of occlusion of the lumen?

Stool (fecalith), inflamed or swollen lymphoid follicles, pinworm, or carcinoid tumors

What are the common symptoms?

1. Abdominal pain: starts as generalized pain caused by irritation of visceral pain fibers; pain migrates to right lower quadrant as worsening appendicitis causes local irritation of peritoneum.
2. Nausea and vomiting: usually follows onset of pain
3. Fever: usually, but not always present
4. Anorexia

What are the common findings on physical examination?

1. Localized guarding and pain to the right lower quadrant—particularly midway between the umbilicus and the anterior-superior iliac spine **(McBurney's point);** diffuse tenderness if appendix is ruptured. Tenderness may be less severe if the appendix is retrocecal or in another location if the appendix is malpositioned (e.g., malrotation). Localized tenderness may resolve temporarily at the time point of perforation. It will then return and likely become generalized.
2. Palpation on the left lower abdomen causes greater referred pain to the right lower abdomen **(Rovsing sign)**
3. Diminished bowel sounds
4. Pain on iliopsoas extension **(Psoas sign)**
5. Possible mass or tenderness anteriorly on rectal examination
6. Possible palpable mass in right lower quadrant if the rupture is contained

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List 6 potential diagnostic studies and what each shows.

1. WBCs: usually elevated with left shift
2. Urinalysis: a few (3–5) WBCs may be present secondary to local irritation of ureter or bladder.
3. Abdominal radiograph: usually normal, but it may show fecalith, ileus or sentinel loop, loss of fat stripe in right lower quadrant, slight concavity of spine to right, air-fluid level in right lower quadrant suggestive of abscess
4. Ultrasound: may show thickened appendix or mass consistent with an abscess. If imaging is felt to be needed, this is a good first choice to potentially avoid the radiation of the CT scan.
5. Contrast enema: nonfilling of appendix with irregularity of cecum
6. CT scan: best for delineating complex abscess; is also now used commonly in many centers to diagnose (or rule out) appendicitis.

What is the treatment for uncomplicated appendicitis?

Surgical removal of appendix with perioperative antibiotic coverage

For ruptured appendix?

Surgical removal of appendix and longer coverage with antibiotics (5–10 days)

For complex abscess?

May require initial drainage with later removal of appendix at about 6–8 weeks (interval appendectomy)

PANCREATITIS

What are the common causes of pancreatitis in children?

Infection, blunt trauma to the mid and/or upper abdomen, cholelithiasis, CF, and congenital anomalies (such as choledochal cyst or pancreas divisum). Pancreatitis can also be seen in patients with inborn errors of metabolism, such as methylmalonic aciduria. A wide variety of medications can also cause pancreatitis.

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List 4 common signs and symptoms.

Midepigastric pain and **tenderness, vomiting.** A **palpable epigastric mass** may be present with extensive inflammation of the pancreas or pseudocyst formation.

What diagnostic studies are used?

Laboratory studies include serum amylase, lipase, and urine clearance of amylase. The extent of the elevations of amylase and/or lipase does not correlate well with the severity of symptoms or the clinical outcome. **Ultrasound** and **CT scan** are the best initial imaging studies. Magnetic resonance cholangiopancreatography (MRCP) may help define ductal anatomy. Endoscopic retrograde cholangiopancreatography (ERCP) can be performed for both diagnostic and therapeutic purposes (this is usually done when the pancreatitis has subsided and a cause is still unknown).

List 3 components of treatment.

IV fluids, cessation of oral intake, pain management; meperidine or morphine are the medicines of choice. Slowly resolving or severe cases of pancreatitis may require a prolonged period of nothing per mouth with IV nutrition or placement of a feeding tube beyond the ampulla of Vater.

Is surgical treatment needed?

Pancreatitis usually resolves with medical treatment.

List 5 indications for surgery.

1. Persistent pseudocyst may require drainage via a percutaneously placed catheter, or internal drainage via cyst-gastrostomy, cyst-duodenostomy, or cyst-jejunostomy.
2. Chronic pancreatitis with pancreatic ductal dilatation may require a pancreaticojejunostomy (Puestow procedure).
3. Extreme hemorrhagic pancreatitis may require debridement of the pancreas.
4. Trauma may disrupt the main pancreatic duct, requiring placement of a stent via ERCP or distal pancreatectomy with attempted spleen preservation.

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5. Correction of congenital anomalies such as choledochal cyst or pancreas divisum that are causing pancreatitis

HEMOLYTIC UREMIC SYNDROME

What is it?

A clinical syndrome of microangiopathic hemolytic anemia, thrombocytopenia, and acute renal failure. **HUS is the most common cause of acute renal failure in children** (Ch 21, p. 328).

What causes HUS?

Two-thirds of cases are preceded by an infection with vero-cytotoxin–producing ***Escherichia coli* O157:H7.** However, HUS may occur after infection with other bacterial pathogens, including *Shigella*, *Campylobacter*, *Salmonella*, and *Yersinia*, and rarely, *Pneumococcus.*

What is the pathogenesis of HUS?

A variety of different bacterial toxins induce endothelial damage, which in turn initiates intravascular platelet activation, causing a diffuse small-vessel thrombosis throughout numerous organ systems. The use of antibiotics during acute infectious diarrhea was initially thought to increase the risk of HUS, but subsequent studies have brought this into question and the topic remains controversial.

What are the signs and symptoms?

1. 95% of HUS cases are preceded by gastroenteritis, and in nearly 75% of cases, the associated **diarrhea** is bloody.
2. Diffuse **abdominal pain** and **vomiting** are common.
3. Resolution of the GI symptoms is associated with the abrupt onset of **pallor, easy bruising, petechiae,** and **oliguria** secondary to acute renal insufficiency. (Do not confuse this with dehydration!)

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4. CNS symptoms are common, including irritability and encephalopathy. **Seizures** may occur and are usually caused by severe hypertension, hyponatremia, or both.

How is HUS diagnosed?

It is a syndrome diagnosed on clinical grounds.

What are the common laboratory findings?

1. Thrombocytopenia
2. Anemia
3. Elevated BUN and creatinine
4. Elevated hepatocellular enzymes (50% of patients)
5. Elevated serum amylase and lipase (25% of patients)
6. RBCs and WBCs present on stool smear
7. Positive stool cultures for *E. coli* O157:H7, *Campylobacter*, *Shigella*, or *Salmonella* (or *Yersinia* in some cases)

What are the radiographic and colonoscopic findings?

Contrast enema generally demonstrates intestinal ischemia with "thumb printing," mucosal irregularity, and ulcerations.

Colonoscopic findings are nonspecific—primarily hyperemic mucosal edema and friability.

Contrast enema and colonoscopy are not routinely performed unless the diagnosis is uncertain, as these maneuvers may exacerbate the colitis.

What is the treatment?

Supportive. Patients with severe GI symptoms receive **bowel rest** and **parenteral nutrition.** When clinically indicated, **RBC transfusions** and **diuretics** are administered, and **fluid intake is restricted.** Progressive renal insufficiency may require **peritoneal dialysis** or **hemodialysis.**

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What percentage of children recover renal function?	95% within 2–3 weeks
List 4 possible long-term complications.	Chronic renal insufficiency; intestinal fistulae or strictures; stroke with CNS deficits; chronic exocrine pancreatic insufficiency

CONSTIPATION

What is it?	A symptom, not a disease. It is the painful passage of large or hard bowel movements or the inability to expel a bowel movement.
What are normal childhood bowel habits?	
Infant?	Average stool frequency is 4 per day; 95% of children have from 1 stool every other day to 4 stools per day.
6 months to 2 years?	Average stool frequency is 2 per day; 95% of children have from 1 stool every other day to 4 stools per day.
Older than 2 years?	Average stool frequency is 1 per day; 95% of children have from 1 stool every third day to 3 stools per day.
What is the relation between constipation and frequency of bowel movements?	The frequency of defecation is influenced by diet and social custom. "Constipation" refers to the character of the stool and the symptoms associated with defecation rather than the frequency of defecation.
How common is constipation in children?	As many as 20% of children ≤5 years will be brought to medical attention because of constipation. **Constipation is the most common reason children are referred to a pediatric gastroenterologist.**

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List 3 main symptoms.

1. Pain associated with the passage of bowel movements
2. Bowel movements that are large, hard, or both
3. Infrequent bowel movements

See additional symptoms in the following text.

What are the other symptoms children may experience?

Intermittent crampy abdominal pain, abdominal distension, intermittent vomiting, early satiety, decreased appetite, "heartburn," water brash, urinary hesitancy or urgency

What are the physical signs?

A distended abdomen with palpable stool in the colon on rectal examination. Anal fissures may be present, and the rectum is generally enlarged and filled with stool. Rectal prolapse may be present. Chronic constipation is the most common cause of rectal prolapse in children.

Why do children develop constipation?

In otherwise healthy children, >99% of cases have no obvious cause and are said to have "functional constipation." In most children, constipation develops after the passage of several large or hard bowel movements with associated pain. This often occurs after weaning, a change in diet, school entry, an episode of gastroenteritis, or during toilet training.

What is the treatment?

The primary goal is to eliminate the pain associated with defecation. This generally means softening the stools.

In young children?

Fruit juices or dark corn syrup (such as Karo syrup) are often used; however, there is no good evidence supporting their efficacy. Osmotic cathartics, such as polyethylene glycol (MiraLax or GlycoLax), magnesium hydroxide (milk of magnesia), and lactulose, are safe and effective.

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In older children with chronic constipation?

Dietary measures are often inadequate, and laxatives polyethylene glycol (MiraLax or GlycoLax), magnesium hydroxide (milk of magnesia), mineral oil, lactulose, senna derivatives, and dioctyl sodium sulfosuccinate (Colace) must be used.

In the most severe and chronic cases?

High doses of oral cathartics or a series of enemas to evacuate the colon may be needed before oral laxative therapies can be effective. Rarely, manual disimpaction under anesthesia is required.

What are the risks of laxative use?

In an otherwise healthy child, the use of any over-the-counter laxative is safe. The major side effects of laxative overdosage are diarrhea, nausea and vomiting, and abdominal cramps.

Despite common belief, long-term use of laxatives does not cause dependency or "cathartic colon."

Is there a role of Botox injection for constipation?

Botox injection into the anal sphincter may allow temporary relaxation (about 1–3 months) of the sphincter. This may allow less painful passage of stool and recovery of a more normal bowel movement pattern. Noted success is still primarily anecdotal.

How are functional constipation and Hirschsprung disease differentiated?

In most cases, they can be differentiated on the basis of the history and physical examination. Functional constipation is far more common than Hirschsprung disease (see Hirschsprung disease, p. 293). Useful differentiating points are that constipated children will still pass flatus relatively easily, whereas children with Hirschsprung disease will not. Constipated children will often experience fecal soiling, whereas children with Hirschsprung disease rarely soil themselves. Constipated children will typically feed well and gain weight appropriately, whereas children with Hirschsprung disease will not.

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ENCOPRESIS

What is it?	Fecal incontinence or fecal soiling
How common is encopresis in children?	Among children older than 4 years, between 1% and 3% will be incontinent of stool more than once weekly.
What is the main cause of encopresis?	Prolonged constipation with resulting overflow incontinence
What are some of the other causes of fecal incontinence in children?	In rare circumstances, it is attributable to an underlying neurologic deficit (e.g., very low meningomyelocele or tethered spinal cord).
Is encopresis primarily a psychological problem?	Although many children with chronic encopresis have psychological and behavioral difficulties, many of these problems are a result, rather than the cause, of their fecal soiling.
What is a typical history?	There is usually at least a remote history of constipation. Soiling generally begins as small streaks of stool in the underwear. As the problem progresses, the volume and frequency of the soiling increase. The child denies any sense of the need to defecate before the accidents. If soiling occurs several times daily, it is often confused with diarrhea.
What are the features commonly found on physical examination?	A protuberant abdomen, with stool palpable throughout the colon. The rectal sphincter is often lax, and the rectum is large and filled with soft stool. An abdominal radiograph shows abundant stool throughout the colon.

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List 2 components of treatment.

Treatment is essentially the same as that for severe, chronic constipation.

1. The colon must be completely evacuated with large doses of oral cathartics, enemas, or by manual disimpaction.
2. When the colon is completely empty, laxatives are started in doses sufficient to produce 1 or 2 soft stools daily.

The child should be encouraged to sit on the toilet for 5–10 minutes after breakfast and dinner. Some children may benefit from biofeedback therapy.

FUNCTIONAL ABDOMINAL PAIN IN CHILDREN

What is functional abdominal pain?

Functional abdominal pain is one of a number of **functional gastrointestinal disorders (FGIDs)**. FGIDs include cyclic vomiting syndrome, irritable bowel syndrome, and functional constipation. With all these disorders, symptoms cannot be explained by known structural or biochemical abnormalities. The **ROME III** criteria can be used to further define these disorders.

What are the primary symptoms of functional abdominal pain?

By definition, the primary symptom is recurrent abdominal pain that has occurred at least once a week for 2 consecutive months. There must be no evidence of any anatomic, inflammatory, infectious, or neoplastic processes that could explain the pain. Depending on where the child localizes the pain and whether they have associated symptoms, recurrent abdominal pain may be subcategorized as functional dyspepsia (pain in the epigastrium), irritable bowel syndrome (pain associated with a change in bowel movements), or functional abdominal pain (pain usually in the periumbilical region and not associated with changes in bowel habits).

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Are there other symptoms?

As many as 20% of affected children have other somatic complaints. Headache, fatigue, chest pain, and pallor are particularly common.

How common is it?

The reported prevalence varies between 2% and 25%. Chronic abdominal pain accounts for at least 2% of pediatric office visits.

Is there any age or sex predilection?

Chronic abdominal pain is most common in children between **5 and 12 years of age.** In older children, chronic abdominal pain is much more common in **girls.**

Is functional abdominal pain the same as irritable bowel?

According to the ROME III criteria, functional abdominal pain and irritable bowel are 2 separate subgroups of abdominal pain-FGIDs. This is a theoretical construct and clinically there is a great deal of overlap. Typically, irritable bowel is characterized by improvement of symptoms following defecation, and the onset is associated with change in both stool frequency (constipation, diarrhea, or both) and stool consistency (hard, small or large, loose, watery).

What is the differential diagnosis?

The differential diagnosis includes constipation, Crohn disease, *H. pylori*–associated gastritis/duodenitis, pancreatitis, acid peptic disease, renal or urologic disease, pelvic inflammatory disease, biliary tract disease, abdominal lymphoma and abdominal migraine/migraine equivalent, undiagnosed malrotation, and chronic appendicitis (rare).

Does lactose intolerance cause chronic abdominal pain?

The prevalence of lactose intolerance is the same in asymptomatic children and children suffering from chronic abdominal pain.

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List 4 "red flags" or alarm signs suggesting the child's pain is not due to functional abdominal pain.

1. Blood in the stools
2. Bilious or persistent vomiting
3. Dysphagia
4. Involuntary weight loss

HIRSCHSPRUNG DISEASE

What is it?

Aganglionosis of the rectum or colon, causing a functional obstruction. The aganglionosis extends from the rectum proximally in a contiguous manner to some level, usually the upper rectum or left colon.

What is the transition zone?

The point where aganglionic bowel meets ganglionic bowel. It can be anywhere but is usually in the rectosigmoid region. Occasionally, the entire colon may be aganglionic, resulting in total colonic Hirschsprung disease.

What are the common signs and symptoms?

They may be insidious. Initial manifestation may be an infant's failure to pass meconium within the first 24 hours of life (95% of patients; see Ch 9, p. 86). Subsequently, there is increasing difficulty with passing stool and flatus, leading to severe stool and flatus retention, abdominal distention, overflow diarrhea, and sometimes enterocolitis and sepsis.

List 4 associated conditions.

Down syndrome; Waardenburg syndrome; cartilage-hair hypoplasia; neonatal appendicitis

How is it diagnosed?

1. **UNPREPPED single-contrast enema** is used to search for the transition zone. The ganglionic portion will be dilated. This may not be evident early in the course of the disease (i.e., in neonates).

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2. Definitive diagnosis is established by the absence of ganglion cells and increased acetylcholinesterase staining on a **rectal biopsy** specimen. Cholinergic and adrenergic nerve endings may also be evident. In infants, biopsy can be performed at the bedside or in the clinic with a suction biopsy device.

How is it treated?

1. Currently, operative correction (excision of aganglionated bowel and pull-through of ganglionated bowel to the anus) is undertaken in 1 stage through a transanal approach. Serial biopsies may be taken to determine the level of the transition zone in this manner. Laparoscopy or a small abdominal incision may facilitate multiple biopsies for faster identification of the transition zone, and may also facilitate mobilization of the bowel to be pulled through.
2. If a child is compromised at presentation (e.g., enterocolitis, malnutrition), a traditional approach may be warranted. A leveling colostomy (i.e., a colostomy immediately proximal to the level of the transition zone) is performed to allow bowel decompression and patient recovery. Later, a colon pull-through procedure (Swenson, Soave, Duhamel) may be performed.

What are the outcomes?

With appropriate surgical care, 85% of patients will have normal or nearly normal bowel function. Others may have functional motility difficulties despite the presence of ganglion cells.

MALROTATION

What is it?

Failure of the gut to make its normal 270-degree counterclockwise rotation during in utero development

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What are the 3 anatomic results?

1. Unrotation of gut to varying degrees with the ligament of Treitz to the right of, or at, the midline, and a mobile cecum
2. Ladd's bands
3. Narrow mesenteric pedicle

What are Ladd's bands?

Peritoneal attachments of the now-mobile ascending colon to the right abdominal wall. They may stretch across the duodenum, causing obstruction.

What is the most dangerous aspect of malrotation?

The narrow pedicle may cause **volvulus** and subsequent loss of part or all of the bowel. **This may be lethal!**

At what age does a patient present with malrotation?

At any age! However, about 80% of patients with malrotation who have symptoms do so within the first 4 months of life.

List 4 conditions in which malrotation is commonly found.

Diaphragmatic hernia; gastroschisis; omphalocele; duodenal and intestinal atresia

List 3 potential symptoms.

1. Vomiting, usually of bilious material **(Vomiting of bilious material in an infant ≤ 4–6 months is malrotation with volvulus until proven otherwise!)**
2. Intermittent abdominal pain
3. Systemic collapse if volvulus has progressed to frank bowel necrosis

List 4 potential signs.

Patient may have a distended abdomen; usually, no peritoneal irritation unless bowel injury is present; dehydration; weight loss. **However, an infant with volvulus may look "normal" with the only symptom being bilious vomiting!**

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What is the definitive diagnostic study?

An **upper GI series** will identify a malposition of the ligament of Treitz and associated volvulus if present. **This study must be done emergently if an infant presents with bilious vomiting.** A contrast enema may show malposition of cecum, but the position may be normal even if the cecum has inappropriately oriented peritoneal attachments.

What is the treatment?

Surgical correction with the Ladd's procedure

What is involved in the Ladd's procedure?

Ladd's bands are divided, the colon is mobilized to the left with the cecum situated near the sigmoid colon, the duodenum is mobilized and straightened, and an appendectomy is performed.

What is done if volvulus is present?

The bowel is turned **counterclockwise** on its mesentery until the volvulus is relieved. If there is necrotic bowel, this is resected, and anastomoses or stomas are performed as appropriate.

What is the outcome?

Usually very good. However, if volvulus is serious, an extensive loss of bowel may result in short-gut syndrome.

CONGENITAL DUODENAL OBSTRUCTION

What is it?

An obstruction of the duodenum secondary to failure of the recanalization process of the fetal duodenum. It occurs during the eighth to tenth week of development.

List the 3 most common types of congenital duodenal obstruction.

Duodenal atresia; duodenal stenosis; duodenal web

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What is annular pancreas?

A failure in the proper rotation of the pancreas may cause an annular pancreas, which also causes duodenal obstruction. Some researchers theorize that annular pancreas is an anatomic phenomenon secondary to one of the primary duodenal conditions noted earlier.

What is the usual location of duodenal obstruction?

The first or second part of the duodenum. It may involve the entrance of the common bile duct.

What are the typical signs and symptoms of duodenal obstruction?

Vomiting, which may be bilious or nonbilious depending on where the bilious drainage is relative to the obstruction; abdominal distension secondary to distended stomach and duodenum

What are the 2 prenatal ultrasound findings?

Polyhydramnios and a **dilated duodenum**

What is the characteristic radiographic finding?

"Double-bubble" sign of dilated stomach and duodenum

How is a definitive diagnosis made?

A contrast study shows total or partial duodenal obstruction with a rounded dilated duodenum (as opposed to the beaklike appearance found in malrotation with volvulus).

Which conditions and malformations are sometimes found with congenital duodenal obstruction?

One-third of affected infants have Down syndrome. Other anomalies include intestinal atresia, malrotation, imperforate anus, cardiac anomalies, and other anomalies of VACTERL association (see p. 301).

How is it treated preoperatively?

NG drainage with rehydration

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What are the 3 surgical options?

Surgical correction may be done semielectively unless malrotation is suspected, which makes repair more urgent.

1. Duodenoduodenostomy (most common)
2. Vertical duodenotomy through an involved web or stenosis with transverse duodenoplasty
3. Duodenojejunostomy (rarely done)

Malrotation or other atresias must be sought at operation.

What is the outcome?

Usually good, although return to full feeding is often slow as the duodenum recovers motility.

INTESTINAL ATRESIA

What is it?

A congenital condition in which the lumen of the bowel is interrupted

Where may an intestinal atresia occur?

Anywhere from the jejunum to the rectum; atresias may be multiple.

List 5 classifications and their characteristics.

Type I: The mucosa is interrupted by a web, but the proximal dilated loop of bowel and decompressed distal loop are still connected at the serosal level.

Type II: The proximal dilated bowel and distal decompressed bowel are connected by a fibrous atretic cord.

Type IIIa: The proximal and distal portions of bowel are separated, as is the mesentery to these two portions of bowel.

Type IIIb: The distal atretic bowel is spiraled around a segmental artery in an apple-peel form.

Type IV: Multiple atresias are present (Fig. 19–2).

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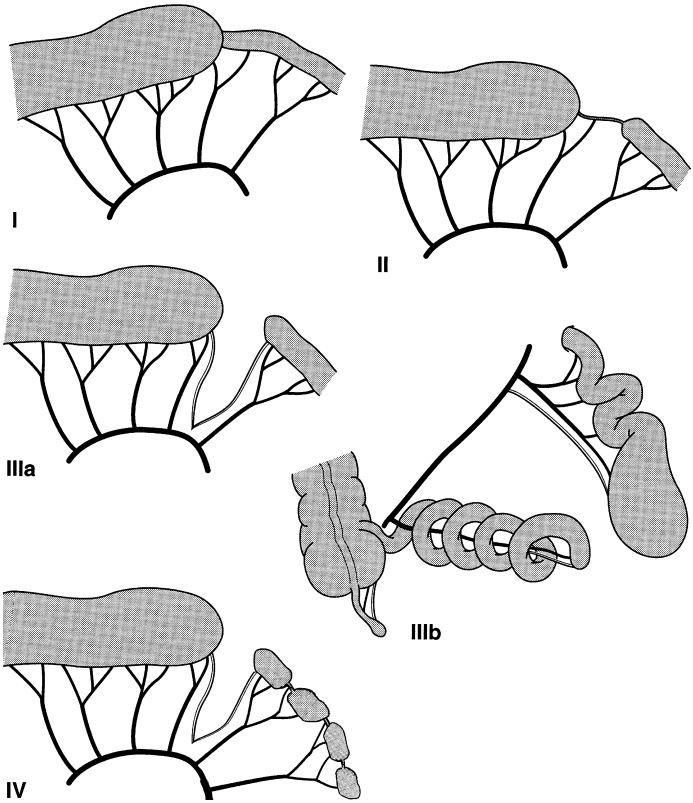

Figure 19–2. Classification of intestinal atresia.

How do they occur?

They are believed to be the result of a vascular accident in utero.

How may a patient present?

1. Newborns generally develop a distended abdomen soon after birth, if it is not already present at birth.
2. Bilious vomiting ensues, or if an NG tube has already been placed, voluminous bilious output is present.
3. Infants may pass meconium because atresias often develop after meconium has passed to the distal bowel in utero.
4. Rarely, the proximal portion of bowel may perforate in utero.

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What is the differential diagnosis?

Malrotation with volvulus, bowel duplication, internal hernia, adynamic ileus with sepsis, meconium ileus, Hirschsprung disease, small left colon syndrome

What is the method for diagnosis?

A contrast enema is performed in the newborn presenting with evidence of distal bowel obstruction. A microcolon will be visualized without connection to the proximal dilated bowel.

What is the treatment?

Surgical correction is necessary.

What is involved in surgical correction?

For jejunal, ileal, or proximal colonic atresia, resection of the dilated portion of the proximal bowel and the atretic tip of the distal bowel is performed, followed by anastomosis in most cases. If the bowel caliber discrepancy is too great, or the atresia is in the distal colon, an end stoma is typically performed first, and later an anastomosis is performed.

What is the outcome?

The current survival rate is 90–100%.

ESOPHAGEAL ATRESIA OR TRACHEOESOPHAGEAL FISTULA

What is it?

A spectrum of anomalies consisting of discontinuity of the esophagus with or without fistula(e); or a fistula between the esophagus and the trachea with esophageal continuity maintained (i.e., isolated fistula)

List the 5 types and their characteristics.

Type A: esophageal atresia without fistula

Type B: esophageal atresia with fistula between the upper portion of esophagus and trachea

Type C: esophageal atresia with fistula between the lower portion of esophagus and trachea

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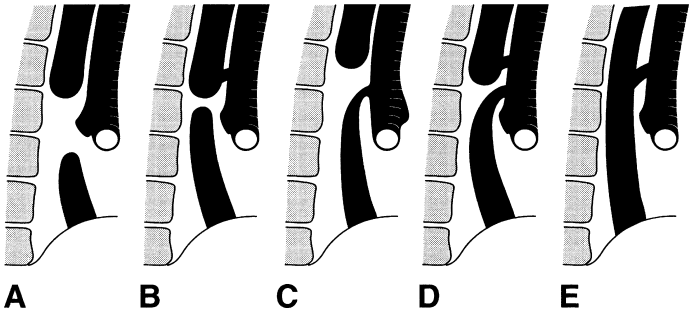

Figure 19–3. Tracheoesophageal fistula.

Type D: esophageal atresia with a fistula between the upper portion of esophagus and trachea and a fistula between the lower portion of esophagus and trachea

Type E: esophagus in continuity with isolated fistula between esophagus and trachea

(Fig. 19–3)

Which type is most common?

Type C (85%)

What is the incidence?

1 in 3,000 births, with an equal incidence between boys and girls. Prematurity is common among patients.

What is the VACTERL association?

A constellation of anomalies that commonly occur together, either entirely or in part, more often than would be expected by "chance."

Vertebral

Anorectal (imperforate anus)

Cardiac

Tracheal

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Esophageal

Renal or genitourinary

Limb or lumbar

Infants need to be screened for all of these anomalies when any one of them is present.

List 2 other anomalies that are commonly associated with TEF.

Duodenal atresia and bowel atresias

What are the symptoms of TEF?

Types A, B, C, D?

Excessive drooling; feeding induces choking, coughing, regurgitation, and cyanosis.

Type E?

Presentation usually delayed; feeding induces coughing and choking; repeated episodes of pneumonia

What are the chest and abdomen radiographic findings for each type?

Type A: dilated upper esophageal pouch with gasless abdomen

Type C: dilated upper esophageal pouch with normal bowel pattern

Types B, D, E: may be normal

How is it diagnosed?

Types A, B, C, D (list 3 components):

1. Inability to pass suction catheter beyond upper esophagus
2. Presence of gas in bowel indicates fistula between distal esophagus and trachea.
3. Instillation of thin barium into esophageal pouch confirms atresia of esophagus and assesses for the presence of a proximal fistula. Bronchoscopic examination may be used to determine the presence of proximal fistula.

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Type E:

Barium swallow; physician may need to instill barium into esophagus with patient in prone Trendelenburg position to demonstrate fistula as it typically courses in a cephalad direction from the esophagus to the trachea.

What is the treatment?

Type A:

1. Placement of gastrostomy tube
2. Drainage of the upper pouch with a Replogle tube, and attempt at primary closure at around 12 weeks of age after the esophagus has had time to grow.
3. If anastomosis of esophagus is not possible, options include esophagostomy with later colon interposition, jejunal interposition, gastric pull-up, or gastric tube formation.

Types B, C, D:

1. Thoracotomy via fourth intercostal space
2. Extrapleural approach
3. Division of fistula(e) with anastomosis of esophagus
4. Repairs of type C have been reported using thoracoscopic technique.

Type E (List 1):

Division of fistula via right cervical incision

List 4 possible complications.

Infection, stricture of the esophagus, leak of the esophageal anastomosis, recurrence of the fistula

What feeding precautions are necessary for children with repaired TEF?

The esophagus motility is impaired. Ultimately, feeding is relatively normal, but chunky foods should be avoided until the child can chew well. The esophagus caliber is usually large enough to handle all foods by that time.

What may be a long-term consequence of TEF?

Most of these patients have gastroesophageal reflux. Acid inhibitors may be helpful, and there may be an emerging role for esophageal surveillance to assess for mucosal changes.

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PYLORIC STENOSIS

What is it?	Hypertrophy of the pyloric muscle, causing gastric outlet obstruction
What is the incidence?	1 in 500–1,000. Male-to-female ratio is 4:1.
What are the etiologic factors?	Unknown. Suggested processes include decreased number of ganglion cells, hypergastrinemia, edema secondary to feeding, and a decrease in nitric oxide synthase. It is probably a "multifactorial" condition.
What is the most common symptom?	Progressive, projectile, **nonbilious** vomiting that occurs after feeding (The infant is then very hungry again.)
List 4 physical signs.	1. Abdominal protuberance possibly present secondary to gastric distension 2. Gastric waves visible through abdominal wall 3. Palpable pylorus deep in epigastrium ("olive sign") 4. Dehydration sometimes present
What metabolic abnormalities may be noted?	Dehydration; hypokalemic, hypochloremic metabolic alkalosis; and hypoglycemia. Rehydration and correction of electrolytes are necessary before surgery. Potassium and glucose need to be included judiciously in resuscitative fluid.
List 2 ways in which it is diagnosed.	Ultrasound (the best study); upper GI series: the "string sign" (i.e., a string of contrast passing through pylorus) is seen. (**Caution:** Pyloric spasm may mimic pyloric stenosis on upper GI series. An upper GI series can definitively rule out pyloric stenosis but cannot definitively rule it in.)
How is it treated?	Ramstedt pyloromyotomy via open incision or laparoscopic technique.
What is the outcome?	Excellent. Babies begin feeding 4–6 hours after surgery.

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INTESTINAL POLYPS

What are they?	Tumors that protrude into the lumen of the bowel
What are the most common polyps in children?	Juvenile inflammatory polyps (sometimes called "juvenile retention polyps")
What are juvenile inflammatory polyps?	Mucosal lesions consisting of dilated and tortuous mucus-filled glands, with a prominent inflammatory infiltrate in the lamina propria. The glands are composed of well-differentiated, mucus-secreting cells.
What size are they?	The polyps are erythematous pedunculated masses 0.5–3.0 cm in diameter.
List 2 other characteristics.	They are often quite friable and bleed when manipulated.
Where in the bowel are they most commonly found?	Although juvenile inflammatory polyps may develop anywhere in the large intestine, nearly two-thirds are located in the distal colon beyond the splenic flexure.
Are polyps solitary or multiple?	Either. More often they are multiple.
Are juvenile inflammatory polyps considered precancerous?	No
What is the most common symptom?	Intermittent painless rectal bleeding in children 1–10 years of age. The blood is generally bright red and either streaked on or intermixed with the stool. A polyp may rarely be a lead point for intussusception.
List 5 less common clinical features.	Intermittent abdominal pain; vomiting; colocolonic intussusception; prolapse of the polyp through the anal canal; iron-deficiency anemia

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How common are juvenile inflammatory polyps?

They are the most commonly identified cause of painless rectal bleeding in children 1–10 years of age.

How are they diagnosed?

Although polyps can be identified on air-contrast enema, flexible colonoscopy is the test of choice. Polypectomy can usually be safely performed with snare electrocautery through the scope.

Must all juvenile inflammatory polyps be removed?

Most ultimately outgrow their vascular supply and autoamputate. The diagnosis is sometimes first suspected when a child passes a polyp in the stool. Because polyps are often asymptomatic and have no malignant potential, colonoscopic intervention is not always warranted.

What other conditions may be associated with intestinal polyps in children?

Peutz-Jeghers syndrome, adenomatous polyposis coli (sometimes called "familial polyposis coli" or "familial adenomatous polyposis"), Cowden syndrome, and Gardner syndrome

Can these polyps become malignant?

Yes. To prevent malignancy, total colon resection with ileoanal pull-through may be needed (especially in adenomatous polyposis coli and Gardner syndrome).

BEZOAR

What is it?

A mass of ingested material, usually hair and vegetable matter that has congealed and settled in the stomach

What is the cause?

It usually results because of children (more commonly boys) eating their own hair. (Often, emotionally disturbed children will do this, although not all develop bezoars.)

List 5 symptoms.

Nausea, vomiting, early satiety, inability to eat, weight loss

What are the physical signs?

Mass in epigastrium or left upper quadrant

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How is it diagnosed?

Usually with an upper GI series

List 2 treatments.

1. Removal of the bezoar is performed via open gastrotomy.
2. Meat tenderizer or removal with endoscopy may be used for small bezoars.

Note: Any underlying emotional disorders should be evaluated and treated.

PICA

What is it?

Persistent eating of significant amounts of nonnutritive substances (e.g., dirt, clay, paint chips)

Does pica always indicate a serious problem?

Not always. During the first 2 years of life, mouthing and eating a wide variety of objects is normal exploratory behavior. In many older children and adults, the chewing and eating of nonnutritive substances (e.g., fingernails, pencils, ice cubes) represent a habit rather than a serious medical problem.

What are the causes?

The pathophysiology is not understood. Pica is a symptom, not a disease, and its presence may indicate an underlying disorder. Pica is most commonly observed in children with developmental disabilities, autism, or mental retardation.

What are the complications of pica?

Complications are related to the substances ingested. The most serious complications of pica are **intestinal obstruction** and **lead poisoning.** Iron-deficiency anemia is seen in some patients.

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MECKEL DIVERTICULUM

What is a Meckel diverticulum?	A remnant of the embryonic vitelline or omphalomesenteric duct that is a true diverticulum (contains all layers of bowel wall)
What is the incidence?	2% of the population
Where is it located?	The antimesenteric border of the ileum, usually within 2 ft of the ileocecal valve in an adult
What types of ectopic tissue may be present?	Pancreatic or gastric in 25% of cases
What are the symptoms of Meckel diverticula?	Most are asymptomatic throughout life. However, symptoms may include: 1. Profuse bleeding per rectum (the most common symptom) 2. Abdominal pain caused by inflammation 3. Obstruction caused by intussusception
What causes bleeding from a Meckel diverticulum?	Erosion of mucosa opposite the diverticulum caused by production of acid from ectopic gastric mucosa
How is it diagnosed?	A symptomatic Meckel diverticulum can often be detected with a technetium-99m pertechnetate scan ("Meckel scan"), which images gastric mucosa. It may sometimes be discovered during laparotomy or laparoscopy for nonreducible intussusception or peritonitis, or incidentally at surgery.
What is the treatment?	Surgical excision of the diverticulum with primary bowel closure
What is a Littré hernia?	An umbilical or inguinal hernia containing a Meckel diverticulum

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PERIANAL AND PERIRECTAL ABSCESS

What is a perianal abscess?	An infected subcutaneous collection in the perianal region
Who is most commonly affected in the pediatric population?	Infants
What are the most common causes?	An infected diaper rash, or a small mucosal tear in the anal canal from a large or firm stool.
What are the typical signs and symptoms?	A firm, red, fluctuant area in the perianal region. The infant may have a fever and may be irritable.
List 2 components of treatment.	1. Lancing the area for drainage of pus 2. Antibiotics may be necessary to resolve the surrounding cellulitis.
What is a perirectal abscess?	An infected collection that extends within the intersphincteric region
What are the signs and symptoms?	Fever and perirectal pain. External signs may be minimal initially because the infection is more recessed from the perianal region.
Who is most commonly affected in the pediatric population?	True perirectal abscesses are rare in the pediatric population. They are likely most common in children with other predisposing conditions.
List 2 components of the treatment.	1. Lancing for adequate drainage of pus, and possible drain placement 2. Antibiotics are necessary for perirectal abscesses.
Can perianal or perirectal abscesses recur?	Yes. If they do recur, then a fistula is present.

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How are these treated?

A probe is placed through the skin opening to the open anal crypt. The overlying skin and muscle are divided and the fistula is curetted. Very deep perirectal abscesses may be better treated with a Seton wire.

List 4 conditions that can predispose older children to perianal and perirectal abscess.

Crohn disease (see p. 260), leukemia, immunodeficiency disorders, diabetes

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Chapter 20 Liver and Hepatobiliary Disorders

CHOLELITHIASIS (GALLSTONES)

What are the 3 types of gallstones?	Cholesterol; pigmented; mixed-type stones
Which stones are most common in children?	**Cholesterol stones**
What causes cholesterol stones?	An imbalance of **lecithin, bile salts,** and **cholesterol** in bile
List 3 causes of pigmented stones.	1. Breakdown products from blood in hemolytic diseases or after heart surgery 2. Abnormal absorption of bile after ileal resection 3. Cholestasis resulting from TPN
What is the incidence of gallstones in children?	2 cases per 1,000 children
List 5 risk factors for gallstones in neonates and infants.	Prematurity, ileal resection, CF, TPN, prolonged fasting. Some neonates have idiopathic stones.
What are the predisposing conditions in older children?	Hemolytic disorders or idiopathic cholesterol gallstones
List 3 ways in which a patient with gallstones presents clinically.	1. **Biliary colic:** intermittent right upper quadrant or epigastric pain, associated with eating, which results from transient obstruction of the cystic duct by a stone, or by passage of a stone through the biliary system to the intestine

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2. **Cholecystitis:** inflammation of the gallbladder secondary to persistent cystic duct obstruction from a stone
3. **Obstruction of the common bile duct (CBD):** may cause persistent right upper quadrant or epigastric pain, jaundice, acholic stools, dark urine, and cholangitis

What is the most useful imaging study?

Ultrasound is best for detecting stones within the gallbladder as well as for evidence of extrahepatic biliary ductal dilatation.

List 5 useful laboratory values and what they show.

1. **Serum bilirubin** may be elevated in hemolytic disorders, CBD obstruction, or cholecystitis.
2. **Alkaline phosphatase** may be elevated with CBD obstruction.
3. **Hepatocellular enzymes** may be elevated in cases of cholestasis, cholecystitis, or prolonged CBD obstruction.
4. **γ-Glutamyl transpeptidase** may be elevated in the presence of an obstructing CBD stone.
5. **Elevated serum amylase** may indicate the presence of associated pancreatitis or CBD obstruction, or both.

List and describe 2 kinds of treatment.

1. **Surgery:** Laparoscopic removal of the gallbladder (with an intraoperative cholangiogram when indicated to define biliary tree anatomy or assess for CBD stones). CBD stones may be removed via endoscopic retrograde cholangiopancreatography (ERCP) before (preferred) or after surgery. If ERCP is not available, CBD stones may be removed at surgery through the cystic duct or by opening the CBD. This may be done with advanced laparoscopic skills or through an open incision. Surgical CBD exploration is not usually done anymore.

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2. Expectant management: In an infant or child who develops sludge or gallstones from TPN cholestasis and who is otherwise asymptomatic, stones may be expected to resolve after the TPN has been discontinued. In addition, infants with idiopathic stones can usually be observed because these stones are a result of the mother's metabolic state and will usually pass or resolve without symptoms and not recur.

HEPATITIS

What is hepatitis?	Inflammation of the liver (hepatocytes)
Are all types of hepatitis infectious?	No
List 5 causes of hepatitis other than viruses.	Trauma, metabolic diseases (e.g., galactosemia and α_1-antitrypsin deficiency), Reye syndrome, vascular obstruction, chemical toxicity (including certain medications)
What is chronic hepatitis?	Persistent inflammation of the liver
List 3 causes of chronic hepatitis.	Infection, immune (autoimmune) disorders, metabolic disorders
Which hepatitis viruses are associated with chronic hepatitis?	Usually the parenteral viruses: hepatitis B, C, and D
How do chronic active hepatitis and chronic persistent hepatitis differ?	Histologically, chronic active hepatitis involves the limiting plate of the portal triad, whereas chronic persistent hepatitis does not.
Which has a better prognosis?	Chronic persistent hepatitis

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What viruses cause hepatitis?	Hepatitis viruses A, B, C, D, and E; herpes simplex virus; varicella-zoster virus; cytomegalovirus; Epstein-Barr virus; adenovirus; rotavirus; enterovirus; rubella virus; parvovirus; influenza viruses
How does a child with viral hepatitis present?	Variable. The manifestations may range from subclinical ("anicteric") to overwhelming liver necrosis and failure.
What is the typical clinical presentation of symptomatic viral hepatitis?	
In the preicteric phase? (list 5 symptoms)	Fever, malaise, loss of appetite, abdominal pain, right upper quadrant tenderness
Icteric phase?	Jaundice, light (clay-colored) stools, dark urine (secondary to bilirubinuria), increased serum levels of hepatic transaminases. **An elevated prothrombin time/INR and a decreasing serum albumin level indicate there is decreased hepatic synthetic function and this reflects more severe liver injury.**

HEPATITIS A

How is it spread?	Usually via the fecal-oral route
List 3 sources of infection.	Contaminated water; foods (including raw shellfish); person-to-person contact (particularly among younger children)
What is the common name for hepatitis A?	Infectious hepatitis
What is the chance of an infected child passing the disease to another household member?	About 10–20%
What is the incubation period?	About 4 weeks, although it can range from 10 to 50 days

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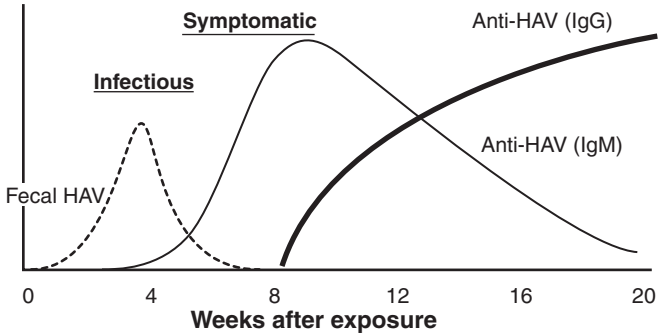

Figure 20–1. Pattern of response to hepatitis A virus (HAV) infection. IgM, immunoglobulin M; IgG, immunoglobulin G. (Reprinted with permission from Behrman RE, Kliegman RM, Jenson HB, eds. Nelson Textbook of Pediatrics. 16th ed. Philadelphia: WB Saunders, 2000:770.)

How is a child's presentation different from an adult's?

Children are much more likely to have anicteric hepatitis, with subclinical disease. Most children with serologic evidence of prior hepatitis A infection have no history of clinical jaundice beyond the neonatal period.

See Figure 20–1 for the pattern of response to hepatitis A virus (HAV) infection.

What is the laboratory diagnosis?

Demonstration of IgM anti-hepatitis A antibodies in the serum

Is demonstration of IgG anti-hepatitis A antibodies useful?

Somewhat. These titers rise later than IgM and may persist, so the positive IgG antibody does not necessarily represent acute infection.

List 3 ways in which hepatitis A infection can be prevented.

1. Good hygiene practices
2. Good public health measures including vaccination against hepatitis A
3. Immune globulin injections within 2 weeks of exposure can prevent infection or reduce disease.

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Is there a vaccine for hepatitis A?	Yes, it is given in 2 parts. Current recommendations are that the first immunization be given at 12 months of age and the second 6–18 months after the first immunization.
Does hepatitis A lead to chronic hepatitis?	Rarely
What is the treatment of hepatitis A?	Usually supportive

HEPATITIS B

What is another name for hepatitis B?	Serum hepatitis
List 3 usual routes by which it is spread.	Usually via **parenteral** routes, including blood and blood products; sexual transmission; maternal-child transmission
What is hepatitis Be antigen?	A secreted soluble antigen that is highly associated with infectivity
What is hepatitis B surface antigen (HBsAg)?	The major envelope protein of the virus
What is hepatitis core (HBc) antigen?	The major protein in the viral capsid. See Figure 20–2 for the pattern of response to hepatitis B virus infection.
Which antigen correlates best with infectivity?	Hepatitis Be antigen
What is the risk of transplacental infection to the infant of a known hepatitis B–infected mother?	The risk depends on the mother's hepatitis Be antigen status. If positive, the risk of transplacental infection is 65–85%. If negative, the risk is 10–20%.
When does mother-to-infant transmission usually occur?	During delivery

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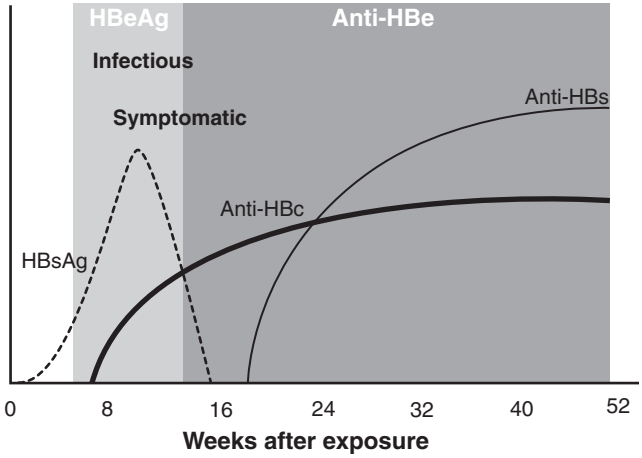

Figure 20–2. Pattern of response to hepatitis B virus infection. HbeAg, hepatitis Be antigen; HBsAg, hepatitis B surface antigen; HBc, hepatitis B core antigen. (Reprinted with permission from Behrman RE, Kliegman RM, Jenson HB, eds. Nelson Textbook of Pediatrics. 16th ed. Philadelphia: WB Saunders, 2000:772.)

What is meant by "chronic carrier state"?	Patients infected with hepatitis B who have a persistent viral infection
Who are most likely to become chronic carriers?	Infected infants
List 3 risks to chronic carriers.	Persistent infectivity; chronic liver disease; hepatocellular carcinoma
How is hepatitis B infection usually diagnosed?	Usually via serologic testing for **serum antibodies against HBsAg and HBc** and for the **presence of the HBsAg**
What is the first serum marker of hepatitis B infection?	Presence of HBsAg
Which antibody usually appears first after an acute hepatitis B infection?	Anti-HBc (detectable at around 15 weeks)

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List 4 ways in which hepatitis B infection can be prevented.

1. Universal precautions for health care workers
2. Decreased exposure by high-risk individuals
3. **Hepatitis B vaccine**—available and effective
4. **Hepatitis B immune globulin** for passive immunization

List 2 components of treatment for an infant born to an infected mother.

Hepatitis B immune globulin; hepatitis B vaccine within 12 hours of birth

Is hepatitis B vaccine recommended for all infants?

Yes

What is the expected immunologic response to the hepatitis B vaccine?

Antibodies are developed to HBsAg, but not to the core or e antigen. Antibody titer may wane over a long period of time, but the immunologic protection from the vaccine may be maintained.

Is there any effective treatment for chronic hepatitis B infection?

Interferons (INF alfa-2b and pegylated INF alpha-2a) appear to have limited effectiveness. Lamivudine, adefovir, and entecavir may induce remission but there is evidence of emerging resistance and combination therapy may prove better than monotherapy. Remission rates remain relatively low with therapy, so prevention is the best treatment strategy.

HEPATITIS C

List 3 ways in which it may be spread.

1. Usually via parenteral routes (including blood transfusion)
2. Possibly via sexual transmission
3. Mother-to-infant transmission—estimated to occur in <10% of cases, which is far less common than with hepatitis B

List 2 ways it is usually diagnosed.

Serology: enzyme-linked immunosorbent assay (ELISA); polymerase chain reaction (PCR)

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When does the ELISA test become positive, if infection is present?	It may be up to 8–12 weeks after the infection before the ELISA is positive, although it may be positive earlier.
What is the treatment?	Prolonged therapy with interferon alpha (INF-α) and pegylated interferon alpha in combination with Ribavirin may lead to sustained virologic response in up to 80% of patients infected with genotypes 2 and 3 but only 40% of patients infected with genotype 1.
What is the risk for chronic liver disease?	Risk may be as high as 70% of infected patients, although there is emerging evidence that the risk of progression is much lower in otherwise healthy children and young adults.

HEPATITIS D

What is another name for the hepatitis D virus?	Delta virus
By what route is it usually transmitted?	Usually via blood products
What is its relationship to hepatitis B infection?	Infection with hepatitis B virus (either previous or concurrent) is required.
In what way is it diagnosed?	Demonstration of anti-hepatitis D virus antibodies

HEPATITIS E

How is it spread?	Fecal-oral route
What is the incubation period?	2–9 weeks
How is it diagnosed?	Diagnosis is usually based on epidemiology and exclusion of other viruses.

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BILIARY ATRESIA

What is it?	Abnormality of the intrahepatic or extrahepatic bile ducts, or both, in which the ducts are microscopic in size, fibrous cords, or completely absent
What is the incidence?	1 in 15,000
List 5 associated defects.	Congenital heart disease (Ch 16); absent inferior vena cava; preduodenal portal vein; intestinal malrotation (Ch 19, p. 294); and polysplenia
What are the etiologic factors?	Unknown at this time. Biliary atresia appears to be a condition acquired after birth and may be caused by a reovirus infection. It appears to be an inflammatory process.
What is so-called correctable biliary atresia?	Biliary atresia in which the intrahepatic and proximal (i.e., those closer to the liver) extrahepatic ducts are patent. This is not common.
What is the natural history of biliary atresia?	Biliary duct obstruction with progressive cirrhosis, portal hypertension, hepatomegaly, and jaundice. Infants with uncorrected biliary atresia are likely to die before 2 years of age.
List 6 presenting signs and symptoms.	Jaundice, hepatomegaly, acholic stools, dark urine, elevated direct bilirubin and alkaline phosphatase
How is it diagnosed?	Ultrasound may show a contracted or absent gallbladder and no evidence of extrahepatic bile ducts. Nuclear scan shows obstruction of biliary flow. Definitive diagnosis is made at laparotomy.
What is the treatment?	**Surgical correction** is the first-line treatment.

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Describe the 2 components involved in surgical correction.

1. **Resection** of the atretic gallbladder and biliary ducts up to the point of the liver that lies within the branches of the portal vein (the periportal plate). This is normally the area where the common hepatic duct would branch into the right and left hepatic ducts.
2. Roux-en-Y **hepaticojejunostomy** with the jejunum anastomosed directly to the periportal plate

What are the outcomes?

Approximately 33% of infants have a successful outcome; 66% of infants ultimately require liver transplantation for survival.

When is the surgical correction of biliary atresia most likely to be successful?

Before the infant is 8 weeks of age and when bile flow is established after the operation

What is the surgical procedure typically called?

The Kasai portoenterostomy

CHOLEDOCHAL CYSTS

What are they?

Abnormal dilatations of the extrahepatic biliary system, the intrahepatic biliary system, or both; thought to be congenital

What are the etiologic factors?

Unknown. However, there is often an abnormal entrance of the pancreatic duct into the CBD above the sphincter of Oddi. It is postulated that reflux of pancreatic enzymes may cause the cysts.

List the 5 types of choledochal cysts and their characteristics.

Type I: cystic dilatation of the CBD

Type II: diverticulum extending off of the CBD

Type III: a choledochocele at the level of the sphincter of Oddi

Type IV: cystic dilatation of the extrahepatic and intrahepatic CBDs

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Type V: single or multiple dilatations of the intrahepatic bile ducts

Who is most prone to getting choledochal cysts?

Females and people of Asian ancestry

How does an infant present with choledochal cysts?

Jaundice is usually the first presenting sign.

Older children?

A classic triad of **abdominal pain, jaundice,** and a **palpable mass** has been described. However, abdominal pain and jaundice are the 2 most common presenting signs. Older children may present with **pancreatitis.**

List 3 typical laboratory findings.

Elevated bilirubin, alkaline phosphatase, and amylase (especially with bile duct obstruction)

What are the most useful diagnostic studies?

Ultrasound is the most useful initial study. CT scan or magnetic retrograde cholangiopancreatography (MRCP) are usually done to better delineate the anatomy. ERCP may be an alternative imaging modality in older children but this is not commonly needed.

What is the treatment?

Surgical excision of the gallbladder and the dilated portion of the CBD and formation of a Roux-en-Y choledochojejunostomy. If the cyst cannot be removed in its entirety, the inner lining of the cyst should be shelled out from the outer wall. This approach works for types I, II, and IV. A type III cyst is usually simply unroofed within the duodenum. Type V cysts represent the most difficult surgical challenge. These are usually drained into a Roux-en-Y jejunal limb.

What is the risk of leaving choledochal cyst tissue behind?

Cyst epithelium can develop **adenosquamous carcinoma.** Type V cysts are the greatest threat for this occurrence, because they cannot be entirely removed.

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What are the outcomes?

The result of surgery for types I, II, III, and IV cysts is quite good. Patients with type V cysts have an increased risk of developing carcinoma and often have recurrent cholangitis. Liver transplant may be considered for some of these patients.

PORTAL HYPERTENSION

List the 3 types of obstruction that may cause portal hypertension in children.

Extrahepatic obstruction (portal vein thrombosis); intrahepatic venous obstruction; suprahepatic venous obstruction (Budd-Chiari syndrome)

What is the most common type of portal hypertension in children?

Extrahepatic (portal vein thrombosis)

List 6 common causes of extrahepatic portal hypertension in children.

Neonatal omphalitis (Ch 10, p. 106), intra-abdominal infections, dehydration (Ch 3, p. 16), umbilical vein catheterization (UVC) (Ch 8, p. 66), enterocolitis, and congenital abnormalities of the portal area

What are the causes of intrahepatic portal hypertension?

Usually caused by **cirrhosis,** which may be secondary to the following conditions: biliary atresia (see p. 320); congenital hepatic fibrosis (see p. 325); cystic fibrosis (Ch 17, p. 258); α_1-antitrypsin deficiency; radiation or chemotherapy changes; hepatitis (see p. 313); sclerosing cholangitis; histiocytosis X; galactosemia; congenital biliary cirrhosis; hepatic hemangioma (Ch 27, p. 452); glycogen storage disease; cholestasis from prolonged total parenteral hyperalimentation; Alagille syndrome

What are the common causes of suprahepatic portal obstruction (Budd-Chiari syndrome)?

In most cases, the cause cannot be identified. Occasionally, oral contraceptives and granulomatous disease (usually outside the United States) may be causes.

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List 8 common signs and symptoms of portal hypertension.

Hepatomegaly; esophageal variceal hemorrhage; splenomegaly with possible subsequent hypersplenism; ascites; caput medusa; spider telangiectasia; palmar erythema; ultimately, liver failure

TREATMENT

How is portal hypertension caused by portal vein thrombosis treated?

Anticoagulation is the initial therapy. If a portosystemic shunt is required, a **distal splenorenal** or **mesocaval shunt** are the 2 most common shunt operations used.

What is a typical complication of portal hypertension from portal vein thrombosis?

Esophageal variceal bleeding is usually the most troublesome complication but rarely requires emergent operative intervention. Usually, the patient is hospitalized and given **IV fluids, vitamin K, H_2** inhibitors, and **blood transfusion,** if necessary, to provide adequate support until the bleeding stops. In some cases, **injection of a sclerosing agent,** via direct visualization of the varices, is required.

How is portal hypertension caused by liver cirrhosis treated?

The ultimate course of treatment depends on the prognosis of the hepatic disease. Variceal bleeding is treated with **sclerosing agents.** Worsening hypertension may require a **distal splenorenal or a mesocaval shunt, or TIPS (transjugular intrahepatic portosystemic shunt) procedure.** In some cases, **liver transplantation** is required.

How is portal hypertension caused by suprahepatic obstruction treated?

Shunts from the portal system to the right atrium are occasionally successful. **TIPS procedure may be considered.** In most cases, **liver transplantation** is the treatment of choice.

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CONGENITAL HEPATIC FIBROSIS

What is it?

A disease characterized by diffuse periportal and perilobular fibrosis. These may form ductlike structures but do not communicate themselves with the biliary system.

List 3 associated conditions.

Renal tubular ectasia; autosomal recessive polycystic renal disease; and nephronophthisis

What are the 2 signs and symptoms?

This condition usually becomes evident in early childhood. **Hepatosplenomegaly** and **esophageal variceal bleeding** secondary to portal hypertension are the characteristic conditions.

What is the treatment?

Usually the hepatocellular function is normal. Treatment focuses on **control of esophageal bleeding.** Bleeding episodes can be controlled by **supportive care** or **sclerotherapy** using endoscopy. In some cases, a **portosystemic shunt,** TIPS procedure, or liver transplantation may be required.

What are the outcomes?

With appropriate intervention, liver function may remain good. However, associated renal conditions may limit long-term survival.

ALAGILLE SYNDROME

What is it?

An inherited disorder characterized by neonatal cholestasis caused by paucity of intrahepatic ducts, cardiac abnormalities, skeletal abnormalities, ocular abnormalities, and a characteristic facial configuration

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What causes it?

Alagille syndrome is inherited as an autosomal dominant trait. The majority of affected individuals have abnormalities of the *JAGGED-1* gene on chromosome 20. A smaller number of affected individuals have abnormalities of the *NOTCH2* gene on chromosome 1.

How do affected children usually present?

With cholestatic liver disease in the neonatal period

What is the most common cardiac abnormality associated with Alagille syndrome?

Peripheral pulmonic stenosis. Less common lesions include pulmonary valve stenosis, atrial septal defect, ventricular septal defect, tetralogy of Fallot, and patent ductus arteriosus.

What other abnormalities can be present?

As they get older, many children develop a characteristic triangular face with a broad forehead, deep-set eyes, pointed chin, and elongated nose with bulbous tip. Butterfly vertebrae are present in 50%, and smaller numbers have abnormalities of the ribs and hands. Seventy-five percent of affected individuals have ophthalmologic findings, most typically posterior embryotoxon, which is prominence of Schwalbe's line just inside the temporal limbus.

What should be included in the differential diagnosis?

Other causes of neonatal cholestasis including biliary atresia, α_1-antitrypsin deficiency, CF, metabolic diseases, bacterial infections particularly with group B streptococcus or gram-negative bacteremia, and TORCH infections

How is the diagnosis made?

The definitive diagnosis is usually made by liver biopsy, which demonstrates intrahepatic cholestasis and paucity of intrahepatic bile ducts with an intact extrahepatic biliary tree.

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How do we treat it?

Treatment is supportive and includes prevention or correction of fat-soluble vitamin deficiencies **(vitamins A, D, E, K)** and management of pruritus when it occurs. In approximately half of affected children, the cholestasis improves as they get older. Some children go on to develop end-stage liver disease and require transplantation.

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Chapter 21 Renal Diseases

ACUTE KIDNEY INJURY

What is it?	A reversible increase in the concentration of creatinine and nitrogenous waste products in the blood and the inability of the kidney to normally regulate fluid and electrolyte homeostasis (has been historically termed "acute renal failure")
What are the most common causes of acute kidney injury?	
In infants? (list 6)	Sepsis, asphyxia, hypotension, congenital heart disease (as a complication of surgery), congenital urinary tract anomalies, renal arterial thrombi
In older children? (list 4)	Hemolytic-uremic syndrome (HUS); acute glomerulonephritis (AGN; usually poststreptococcal); trauma; sepsis
What is the appropriate urine output for children?	Infants: ~2 mL/kg per hour Toddlers: ~1–2 mL/kg per hour School-age children: ~1 mL/kg per hour Adolescents and adults: ~0.5 mL/kg per hour
What is oliguria?	Urine output that is below the appropriate amount for a given age group
List the 3 classifications of oliguria.	1. **Prerenal:** volume depletion or poor cardiac output 2. **Renal (or intrinsic):** glomerular or tubular injury 3. **Postrenal (or obstructive):** congenital or acquired urinary tract obstruction (must be bilateral)

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How can one differentiate between prerenal oliguria and intrinsic renal failure in infants?

In prerenal oliguria, the kidney responds to hypoperfusion by increasing sodium and water reabsorption. The urine is concentrated (specific gravity > 1.010) and the fractional excretion of sodium is low (<1%). When glomerular or tubular damage has occurred, these functions are not maximized. The urine will be isosthenuric (specific gravity = 1.010), and fractional excretion of sodium will be high (>2%).

What are the cardiac manifestations of hyperkalemia?

Peaked T waves are seen first, followed by prolonged PR intervals, flattened P waves, and widened QRS complexes. Terminally, ventricular tachycardia and fibrillation develop.

What is the treatment of hyperkalemia?

Membrane stabilization: Calcium gluconate (100 mg/kg IV) or calcium chloride (10 mg/kg IV)

Redistribution: Sodium bicarbonate ($NaHCO_3$; 1–2 mEq/kg IV) drives K^+ into cells in exchange for H^+ extruded to buffer the bicarbonate. β-Agonists (10 mg of albuterol by nebulizer) as well as insulin (0.1 U/kg IV) plus glucose (0.5 g/kg IV) stimulate cellular uptake of K^+.

Removal: Na^+-K^+ exchange resin given by mouth or by rectum binds K^+ for later excretion; **dialysis** is the most effective method of removing K^+.

Why are patients with acute kidney injury commonly hypertensive?

Fluid overload is the most common cause of hypertension, although acute glomerular diseases such as AGN and HUS are also associated with **high renin** output.

How is hypertension best treated in renal injury?

Appropriate fluid management is mandatory; diuresis if possible, and dialysis if necessary

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Vasodilators (e.g., calcium-channel blockers, sodium nitroprusside, diazoxide)

Angiotensin-converting enzyme inhibitors (must be used with care as they exacerbate decreased glomerular filtration rate)

What is the proper fluid replacement prescription for a child with acute kidney injury?

Combine insensible fluid losses, urine output, extrarenal fluid losses (e.g., nasogastric drainage, stool losses), and estimated losses of Na^+ and other electrolytes.

What methods of dialysis are used for children with acute kidney injury?

1. **Peritoneal dialysis (PD)** is the most common, although hemodialysis may be feasible for larger children and adolescents.
2. **Continuous venovenous hemodiafiltration** uses a blood pump to drive fluid and electrolyte transfer across an extracorporeal membrane. It is most commonly used for children with extreme fluid overload, those with unstable cardiovascular status, or children in whom PD is not possible.

CHRONIC KIDNEY DISEASE

What is creatinine clearance (Ccr)?

An estimate of glomerular filtration rate:

Ccr = UV/P × 1.73/SA = mL/min per 1.73 m^2, where

SA = body surface area (m^2)

U (mg/mL) = urinary creatinine concentration

V (mL/min) = total urine volume (mL) divided by time (min)

P (mg/mL) = serum creatinine

A correction factor of 1.73 is used to normalize adult and child CCr values because a normal adult body surface area is 1.73 m^2.

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What is a normal Ccr for an infant?	A normal newborn's Ccr is ~20 mL/min per 1.73 m^2.
At what age is the adult level reached?	Normal adult values (80–120 mL/min per 1.73 m^2) are reached by 2 years of age.
What level of Ccr denotes ESRD?	Ccr < 10 mL/min per 1.73 m^2
What are the most common causes of chronic kidney disease or ESRD in the pediatric population?	
In infants and preschool children? (list 4)	Congenital structural anomalies, obstruction, hypoplasia, dysplasia
In older children and adolescents?	**Acquired glomerular diseases,** including glomerulonephritis, HUS, reflux nephropathy, and systemic lupus erythematosus **Inherited disorders,** including Alport syndrome and polycystic kidney disease
How well do children with advanced chronic kidney disease grow?	Poorly, both in weight gain and linear growth
List 7 potential causes of poor growth.	Steroid treatment, protein losses (in nephrotic syndrome), sodium wasting, chronic acidosis, renal osteodystrophy, recurrent illness, malnutrition
List 5 treatments of growth failure.	Aggressive nutritional support, medical management of electrolyte abnormalities, dialysis, recombinant growth hormone. **Early renal transplant may be recommended.**

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What is renal osteodystrophy?

Bone demineralization caused by decreased renal function, leading to decreased production of vitamin D and elevated parathyroid hormone levels; demineralization is often severe enough to impair growth and increase the risk of fracture.

Why are patients with advanced kidney disease anemic?

Because failing kidneys decrease the production of erythropoietin

How is anemia treated?

Administration of subcutaneous or IV recombinant erythropoietin reduces the need for transfusions.

List 5 hallmark electrolyte abnormalities in chronic kidney disease.

Hyperkalemia, uremia (increased BUN), hyperphosphatemia, hypocalcemia, acidosis

List 3 treatment options for children with ESRD.

1. **Peritoneal dialysis** (the favored modality for children) uses the peritoneal membrane for exchange with dialysate and may be done in an automated fashion by the parents at home.
2. **Hemodialysis** is harsher, requires in-hospital treatments, and uses needles or large-bore venous catheters.
3. **Renal transplantation** (either from living or deceased donors) provides long-term and more physiologic renal replacement; there may be problems with rejection or recurrent disease.

NEPHROTIC SYNDROME

What 4 features characterize it?

Edema, proteinuria (>4 mg/kg per hour), hypoalbuminemia (<2.0–2.5 g/dL), hypercholesterolemia (>200 mg/dL)

List 6 diseases that may cause nephrotic syndrome.

Minimal-change disease, focal segmental glomerular sclerosis, membranoproliferative glomerulonephritis, membranous nephropathy, systemic lupus erythematosus, Henoch-Schönlein purpura (Ch 19, p. 277)

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What is minimal-change disease?

A form of nephrotic syndrome in which light microscopy and immunofluorescence are normal, and electron microscopy shows only effacement of the podocyte foot processes. Approximately 85% of children with nephrotic syndrome have minimal-change disease.

What is the source of the edema in nephrotic syndrome?

Although not completely understood, edema is most likely attributable to decreased plasma oncotic pressure secondary to protein loss into the urine. Fluid leakage into the extravascular space also causes decreased perfusion pressure, which results in increased sodium and water reabsorption by the kidney.

How is nephrotic syndrome treated?

Greater than 95% of children with minimal-change disease go into remission with corticosteroid therapy within 4 weeks. Two-thirds of patients will have relapses, some frequently. Adverse effects of steroids, steroid dependence, or both may lead to treatment with cytotoxic agents or calcineurin inhibitors. When nephrotic syndrome is a secondary process, treatment or resolution of the primary process is usually curative.

What is the prognosis of minimal-change disease?

Usually resolves spontaneously after puberty without renal dysfunction

What are the most common complications of nephrotic syndrome?

Infection: Pneumococcal and gram-negative infections are the most common. Increased susceptibility is attributable to poor nutrition, loss of immunoglobulins, and immunosuppressive therapy.

Hypercoagulability: Hyperlipidemia, urinary loss of anticoagulant proteins, and intravascular volume depletion all contribute.

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GLOMERULONEPHRITIS

What is the difference between nephrosis and nephritis?	**Nephrosis (or nephrotic syndrome):** proteinuria, hypoalbuminemia, and edema; implies no inflammation **Nephritis:** hematuria, decreased CCr, and hypertension; implies renal inflammation. Many chronic glomerular diseases present a picture of nephritis with or without nephrosis.
List 4 features of the typical presentation of acute poststreptococcal glomerulonephritis.	Gross hematuria, hypertension, mild edema, and decreased renal function, 7–14 days after a skin or throat infection with **group A streptococcus**
What is the laboratory profile of poststreptococcal glomerulonephritis?	**Decreased C3 (third component of complement) and elevated anti–streptococcal titers** (ASO, Streptozyme, or anti-DNase B)
What is the clinical course?	Varying degrees of renal insufficiency and hypertension, with resolution of clinical signs and symptoms after 1 month
How long can urinary abnormalities last?	Up to 2 years
How many children completely recover?	95%; children with severe involvement have all the complications of acute kidney injury.
Does penicillin help?	It may prevent further spreading of nephritogenic strains, but treatment of streptococcal infections does not decrease the risk of poststreptococcal glomerulonephritis.
List 7 chronic forms of glomerulonephritis that most commonly affect children.	IgA nephropathy, Henoch-Schönlein purpura (Ch 19, p. 277), focal segmental glomerulosclerosis, lupus nephritis, membranoproliferative glomerulonephritis, membranous nephropathy, Alport syndrome

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VESICOURETERAL REFLUX

What is it?	It is "backwash" of urine from the bladder into the ureter or kidney and is caused by incompetence of the ureterovesical junction.
How does it cause damage?	By exposing the kidney to high pressure during voiding and by increasing the risk of pyelonephritis in the presence of a lower UTI. Dilatation and scarring of the collecting system and renal parenchyma may lead to chronic kidney diseases (including ESRD) and hypertension.
What are the etiologic factors?	It may be primary and isolated (congenital incompetence) or **associated with other urinary tract abnormalities.** Secondary reflux may be caused by increased bladder pressure, inflammation, obstructing lesions (e.g., posterior urethral valves), or previous surgical procedures.
When is it usually recognized?	Usually during an evaluation for a UTI, renal insufficiency, hypertension, or voiding problems
How is it diagnosed?	**Voiding cystourethrogram (VCUG),** in which radiopaque dye is instilled into the bladder until full and the dye is observed during voiding
What is the grading system?	**Grade I:** reflux into a nondilated distal ureter **Grade II:** reflux into the upper collecting system without dilatation **Grade III:** reflux into a dilated collecting system without blunting of calyces **Grade IV:** reflux into a dilated system with blunting of calyces **Grade V:** massive reflux with gross dilatation and distortion of the ureter and collecting system

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What is the natural history?

Risk of renal scarring increases with the degree of reflux. Primary grades I and II reflux resolve spontaneously with maturation in 80% of children. Higher grades are less likely to resolve. Secondary reflux has a less favorable outcome across all grades.

What is the treatment?

Prevent infection with **antibiotic prophylaxis** (commonly, trimethoprim/sulfamethoxazole). If expectant management is undertaken, uroprophylaxis continues as long as the reflux persists.

Follow-up VCUGs should be performed every 1–2 years to evaluate the progression or regression of the reflux.

Children with severe degrees of reflux, breakthrough infections while on uroprophylaxis, or evidence of renal scarring are candidates for surgical correction via ureteral reimplantation or constriction of the ureteric orifice by endoscopic injection of synthetic material (e.g., dextranomer macrospheres or polydimethylsiloxane with polyvinyl pyrroline) around the orifice.

RENAL TUBULAR ACIDOSIS

What is it?

Systemic hyperchloremic (i.e., normal anion gap) acidosis resulting from abnormal urinary acid-base homeostasis

How does a patient usually present?

Growth failure in the first year of life

What is type 1 (distal) RTA?

The distal tubule has deficient H^+ excretion capability, which leads to excess body H^+.

What is type 2 (proximal) RTA?

An inability of the proximal tubule to reabsorb filtered bicarbonate leads to the loss of buffering capacity and acidosis.

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What is type 4 RTA?

Distal tubular damage, commonly caused by obstructive uropathy, leads to decreased responsiveness to aldosterone. The restricted ability to excrete H^+ and K^+ leads to hyperkalemic acidosis.

In what 3 ways can the physician differentiate among these types?

Types 1 and 2 usually cause hypokalemia, whereas type 4 tends to cause hyperkalemia.

Types 1 and 2 can be differentiated in an acidotic patient by urine pH: type 2 patients acidify urine to pH < 5.5 when serum HCO_3^- is <16 mEq/L (distal acidifying mechanisms still work).

Type 1 patients cannot acidify the urine because of distal abnormalities, even in the presence of significant systemic acidosis.

How is RTA treated?

Primarily by the replacement of bicarbonate:

Type 1 patients typically require only 1–2 mEq/kg per day of $NaHCO_3$ to maintain acid-base balance because of the small amount of base needed to buffer endogenously formed $\mathbf{H^+}$.

Type 2 patients are unable to reabsorb bicarbonate, and doses of **>10 mEq/kg** per day may be needed for acid-base balance.

Type 4 patients may occasionally require potassium-binding resin therapy for hyperkalemia.

Patients with distal RTA who develop hypercalcemia may benefit from thiazide diuretics.

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FANCONI SYNDROME

What is it?	Generalized aminoaciduria, glycosuria, and phosphaturia. It is often accompanied by bicarbonate wasting, proteinuria, and hyperkaluria, all of which are caused by proximal tubule transport defects.
What are the 2 common clinical manifestations?	Growth failure and vitamin D–resistant rickets
What causes it?	It is usually idiopathic. However, it is a common feature of certain inborn errors of metabolism (e.g., cystinosis, galactosemia, Lowe syndrome) or toxic events (e.g., heavy-metal poisoning).
List 6 laboratory findings.	Normal anion gap hyperchloremic metabolic acidosis, hypokalemia, hypophosphatemia, elevated fractional excretion of phosphate (>15%), glycosuria in the presence of euglycemia, aminoaciduria
List 3 treatments.	Diagnosis and treatment of underlying causes; high doses of vitamin D; phosphate and bicarbonate supplementation

DIABETES INSIPIDUS

What is central DI?	Loss of ADH secretion, resulting in the inability to concentrate urine appropriately despite normal renal function
List 3 consequences.	Increased urine output, hypernatremia, and dehydration
List 8 common etiologic factors.	Idiopathic, posttraumatic, postsurgical, congenital malformation, intracranial tumors, CNS infections, histiocytosis, granulomatous disease

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What are the 4 signs or symptoms?

Polyuria, polydipsia, weight loss, growth failure; patients generally prefer water to other fluids.

What signs may indicate that DI is secondary to a tumor?

Neurologic or visual complaints

List 3 diagnostic laboratory results.

Morning urine specific gravity < 1.010; low urine osmolarity; normal-to-high serum sodium concentration

What is the water-deprivation test?

The test is designed to assess urinary response to water deprivation. An initial water load of 500 mL/m^2 is given. Measurements are taken of:

1. Hourly body weight and urine output
2. Urine specific gravity and osmolarity of each sample
3. Serum sodium and osmolarity every 4 hours

Desmopressin (dDAVP), a long-acting analog of ADH, is given at the end of the test to document responsiveness to ADH.

List 4 indications of a positive test.

Persistence of dilute urine with osmolarity less than that of plasma; a rise in serum sodium to >145 mEq/L; a rise of serum osmolarity to >290 mOsm/kg; weight loss of 3–5%

What 2 radiographic tests should be ordered?

1. **Skull radiograph** investigating for calcification, enlargement of the sella turcica, erosion of the clinoid processes, or increased width of the suture lines
2. An **MRI** to detect lesions of the pituitary gland and hypothalamic-neurohypophyseal tract

What is the treatment?

Desmopressin (dDAVP) once or twice daily

What is the differential diagnosis?

Nephrogenic DI; psychogenic water drinking; impaired thirst mechanism

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NEPHROGENIC DI

What is the difference between nephrogenic and central DI?

Patients with nephrogenic DI synthesize and secrete adequate ADH, whereas patients with central DI do not.

What is the primary defect in nephrogenic DI?

Lack of distal tubular response to ADH, with inability to concentrate the urine

What is the etiologic factor in primary nephrogenic DI?

Primary nephrogenic DI is a rare X-linked recessive disorder with profound effects in males, although females may be mildly affected. In most families, the defect is caused by a mutation in the vasopressin receptor. Autosomal dominant and recessive DI have also been described in which aquaporin 2 (the renal water channel) is defective.

List 4 etiologic factors in secondary nephrogenic DI.

Secondary nephrogenic DI is more common and often less severe than primary DI. Causes include obstructive uropathy, chronic renal failure, sickle cell disease, and drug toxicity.

How does a patient present?

In the more severe forms, signs appear within the first weeks of life, usually including polyuria, polydipsia, failure to thrive, and chronic dehydration. The degree of dehydration is commonly underappreciated because the child continues to urinate. Fever, irritability, and poor feeding are also common.

What are the characteristic laboratory findings?

Hypernatremia, hyperchloremia, urine osmolarity < 200 mOsm/kg in the presence of serum osmolarity > 300 mOsm/kg. ADH levels are normal, and there is no response to exogenously administered vasopressin.

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List 3 treatments.

1. Maintenance of adequate fluid intake (most important)
2. Although somewhat counterintuitive, thiazide diuretics decrease urine output by causing mild sodium depletion, thereby encouraging proximal tubular sodium and water reabsorption.
3. Prostaglandin synthesis inhibitors may decrease urine output (mechanism of action is unclear).

RENAL STONES

List 3 signs or symptoms.

Hematuria (microscopic or gross); abdominal or flank pain; UTI

What is the most common chemical composition of a stone?

Calcium oxalate

List 4 other chemical compositions.

Calcium phosphate, struvite (magnesium ammonium phosphate), uric acid, cystine

List 10 predisposing conditions.

Urinary tract anomalies, recurrent UTIs, hypercalciuria, distal RTA, immobilization, hyperoxaluria, cystinuria, hyperparathyroidism, chronic loop diuretic use, hypocitraturia (citrate is an inhibitor of stone formation)

What are helpful imaging studies?

1. Plain abdominal radiograph may show calcium-containing stones.
2. IV pyelogram or ultrasound examination (U/S) documents the location of radiopaque and radiolucent stones, as well as the degree of obstruction.
3. CT scan (spiral technique required)
4. U/S and VCUG help evaluate urinary tract abnormalities.

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What are useful laboratory studies?

Serum: electrolytes (especially HCO_3^-), calcium, phosphorus, uric acid, creatinine, parathyroid hormone

Urine: urinalysis and culture, urine pH, 24-hour collection for calcium, creatinine, phosphorus, oxalate, uric acid, cystine, and citrate

What is normal calcium excretion?

4 mg/kg per day or spot urinary calcium to creatinine ratio < 0.2

List 4 treatments of hypercalciuria.

Limit calcium intake to the recommended daily allowance; increase fluid intake; limit sodium intake (sodium restriction increases calcium reabsorption). If stones persist, thiazide diuretics may help.

Describe the therapeutic management of renal stones.

Provide **hydration** and **pain management** until stone passes. **Treatment of predisposing conditions** may prevent future stones. If stones persist, **lithotripsy** can pulverize some stones without the need for surgery. Endoscopic, percutaneous, or open **surgery** may be needed.

HYPERTENSION

What defines hypertension in the pediatric population?

BP increases with age:

Significant hypertension is defined as BP greater than the 95th percentile for age, sex, and height.

Severe hypertension is BP greater than the 99th percentile.

What is the appropriate size for a child's BP cuff?

The cuff bladder width should be large enough to cover two-thirds of the upper arm length. Bladder length should be long enough to surround the entire arm circumference. Cuffs that are too small give erroneously high readings, and cuffs that are too large may give erroneously low readings.

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List 3 common causes of acute hypertension in infants.	Renal artery occlusion, medications, hypoxia
In children and adolescents? (list 3)	AGN, HUS, and medications (including illicit drugs)
List the 4 most common causes of chronic hypertension in infants.	Renal arterial thrombi (umbilical catheter complication; Ch 8, p. 66); aortic coarctation (Ch 16, p. 194); obstructive uropathy; medications
In young children? (list 4)	Obstructive uropathy; reflux nephropathy (see Vesicoureteral Reflux, p. 335); glomerular disease (see Glomerulonephritis, p. 334); renal artery stenosis
In adolescents? (list 4)	Essential hypertension; glomerular disease (see Glomerulonephritis, p. 334); reflux nephropathy (see Vesicoureteral Reflux, p. 335); renal artery stenosis
How is the diagnosis of essential hypertension made?	By exclusion of secondary causes
What should evaluation include?	Studies of renal function, kidney anatomy, and urinary sediment Studies for rare but correctable causes, such as pheochromocytoma (Ch 24, p. 391), Cushing disease (Ch 24, p. 390), and aortic coarctation (Ch 16, p. 194) Nuclear scans, arteriography, or both for diagnosis of renal artery stenosis may be needed.
What medications are used to treat chronic hypertension in children?	Essentially all forms of antihypertensives may be used if monitored appropriately.
When are diuretics used?	For patients with underlying renal disease and fluid overload, diuretics are often used in conjunction with other medications.

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Chapter 22 Genitourinary Disorders

HYPOSPADIAS

What is it?	Malformation of the penis with abnormal ventral placement of the urethral meatus along the penile shaft, scrotum, or perineum
What is the incidence?	About 1 per 300 male births; it is the most common penile malformation.
What are some associated malformations?	Chordee (curvature) of the penis, hernias, and cryptorchidism are common. Chromosome abnormalities are uncommon in patients with uncomplicated hypospadias, but more common with complex malformations.
What is the treatment?	Surgical repair, sometimes in stages for severe hypospadias
List 3 common complications.	Chordee, fistula, stricture

CRYPTORCHIDISM

What is it?	Lack of normal descent of the testicle; unilateral in 75% of cases, bilateral in 25%
What is the incidence?	About 1 in 100 male infants
At what age is a boy's testicle considered truly undescended?	After 1 year of age
Why?	Testicles undescended at birth usually descend into the scrotum within the first year.

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What are the 2 consequences of an undescended testicle?

1. After the second year, testicular degeneration occurs, resulting in low spermatogonia counts and degeneration of germinal epithelium. Seminiferous tubules become fibrous. This overall degeneration can cause the formation of sperm antibodies, which can adversely affect fertility even if there is a normal descended testicle on the opposite side.
2. There is an **increased incidence of cancer,** particularly **seminoma,** in an undescended testicle. Orchiopexy may reduce this incidence if performed before puberty and allows examination of the testicle.

What is the treatment?

Hormone therapy may be used to induce the descent of the testicle but is usually unsuccessful.

The major treatment is **orchiopexy.** If the testicle is palpable, an open inguinal approach is appropriate. If the testicle is not palpable (i.e., is inside the inguinal canal or intra-abdominal), the **Fowler-Stevens approach** may be indicated. The spermatic vessels are divided high in the retroperitoneum as a first procedure (laparoscopic approach preferred), with the testicle left in place. Collateral vessels then form along the vas deferens, and the testicle is brought into the scrotum as a second procedure, usually laparoscopically.

When should orchiopexy be performed?

Strong consideration should be given to orchiopexy between 6 months and 1 year of age if the testicle is showing no signs of descent. Orchiopexy should absolutely be performed no later than 18–24 months of age (allowing age correction for prematurity).

What should be done if the testicle is not palpable at all?

Laparoscopic examination to determine the presence or absence of the testicle

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EPIDIDYMITIS

What is it?	Inflammation of the epididymis
What are the 2 most common causes?	Reflux of infected urine; STDs caused by gonococci and *Chlamydia*
How does the patient present?	With unilateral pain in the scrotum
What are the physical findings?	A large, tender, and firm epididymis with a normal testicle
What is the treatment?	Antibiotics appropriate for the identified pathogen
What should be suspected if epididymitis occurs in a non-sexually active child or a prepubertal child?	A urinary tract abnormality. They should be evaluated with a renal ultrasound and a voiding cystourethrogram (VCUG).

TESTICULAR TORSION

What is it?	The testicle twists on its blood supply and the vas deferens. The testicle may become ischemic and necrotic.
What are the 2 types of testicular torsion?	1. **Extravaginal:** torsion is outside of the tunic vaginalis (predominantly in neonates) 2. **Intravaginal:** torsion is within the tunic vaginalis (a.k.a. "Bell-clapper" anomaly—predominantly in older boys)
How do patients with torsion present?	Usually with significant scrotal pain and swelling unilaterally. The testicle may also be high in the scrotum because of the twisted cord.
What is the differential diagnosis?	Epididymitis, orchitis, acute hydrocele, torsion of the appendix epididymis or appendix testes, incarcerated inguinal hernia

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What is Prehn sign?

A potential way of differentiating testicular torsion from epididymitis. Lifting of the testicles may relieve pain associated with epididymitis, but not with torsion.

What is the evaluation for testicular torsion?

If testicular torsion is highly suspected, the patient should be brought directly to the operating room. If the duration of symptoms is approaching or exceeding 6 hours, it is likely that the testicle is necrotic. If the time from onset of the symptoms is shorter and there is a suspicion that an alternative diagnosis is possible, **Doppler ultrasound** can be used to evaluate for torsion, or for an alternative cause of the presenting symptoms and for assessment of viability of the testicle. Radioisotope imaging (not commonly used anymore) can assess for viability of the testicle but does not image the torsion.

What is the treatment?

Surgical exploration of the scrotum is performed through a midline incision in the scrotum. If viable, the affected testicle is unrotated and is fixed at 4 points in the scrotum. If the testicle is necrotic, it is removed. In either case, the opposite testicle is fixed in the opposite side of the scrotum at 4 points as well.

TESTICULAR TUMORS

What are the 4 common types of testicular tumors that affect boys (with examples when appropriate)?

1. **Germ cell tumors,** including yolk sac tumor (endodermal sinus tumor), teratoma or teratocarcinoma, seminoma (rare in children)
2. **Gonadal stromal tumor (non–germ cell tumor),** including Leydig cell tumor, Sertoli cell tumor (often benign), gonadoblastoma, mixed tumors
3. **Leukemic and lymphomatous infiltrates**
4. **Rhabdomyosarcoma** (is paratesticular)

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Which of these types is the most common?

Germ cell tumor

Which is the most common testicular cell tumor in prepubertal males?

The **yolk sac** (endodermal sinus) tumor

What is the typical sign of a testicular tumor?

Presence of a painless testicular mass. Sometimes, a hydrocele may be caused by the tumor and may delay diagnosis.

What are the common chemical markers of germ cell tumors?

The most common is **α-fetoprotein (AFP)**. **β-Human chorionic gonadotropin (β-HCG)** is rarely elevated in prepubertal testicular tumors.

What is the surgical workup and treatment?

Biopsy should be performed through an inguinal incision if possible. Traditionally, positive biopsies obtained through a scrotal incision have required scrotal resection. However, greater attention is now paid to scrotal preservation in these instances. Orchiectomy is performed through an inguinal incision. If the tumor is isolated to the testicle, surgical resection (orchiectomy) is all that is required. Retroperitoneal lymph node dissection is required if there is evidence of clinically suspicious nodes on CT scan. An extensive tumor will require chemotherapy.

In which patients do gonadoblastomas arise?

Patients with mixed gonadal dysgenesis (45,X/46,XY) or intersex with dysgenetic gonads in association with a Y chromosome in the karyotype. These patients may be genotypic males but are usually not phenotypic males.

What is the treatment?

Usually only removal of the gonads, because these tumors are encapsulated and slow-growing. If the gonads are palpable, consideration is given to delayed removal since hormonal effects in infancy may be important in gender imprint on the brain.

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In which patients do seminomas most commonly arise?	Males with cryptorchid testes
What are the 2 components of the treatment of seminoma?	Surgical resection with radiation therapy. These tumors are particularly sensitive to radiation therapy.
How are leukemic and lymphomatous infiltrates treated?	After a transscrotal biopsy, these lesions are treated systemically according to the type of disease found.
Which tumors cause precocious puberty?	**Gonadal stromal tumors,** such as the Sertoli cell and Leydig cell neoplasms. Leydig cells, in particular, secrete excessive testosterone (Ch 24, p. 375, precocious puberty). Gynecomastia may also be seen with Sertoli cell tumors.
What is the treatment?	Surgical resection (orchiectomy)
How is testicular rhabdomyosarcoma evaluated and treated?	Similar to yolk sac tumor. Bone scan and bone marrow aspirates are needed as well. Radiation therapy may be adjunctive but is rarely needed.
What are the outcomes for rhabdomyosarcoma?	Favorable, about 95% 5-year survival rate
What is the treatment for teratoma?	Simple orchiectomy

OVARIAN TUMORS

What are the 4 major categories of ovarian tumors in children?	1. **Germ cell tumors,** including teratoma, germinoma, endodermal sinus tumor, embryonal carcinoma, and choriocarcinoma 2. **Germ cell sex cord-stromal tumors** including gonadoblastoma and other mixed germ cell-sex cord tumors. 3. **Sex cord stromal tumors,** including granulosa-theca tumor and Sertoli-Leydig tumor 4. **Epithelial ovarian tumors,** including mucinous, serous, clear cell, endometrioid, mixed, and undifferentiated

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Which is the most common ovarian tumor in children?	Teratoma
Are most teratomas benign or malignant?	Benign
How does a child with teratoma present?	With an abdominal mass, pain, or both
What may be a pertinent radiographic finding?	The presence of calcification—in 50% of cases
What is the treatment?	If the tumor is within the capsule of the ovary, unilateral salpingo-oophorectomy is all that is needed. However, if the tumor is grade II or greater, there is an increased chance of malignancy, and chemotherapy will be required.
What are germinomas?	Malignant tumors derived from totipotential (i.e., sexually undifferentiated) germ cells. Historically, these have been termed dysgerminoma if found in the ovary, seminomas if found in the testis, and dysgerminoma if in an extragonadal site (e.g., anterior mediastinum and pineal regions).
In what age groups are they most common?	Prepubertal and adolescent girls
Are these tumors biologically active?	Minimally
What is an endodermal sinus tumor?	An aggressive malignant germ cell tumor that originates from undifferentiated and multipotential embryonal cells. It grows rapidly and metastasizes early.
In what age groups do they most commonly present?	Teenage girls and young adult women In young children and infants, this tumor typically presents in the sacrococcygeal area.
Is there a tumor marker?	Yes, AFP

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What is the outcome?

4-year survival rate of approximately 50–70%

What is an embryonal carcinoma?

A rare malignant germ cell tumor that is quite biologically active. It resembles embryonal carcinoma of the adult testis.

What are the tumor markers?

AFP and β-HCG

List 3 characteristics of the typical presentation.

Abnormal vaginal bleeding, hirsutism, and precocious puberty caused by elevated β-HCG

What is a choriocarcinoma?

An aggressive germ cell tumor differentiated toward trophoblastic structures. It generally is hormonally active.

What is the tumor marker?

β-HCG

With what symptoms does the patient present?

Precocious puberty or menstrual irregularity

What are the general treatment guidelines for germ cell tumors?

1. **Surgical:** Staging includes evaluating for peritoneal implants, omental implants, liver metastases, pelvic and para-aortic lymph nodes or any other enlarged nodes, sampling of abdominal fluid for cytology, and examination of the opposite ovary. This evaluation may be done via laparoscopy or open procedure depending on suspicions and findings. If the examination reveals no suspicious findings, unilateral oophorectomy is done. Otherwise, appropriate samples are taken with possible debulking of lymph nodes and removal of the omentum.
2. **Medical:** Tumors that are malignant and beyond stage 1 (confined to ovary) will require adjuvant chemotherapy. Consideration may be given to bilateral oophorectomy. Radiation therapy has largely been abandoned.

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What is a gonadoblastoma?

A tumor that arises in dysgenetic gonads. Most patients have a 46 XY or 45 X/46 XY karyotype.

What is the treatment?

Surgical resection of the gonadoblastoma with consideration given to the removal of the opposite gonad, as it may also be at risk for malignant degeneration. Other treatment principles are as outlined for germ cell tumors.

What is a granulosa-theca cell tumor?

A tumor that has its origin in sex cord or stromal tissue

With what symptoms does the patient present?

An abdominal mass and precocious puberty

What are the pertinent laboratory findings?

Elevated levels of serum and urinary estrogen, inhibin, Müllerian-inhibiting substance

What is the treatment?

Surgical resection for stage I. Advanced-stage disease may require bilateral salpingo-oophorectomy, hysterectomy, chemotherapy, and radiation therapy.

What is a Sertoli-Leydig cell tumor?

Another ovarian tumor of sex cord or stromal origin. These tumors were formerly called "arrhenoblastomas."

What is the characteristic presentation?

An abdominal or pelvic mass with masculinization (because these tumors typically secrete testosterone)

What are the 3 tumor markers?

Elevated CA-125, AFP, lactate dehydrogenase (LDH)

What is the treatment?

Surgical resection for stage I disease. Advanced disease may require strategies similar to those used for granulose-theca cell tumors.

What are epithelial ovarian tumors?

Tumors of typical ovarian tissue; infrequent in children

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What is the primary tumor marker?

Elevated CA-125

What is the most typical presentation in children?

Presence of an abdominal mass

How is surgical staging undertaken?

1. Peritoneal washings for cytology
2. Examination of all peritoneal surfaces and liver
3. Biopsies of the diaphragm and peritoneum
4. Omentectomy
5. Sampling of para-aortic and pelvic lymph nodes

What is the treatment?

Total abdominal hysterectomy, bilateral salpingo-oophorectomy, and omentectomy with the staging procedures as outlined in the preceding text. Adjuvant chemotherapy is needed in all stages above stage I as well as in some stage I cases.

What are the 3 indicators of poor prognosis?

Advanced stage; aneuploidy; C-fms oncogene

VAGINAL ANOMALIES

What is vaginal atresia (a.k.a. "agenesis")?

A condition (sometimes known as "Mayer-Rokitansky syndrome") in which the Müllerian ducts fail to reach the urogenital sinus, which results in the failure of vaginal canalization. Atresia may be proximal (i.e., virtually no vaginal formation) or distal (proximal canalization with distal obstruction).

In vaginal atresia, what is the status of the other reproductive organs?

The ovaries and fallopian tubes are normal. The uterus is usually normal.

What is proximal vaginal atresia attributable to?

Dysplasia of the Müllerian ducts

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What is the status of the reproductive organs?	The fallopian tubes and ovaries are normal. The uterus and cervix are hypoplastic or absent.
What is hydrocolpos?	Filling of the vagina with mucus
What is hydrometrocolpos?	Filling of the vagina and uterus with mucus
What is hematocolpos?	Filling of the vagina with menstrual blood discharge
What is hematometrocolpos?	Filling of the vagina and uterus with menstrual blood discharge
With what symptom does a patient with distal vaginal atresia commonly present?	Colicky abdominal pain once menarche begins because of collection of menstrual blood (i.e., hematocolpos or hematometrocolpos)
How is distal vaginal atresia treated?	Perineal vaginoplasty
How does proximal vaginal atresia present?	It is more likely to present as the lack of menstrual periods because of the hypoplasia or agenesis of the uterus.
Can abdominal pain occur with proximal vaginal atresia?	Yes, in cases in which the uterus is well formed enough to produce the menstrual blood
How is this treated?	Drainage of the uterus and formation of a vagina using any vaginal tissue that may be present, using a pull-through of a portion of bowel, or a combination of these measures

VAGINAL TUMORS

What are the 2 most typical vaginal tumors of childhood?	Rhabdomyosarcoma (sarcoma botryoides), endodermal sinus tumor (germ cell)
How do patients with these tumors present?	Vaginal mass or swelling with possible vaginal bleeding

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List 3 components that the workup for an endodermal sinus tumor should include.

Serum tests for AFP and β-HCG; CT scan of the vagina, pelvis, and abdomen; chest radiograph

What is the treatment?

Usually, biopsy of the tumor is performed with subsequent chemotherapy and completion resection. Resection may include the removal of the uterus if the tumor extends that far.

List 5 components included in the workup of rhabdomyosarcoma.

Chest radiograph, CT scan of the pelvis and abdomen, bone marrow aspiration, bone scan, cystoscopy

How is this tumor treated?

Usually, biopsy is followed by chemotherapy to reduce the tumor and then completion resection. Hysterectomy or pelvic exenteration is rarely required.

What is the 5-year survival for rhabdomyosarcoma of the vagina?

Approximately 90%

IMPERFORATE HYMEN

What is imperforate hymen?

Persistence of an epithelial membrane at the opening of the vagina. It is the most common cause of vaginal obstruction.

How does a patient with imperforate hymen usually present?

It is often not apparent until adolescence. The girl experiences episodes of lower abdominal pain but no menstruation. After a time, a lower abdominal mass may be present, representing hydrometrocolpos, hematometrocolpos, or hematocolpos.

How is this treated?

The hymen is incised with a cruciate incision. The raw edges of the hymen ring are then sutured with absorbable sutures to promote epithelialization of the edges of the new open hymen ring.

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Chapter 23 Hernias and Abdominal Wall Defects

HERNIAS

What is a hernia?	Protrusion of a body component or organ outside its normal respective compartment. A hernia can involve the inguinal canal, abdominal wall, diaphragm, fascia, and mesentery among other areas.
What is the most common type of hernia?	Inguinal hernia (Fig. 23-1)

INGUINAL HERNIAS

What is an inguinal hernia?	A protrusion through the inguinal canal in the groin region. There may also be weakness of the muscle comprising the inguinal canal and floor.
What is an indirect inguinal hernia?	A herniation of the peritoneal sac through the inguinal ring itself. An indirect hernia does not imply any weakness of the muscle in the inguinal region. Therefore, a "defect" is not felt in the abdominal wall. However, the inguinal ring can be stretched by large hernia sacs (particularly in premature infants).
What is the most common sign of an indirect inguinal hernia?	A bulging in the groin or scrotal region. This may be unilateral or bilateral.
Which side is more commonly involved?	The right side

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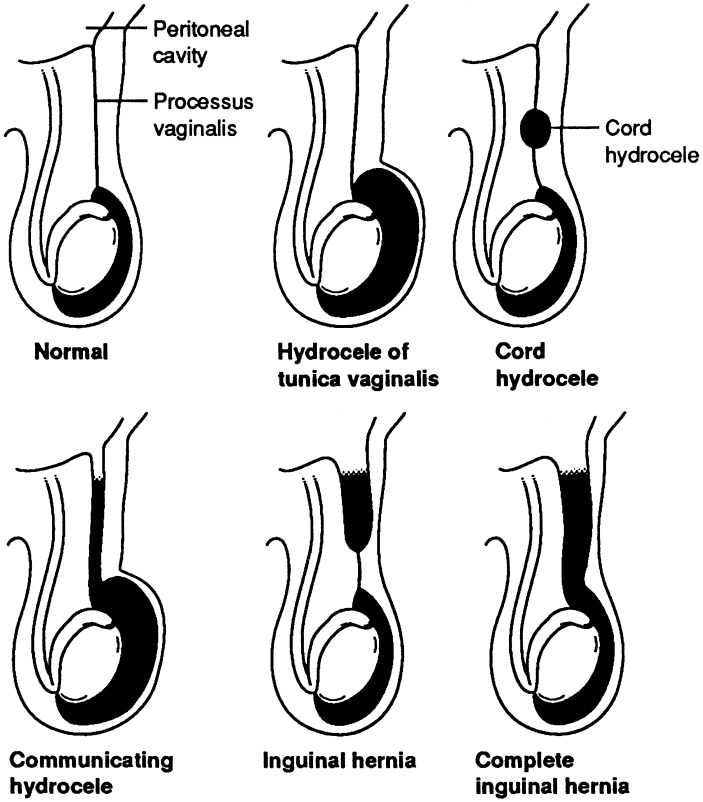

Figure 23–1. From left, configurations of hydrocele and hernia in relationship to patency of the processus vaginalis.

What are the common symptoms?

Most indirect inguinal hernias manifest as a **painless bulge.** If a hernia is symptomatic, **pain** is the most common symptom. If intestines are incarcerated within a hernia sac, **nausea** and **vomiting** may be additional symptoms.

What is the treatment of an indirect inguinal hernia?

Surgical ligation of the hernia sac at the level of the internal inguinal ring. The ring may need to be reinforced if it has been stretched by a large sac (not common beyond premature infants)

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List 3 reasons an inguinal hernia needs to be repaired.

1. It does not resolve on its own.
2. It can become larger with time.
3. Most important, intestines can be entrapped (incarcerated) within the hernia, thus causing a **surgical emergency.**

What is a hydrocele?

A collection of fluid along the testicular cord or within the scrotal sac. Most hydroceles are isolated within the tunica vaginalis and resolve before 1 year of age.

What is a cord hydrocele?

A hydrocele that is isolated within a remnant of the processus vaginalis along the spermatic cord structures. These **tend not to resolve** and usually need to be removed.

What is a communicating hydrocele?

A hydrocele in which the fluid is moving into and out of a patent processus vaginalis

What is a direct inguinal hernia?

In children, this manifests as a weakness in the muscle wall medial to the inferior epigastric vessels.

What are the signs and symptoms of a direct inguinal hernia?

Similar to indirect inguinal hernias; can be difficult to distinguish from indirect inguinal hernias on physical examination

What are the risk factors for direct inguinal hernias?

Conditions that result in chronic abdominal pressure, such as chronic coughing, constipation, heavy lifting, or ascites. Connective tissue disorders may also predispose to these hernias.

Are direct inguinal hernias common in infants and children?

No

How are direct inguinal hernias treated?

The hernias are repaired surgically. If a sac is present, it is ligated at the level of the inguinal ring. In addition, the musculature of the inguinal ring and floor can be repaired through a variety of techniques. Mesh is rarely used in children.

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What is an epigastric hernia?	A weakness in the midline fascia that exists between the area of the umbilicus and the xiphoid process. It is generally between 0.5 and 1 cm in diameter.
How does a patient with an epigastric hernia present?	A small bulge will be noted in the involved area. There may be associated pain because of entrapment of preperitoneal fat (this condition is sometimes termed **"epiplocele"**). It is rare that intestines will be involved in such a small hernia.
How are these hernias treated?	By surgical closure of the fascia
What is a Richter hernia?	A hernia involving an intestinal loop such that only a portion of the intestinal lumen is entrapped by the hernia
What is a sliding hernia?	A hernia in which a portion of the hernia sac is composed of an adjacent organ such as a loop of bowel or a portion of bladder
What is a Littre hernia?	A hernia in which a Meckel diverticulum is protruding into the hernia sac (Ch 19, p. 308)

UMBILICAL HERNIA

What is it?	Congenital fascial defect that persists in the umbilical region. It represents an incomplete closure of the omphalos that is present during fetal development.
What is the incidence?	1 in 6 children
In what 2 groups of children is it most common?	African American children (occurs 9 times more often than in other populations) and premature infants

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What is the natural history of an umbilical hernia?

They usually close spontaneously by 3–5 years of age and therefore can be monitored in the infant and toddler stage. An umbilical hernia is unlikely to cause symptoms in this age group. However, fascial defects > 1.5 cm are not likely to close.

What is the treatment?

Surgical fascial closure

List 5 indications for surgery.

1. Lack of closure by 5 years of age
2. Fascial defect of 1.5 cm or greater
3. Large umbilical proboscis resulting in skin excoriation or other difficulty
4. Discomfort from the hernia
5. Incarceration (rare)

CONGENITAL DIAPHRAGMATIC HERNIAS

(See Ch 17, p. 215.)

ABDOMINAL WALL DEFECTS

OMPHALOCELE

What is an omphalocele?

A centrally located abdominal wall defect that results from the failure of closure of the abdominal wall during development. Omphaloceles can be small or large. The larger the omphalocele, the less well formed is the abdominal wall and abdominal cavity.

Omphalocele defects are covered by an intact sac (Wharton jelly) in most cases. The umbilicus extends from the apex of the Wharton jelly.

What is the incidence?

About 1 in 4,000 live births

List 5 associated conditions.

Congenital heart defects, lung hypoplasia, chromosomal abnormalities, renal abnormalities, malformation syndromes (e.g., Beckwith-Wiedemann syndrome)

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List 2 ways omphaloceles are diagnosed.

Prenatal ultrasound (this allows time for prenatal counseling of parents and investigation for other potential anomalies during the prenatal stage); clinical examination at birth

List 4 components of the initial management of an infant with an omphalocele.

Management is mainly supportive.

1. IV access is established and IV fluids are administered.
2. Antibiotics are administered by IV.
3. An NGT is placed for gastric decompression.
4. The omphalocele is dressed with a moist gauze. A plastic wrap can be placed to cover the gauze, thereby retaining heat and moisture.

Cardiac and renal ultrasounds should be obtained. Chromosomal analysis should also be done.

How is an omphalocele repaired?

If the omphalocele is small?

If no contraindications, by excising the Wharton jelly sac and closing the fascia soon after birth, or when the infant is otherwise stable

If the omphalocele is large?

1. The Wharton jelly can be removed and a temporary silo of prosthetic material such as Silastic can be placed. The silo is then reduced during a 10- to 14-day period, and the abdominal wall is closed, primarily with fascia or with a small prosthetic patch of a material such as Gore-Tex. **Caution:** The abdominal pressure required to close the abdomen using this method may result in respiratory, renal, and hemodynamic compromise.

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2. A large omphalocele can be left intact and be treated with any of a variety of topical agents such as silver sulfadiazine or antibiotic ointment ("paint and wait method"). The Wharton jelly then epithelializes. The abdominal wall defect can then be closed months or even years later when a closure in 1 or more stages can be assured and the patient's medical status is stable. This is now the preferred approach for large omphaloceles.

Is the presence of an omphalocele a surgical emergency?

No, if the **Wharton jelly of the omphalocele is intact.** The Wharton jelly protects the abdominal contents. All other necessary evaluation and management issues should be addressed first. If the **Wharton jelly has been disrupted** in utero or during birth, **surgical treatment is emergently needed.**

What is the outcome of treatment of omphalocele?

Generally, the abdominal wall defect can be successfully managed. Outcome depends more on associated conditions.

GASTROSCHISIS

What is a gastroschisis?

Gastroschisis is an opening in the abdominal wall that is present to the right of the umbilicus. The intestine is exposed. The opening is usually small, and the abdominal wall and cavity are usually quite well formed.

What are the etiologic factors?

Unknown. A common theory is that the defect occurs in the position where a second umbilical vein degenerates during fetal development.

What is the incidence?

About 1 in 6,000–10,000 live births

List 2 ways gastroschisis is diagnosed.

Gastroschisis is often diagnosed on **prenatal ultrasound** or is immediately apparent at birth.

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List 2 associated conditions.

Atresia of the intestine or in utero intestinal perforation

List 5 components of the initial management of gastroschisis.

1. IV fluid administration
2. Administration of IV antibiotics
3. Nasogastric suction
4. Placement of a plastic bag around the infant's body up to chest level. This maintains a moist atmosphere for the exposed bowel.
5. The infant should be lying on the right side so that the mesentery of the bowel is not compromised by hanging over the umbilical region of the abdominal wall.

How is gastroschisis repaired?

Reduction of the exposed intestines should be achieved after the **supportive measures** noted earlier are instituted. This is generally done in the operating room with the infant under general anesthesia and the abdominal wall relaxed. In some cases, this may also be accomplished in the neonatal intensive care unit. If the intestines can be reduced without significantly compromising the infant's respiratory status and without compromise of renal perfusion, the abdominal wall can then be closed primarily.

If reduction cannot be achieved because of bowel swelling, then a **prosthetic silo** (e.g., Silastic) can be placed. Reduction of the intestines can be accomplished gradually during a 7- to 14-day period as bowel swelling resolves. The abdominal wall can then be closed surgically.

What is the outcome for infants with gastroschisis?

Survival is about 98%. Any adverse sequelae are caused by the presence of intestinal atresia, compromise, or perforation and the surgical treatment required for those conditions.

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Chapter 24 Endocrine Disorders

DIABETES

DIABETES INSIPIDUS (SEE CH 21)

DIABETES MELLITUS

What is it?	Absent or diminished insulin secretion or action resulting in hyperglycemia and abnormal energy metabolism
What are the 2 types?	**Type 1:** loss of pancreatic β-cell (insulin-secreting cell of the islets of Langerhans) function, resulting in a loss of insulin secretion; **it is the most common type seen in childhood.** **Type 2:** insulin resistance with insufficient insulin secretion; more common in adults but becoming more common in children
What are the etiologic factors?	Not completely known; associated with a combination of genetic and environmental factors. Hyperglycemia can further impair β-cell function, called "glucose toxicity." **Type 1:** Autoimmune processes are important in type 1, which might be triggered by environmental influences (viral?) in an individual with predisposing genetic factors. Risk of type 1 DM increases if other family members are affected with type 1 DM. Certain HLA haplotypes confer increased risk of type 1 DM: DR3DQ2 and DR4DQ8.

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Type 2: Inherited β-cell defects and propensity for insulin resistance with increasing body mass index are associated. Patients have impaired first-phase insulin release. Genetics are polygenic with varying interactions with the environment. Type 2 DM is more "hereditary" than type 1 DM. Lifetime risk is ~40% if parent has type 2 DM and ~90% if monozygotic twin is affected. Prevalence is higher in African Americans, Native Americans, and Hispanic Americans.

How does a patient with DM present?

Most commonly with **polyuria, polydipsia, and weight loss;** symptoms often occur insidiously over weeks to months. Pediatric patients sometimes present with diabetic ketoacidosis (DKA), including those with type 2 DM.

How is DM diagnosed?

Two random blood glucose values > 200 mg/dL (or 1 if symptomatic); fasting blood glucose > 126 mg/dL; elevated glycosylated Hgb (HbA_{1c}) level ≥ 6.5%.

In type 1 DM, islet cell, insulin, or other autoantibodies are usually present.

In type 2 DM, a fasting insulin level may reveal hyperinsulinemia, indicating significant insulin resistance. However, if children have developed concomitant β-cell insufficiency or failure (indicated by significant hyperglycemia), then insulin levels may not be elevated, although may still be higher than patients with type 1 DM (i.e., c peptide > 0.6 ng/mL).

Why is glycosylated Hgb (HbA_{1c}) level important?

It provides an estimate of the average blood glucose level for the preceding 2–3 months.

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What are the goals of type 1 DM management?

Normal growth and development; prevention of early and late complications; manage glucose variability to prevent hypoglycemia

Specific goals for blood glucose ranges and HbA_{1c} vary by age:

Age	Glucose Range	HbA_{1c}
0–5 years	100–180 mg/dL	<8.5%
6–12 years	90–150 mg/dL	<8%
12–19 years	90–130 mg/dL	<7.5%

What are the goals of type 2 DM management?

Near-normal glycemic control (HbA_{1c} < 7%, fasting BG < 130 mg/dL); reduce factors leading to insulin resistance (manage weight, improve activity); identify and treat comorbidities (hypertension, dyslipidemia, nonalcoholic steatohepatitis); prevent vascular complications

List the 4 main components of management of type 1 DM.

1. **Insulin:** used to provide basal insulin needs (fasting requirements for glucose produced by liver from gluconeogenesis) and to metabolize carbohydrates consumed. Most insulin today is recombinant human insulin or insulin analogs, given as a combination of short-acting (regular, lispro, or aspart) and long-acting (NPH, glargine, or detemir). Methods of administering include as a 2 or 3 injections per day **"fixed" regimen** with NPH and regular/lispro/aspart or as **multiple daily injections** (basal-bolus method) with glargine/detemir supplying basal needs and lispro/aspart dosed on the basis of carbohydrate content prior to consumption. "Fixed regimen" using NPH has drawbacks of requiring "fixed" mealtimes and carbohydrate content each day and fasting hypoglycemia between meals/night/during illnesses. **Continuous insulin**

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infusion via insulin pump can provide tight glucose control in motivated patients by administering short-acting insulin (lispro/aspart) in adjustable basal amounts throughout the day with boluses given for carbohydrates or hyperglycemia. DKA can occur more quickly in pump patients if a problem with the infusion site is not promptly recognized, since no long-acting insulins are "on-board."

2. **Diet:** depends on insulin regimen: consistent carbohydrate intake is required for 2–3 injections per day **fixed insulin regimen**; carbohydrate counting is required for dosing of rapid-acting insulin in **multiple daily injections** and **insulin pump therapy**.
3. **Exercise:** aerobic exercise lowers the blood sugar without additional insulin and aids overall fitness. Patients should monitor blood glucose carefully during exercise. Easily absorbable carbohydrates (i.e., sports drinks) may be consumed during exercise to prevent hypoglycemia.
4. **Glucose monitoring:** at least 4 times daily before meals and bedtime. With insulin dose titration, patients may need to test 2 hours after meals and during the night to assess the response to insulin. Subcutaneous continuous glucose monitors are now available to give minute-to-minute glucose trends to assist with strategies to reduce glucose variability.

List 2 acute complications of type 1 DM and their causes.

1. **Hypoglycemia** can occur with over-insulinization or vigorous exercise, or if the patient skips meals when taking insulin via **fixed-dose regimen**.
2. **DKA** may be caused by poor patient compliance with insulin therapy, or by a severe illness.

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List 10 signs and symptoms of hypoglycemia.

Hunger, diaphoresis, and tremulousness because of sympathetic discharge

Deprivation of glucose to the CNS can lead to headaches, confusion, lethargy, bizarre behavior, slurred speech, loss of consciousness, and seizures.

What is the treatment of hypoglycemia?

If the patient is **alert,** give rapid-acting glucose-containing fluids and foods, or glucose tabs or gel. Repeat glucose check in 10–15 minutes until glucose > 80 mg/dL. Avoid complex carbohydrates and fat until glucose > 80 mg/dL, as they will delay carbohydrate absorption.

If the patient is **unconscious,** give intramuscular glucagon or intravenous glucose in water.

List 5 late complications of type 1 DM.

Retinopathy, nephropathy, neuropathy, large-vessel atherosclerosis, ulcers on lower legs and feet

Can these be prevented?

Tight glucose control can decrease the frequency of these complications by up to 75% and potentially prevent them.

What is DKA?

A **potentially life-threatening condition** occurring in DM that is characterized by **severe hyperglycemia,** with resulting **electrolyte disturbances, dehydration,** and **metabolic acidosis**

What are some signs or symptoms of DKA?

Polyuria, polydipsia, fatigue, dehydration with tachycardia (possible hypotension and hypoperfusion), abdominal pain, nausea, and vomiting; Kussmaul respirations (hyperpnea), obtundation, and coma may occur.

What are the causes of DKA?

A lack of adequate insulin is the primary cause of DKA. DKA may be the initial sign of DM. In patients with known diabetes, DKA may be triggered by illnesses or noncompliance with insulin therapy.

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List 7 metabolic derangements in DKA.

1. Severe hyperglycemia
2. Decreased serum CO_2 (with respiratory compensation)
3. Increased BUN and hematocrit (dehydration with hemoconcentration)
4. Normal or low Na^+ (pseudohyponatremia is artifact, caused by lipemic serum or hyperglycemia)
5. Normal or increased **serum K^+** caused by cellular shifts from acidosis; however, **total body K^+ depletion is present from renal losses and is potentially life-threatening. Be aware that K^+ may drop precipitously with correction of acidosis.**
6. Presence of ketones in serum and urine
7. Low serum phosphorus due to hyperphosphaturia

List 4 components of DKA treatment.

1. **Careful rehydration! IV fluid is the first step.** Patients typically have at least a 7–10% fluid deficit. Gradual rehydration with isotonic fluids may reduce the risk of cerebral edema. Initial rehydration may include 10–20 mL/kg total as boluses of normal saline or lactated ringers. Replete remaining fluid deficit over next 48–72 hours using maintenance to no more than twice maintenance rates.
2. **IV insulin:** infusion at 0.1 units/kg/hr. IV insulin bolus is not generally recommended. Younger children (<2 years) may need 0.05 units/kg/hr. Infusion should be titrated at a rate of no less than 0.05 units/kg/hr (for older children). IV dextrose can be started or increased to prevent hypoglycemia with insulin administration.

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3. **Dextrose** (5–10%) is added to IV fluids when the glucose level reaches 250–300 mg/dL. Dextrose plus insulin is needed to reverse the acidosis from the underlying catabolic process.
4. **$NaHCO_3$** is considered *only* for severe acidosis (serum $CO_2 < 5$ mmol/L or pH < 7.0) as it may increase complications such as cerebral edema.

What are the cautions in DKA treatment?

SLOW correction of hyperglycemia and dehydration is essential. Too much fluid too quickly or rapid shifts in osmolarity may cause cerebral edema and herniation. IV dextrose should be administered with IV insulin to avoid hypoglycemia during the latter phase of DKA treatment.

What is the treatment of type 2 DM?

Children with symptomatic type 2 DM require medication; those with HbA_{1c} values $> 8.5\%$ require insulin therapy. Diet and exercise are first-line treatments for those not yet symptomatic. The only pharmaceuticals approved by the FDA for use in pediatric type 2 DM are metformin and insulin.

DISORDERS OF CALCIUM BALANCE

HYPERPARATHYROIDISM

What is it?

Elevated levels of PTH, causing hypercalcemia and hypophosphatemia

What does PTH do?

PTH mobilizes calcium from bone, increases calcium reabsorption from the gut, and causes decreased renal tubular reabsorption of phosphorus. PTH causes the conversion of inactive 25-hydroxyvitamin D (storage form) to active 1,25-dihydroxyvitamin D by 1α-hydroxylation in the kidney with the assistance of magnesium.

What stimulates PTH secretion?

Hypocalcemia

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What are the signs and symptoms of hyperparathyroidism?

Symptoms related to hypercalcemia: fatigue, nausea, vomiting, anorexia, palpitation, pruritus, constipation, lethargy, weakness, depression, poor memory, confusion, polyuria, polydipsia, bone pain, and fractures

Symptoms secondary to kidney stones: hypertension, renal colic

Mnemonic: "stones, groans, bad bones, and psychiatric overtones"

List 3 diagnostic laboratory findings.

Elevated serum calcium (total and ionized); low phosphorus; inappropriately normal or elevated PTH (relative to elevated serum calcium)

What might the radiographs show?

Subperiosteal bone resorption

List 2 etiologic factors.

Hyperfunction of all the parathyroid glands **(hyperplasia)**—familial in 20%; a solitary **adenoma**—typically nonfamilial

What are the treatments?

Hyperplasia: subtotal parathyroidectomy (usually 3.5 glands) or total parathyroidectomy with reimplantation of pieces of half gland in sternocleidomastoid or brachialis muscle

Adenoma: removal of adenomatous gland

List 2 types of associated syndromes.

Multiple endocrine neoplasia (MEN):

Type I: tumors of the parathyroids, anterior pituitary, and pancreas or gut; autosomal dominant; inactivating mutation of tumor suppressor MENIN

Type IIa: hyperparathyroidism, pheochromocytoma, and medullary thyroid carcinoma; autosomal dominant; activating mutation of proto-oncogene RET

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What is familial hypocalciuric hypercalcemia?	Parathyroid gland insensitivity to the inhibitory effect of calcium, causing a higher Ca level "set-point" for PTH release
List 3 causes of secondary hyperparathyroidism.	Chronic renal disease, hepatic disease, and vitamin D deficiency
List 6 other causes of hypercalcemia.	Malignancy (leukemia, lymphoma, rhabdomyosarcoma, and fibrosarcoma), immobilization, granulomatous disease (extrarenal overproduction of 1,25-dihydroxyvitamin D due to sarcoidosis, tuberculosis, lymphoma, disseminated candidiasis, leprosy, Wegener granulomatosis, cat scratch disease, or *Pneumocystis carinii*), hyperthyroidism, adrenal insufficiency, drugs (lithium, vitamin A or D intoxication)

HYPOPARATHYROIDISM

What is it?	Low levels of PTH
List 2 metabolic results.	Hypocalcemia and hyperphosphatemia
What are the typical signs and symptoms?	Symptoms related to hypocalcemia: tetany, seizures, abdominal pain, numbness of the face or extremities, or carpopedal spasm. Chvostek and Trousseau signs may be elicited.
What is Chvostek sign?	Twitching of the muscles innervated by the facial nerve; it is elicited by tapping 1–2 cm anterior to the earlobe just below the zygomatic process.
What is Trousseau sign?	Carpal spasm that occurs after inflating a blood pressure cuff on the upper arm to above systolic pressure for up to 3 minutes
What is the treatment for hypoparathyroidism?	Active vitamin D (or 1,25-dihydroxycholecalciferol [calcitriol]) and calcium supplementation

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What are the etiologic factors in children?

Sporadic, autoimmune, or agenesis/dysgenesis (i.e., 22q11 deletion syndrome, a.k.a. "DiGeorge syndrome")

What is the pathogenesis of DiGeorge syndrome?

Dysgenesis of the third, fourth, and fifth pharyngeal pouches, resulting in hypoplasia of the thymus and parathyroid glands and anomalies of the great vessels

What is transient neonatal hypocalcemia?

Decreased parathyroid responsiveness caused by sepsis, prematurity, intrauterine growth retardation, hypomagnesemia (especially in infants of diabetic mothers), maternal pre-eclampsia, or maternal hyperparathyroidism

What is the differential diagnosis of neonatal hypocalcemia?

Vitamin D deficiency; high dietary phosphate load (cows' milk or swallowed maternal blood); intestinal calcium malabsorption; hypomagnesemia; permanent hypoparathyroidism

What is pseudohypoparathyroidism?

Disorder with hypocalcemia and hyperphosphatemia, but with high PTH levels, due to end-organ PTH resistance; may also have round face, short stature, obesity, brachydactyly, heterotopic calcification, mental retardation

VITAMIN D METABOLISM

What forms of vitamin D are biologically important?

Vitamin D_3 (cholecalciferol) is made in the skin from sun exposure and is the primary endogenous form. **Vitamin D_2 (ergocalciferol)** comes from plant ingestion and is the primary form in food.

25-hydroxyvitamin D is made from D_2 or D_3 by 25-hydroxylation in the liver and is the storage form.

1,25-dihydroxyvitamin D (calcitriol) is made from 25-hydroxyvitamin D by 1α-hydroxylation in the kidney by PTH and is the active form.

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What are the findings of vitamin D deficiency?

Hypocalcemia (vitamin D deficiency is the most common etiology in children) with associated symptoms (see the preceding text); rickets

List 4 metabolic results.

Hypocalcemia, hypophosphatemia, high serum alkaline phosphatase (most sensitive indicator of rickets), low 25-hydroxyvitamin D

List 7 risk factors for vitamin D deficiency.

Darkly pigmented skin, northern latitude inhabitancy, avoidance of sunlight exposure, exclusive breast-feeding, vitamin D–deficient mother, vegan diet, malabsorptive disorders (cystic fibrosis, short gut, celiac disease, inflammatory bowel disease)

What is the therapy?

Acute *symptomatic* (i.e., tetany or seizures) hypocalcemia is treated with IV calcium, switching as soon as possible to oral calcium and vitamin D. Calcitriol (active vitamin D) may be given to treat acute hypocalcemia, but ergocalciferol or cholecalciferol therapy is needed to replenish vitamin D stores.

Define rickets.

Defective mineralization at the growth plate, associated with defective cortical and trabecular bone mineralization

What are the clinical features of rickets?

Bone pain and splaying of metaphyses (i.e., wrist and ankle), bowing of long bones (once child can stand), rachitic rosary (widened costochondral junctions of ribs), craniotabes (thinning of outer layer of skull, easily depressible at occiput), frontal bossing

What are the radiologic signs of rickets?

Metaphyseal cupping, splaying (widening), and fraying (irregular edges)

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What are the other causes of rickets?

Hyperphosphaturic rickets ("vitamin D–resistant" or x-linked hypophosphatemic rickets); deficient 1α-hydroxylase activity or decreased 1,25-dihydroxyvitamin D receptor binding ("vitamin D–dependent rickets"), may have dental enamel hypoplasia or alopecia totalis, autosomal recessive inheritance; Fanconi syndrome (i.e., cystinosis); hypophosphatasia

PRECOCIOUS PUBERTY

What is it?

Onset of puberty more than 2.5–3 standard deviations earlier than average; traditionally, before age 8 years in females or 9 years in males. One endocrine society now recommends using younger than age 7 years in Caucasians and 6 years in African Americans.

What is usually the first sign of puberty in males?

Enlargement of the testes

What is the first sign of puberty in females?

Thelarche (breast development)

Is precocious puberty more common in boys or girls?

Girls

What is the most common cause of precocious puberty in girls?

Idiopathic precocious puberty

What is meant by central precocious puberty (CPP)?

Precocious puberty secondary to increased release of gonadotropin-releasing hormone from the hypothalamus

List 4 causes of CPP.

1. Idiopathic true or central precocious puberty
2. Chronic exposure to sex steroids
3. CNS pathology (trauma, tumor, hamartoma, Rathke's cleft cyst, or malformation)
4. Postinfectious or postinflammatory conditions

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What is peripheral precocious puberty (PPP, a.k.a. "precocious pseudopuberty")?

Puberty caused by the release of sex steroids by mechanisms other than the usual hypothalamic-pituitary-gonadal axis; typically hormones come from gonads or adrenals, not stimulated by gonadotropins

List 6 causes of PPP.

Exogenous hormones; hormone-producing tumors or cysts (e.g., adrenal, ovarian, testicular, hCG-secreting germinomas); congenital adrenal hyperplasia (CAH); McCune-Albright syndrome; male-limited gonadotropin-independent precocious puberty; primary hypothyroidism

What is McCune-Albright syndrome?

A syndrome consisting of the triad of precocious puberty, hyperpigmented macules, and fibrous dysplasia of the bones; may also have excess growth hormone (GH) or thyroid production; is due to G_s activating mutation

What is premature adrenarche?

Premature appearance of secondary sexual hair due to early activation of adrenal androgen production or due to increased sensitivity to adrenal androgens; thought to be unrelated to gonadotropin production

List 2 ways in which premature adrenarche differs from precocious puberty.

1. Premature adrenarche has no other features of puberty, such as physical changes in genitalia or breasts or increased linear growth velocity.
2. The bone age is not very advanced.

What is premature thelarche?

Premature breast development; it is a form of incomplete puberty and is often seen in the first 2–5 years of life.

In what way does premature thelarche differ from precocious puberty?

In premature thelarche, there are no other signs of pubertal development.

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How are premature adrenarche and premature thelarche evaluated?

Many patients require no laboratory testing. Careful clinical follow-up is the key to excluding precocious puberty, monitoring linear growth and progression of further pubertal development. Sometimes tests of bone age are helpful if there has been recent increased growth. CAH can be excluded in premature adrenarche patients by obtaining a 17-hydroxyprogesterone level.

What is a "bone age"?

The skeletal age of a child, determined by a radiograph, typically of the left hand and wrist, which is compared with standards for a given age and used to examine the degree of epiphyseal maturation

List the 2 components of an initial evaluation of precocious puberty.

Detailed history (including evaluation of previous growth data and current growth velocity); physical examination

What is included in the laboratory evaluation?

Laboratory studies may include serum levels of follicle-stimulating hormone (FSH), luteinizing hormone (LH), estradiol, testosterone, and adrenal androgens (DHEAS and 17-OHP).

What radiographic studies are indicated?

Plain radiographs to assess the bone age may be helpful. If clinical and laboratory test findings suggest CPP, a brain MRI with pituitary visualization sequences (with and without contrast) is needed in any boy and in girls < 7 years. Girls aged 7–8 may need an MRI, if indicated. If findings suggest PPP, ultrasound or CT examination of the pelvis (ovaries) or abdomen (adrenal glands, other masses) may be indicated.

GYNECOMASTIA

What is it?

Benign proliferation of the glandular breast tissue in a male

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What is pseudogynecomastia?

Fat deposition in the breast area *without* glandular proliferation, seen in obese boys and men

Name 5 causes of gynecomastia.

1. Pubertal gynecomastia, seen in up to 50% of boys, peaks between the ages of 13 and 14, may initially be asymmetric on presentation, and is typically gone within 18 months.
2. Infantile gynecomastia, seen in up to 90% of infants in the first 2–3 weeks of life, is secondary to circulating maternal hormones; may have a small amount of galactorrhea.
3. Drugs: use and abuse of marijuana and alcohol, in addition to medications such as spironolactone and ketoconazole. Recently, the use of lavender and tea tree oil as well as hair and skin care products made from placental byproducts has been implicated.
4. Primary hypogonadism, such as that seen in Klinefelter syndrome
5. Neoplasms, especially of the testes and adrenals

Is surgery ever warranted?

Yes, excision of breast tissue may be warranted for gynecomastia when it is painful or when large breast size adversely affects self-image. In the latter instance, excision can greatly improve body image and self-esteem. The placement of the incision will depend on the amount of tissue that is to be removed. Pseudogynecomastia should be evaluated after the patient has adhered to a diet and exercise regimen to lose weight. If gynecomastia persists, resection of breast tissue may be necessary.

DELAYED PUBERTY

What is it?

Delay in the onset of development of secondary sexual characteristics

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After what age is puberty considered delayed?

If there has been no development of secondary sexual characteristics by age 12 years **in females** or 14 years **in males**

What is hypergonadotropic hypogonadism?

Delayed puberty associated with increased levels of FSH and LH

What is its pathophysiology?

The pituitary is functioning, but there is lack of peripheral response from the gonads, that is, primary gonadal failure

What is hypogonadotropic hypogonadism?

Delayed puberty associated with inadequate levels of FSH and LH

What is its pathophysiology?

It implies a central problem in gonadotropin production or release; it may include a structural abnormality, tumor, gene defect leading to hypopituitarism, or nutritional problem/stress (functional).

What is the most common cause of delayed puberty in boys?

Constitutional delay of puberty, a variant of normal growth, which typically has a strong family history

List 8 causes of pubertal delay in boys or girls.

Acquired hypothyroidism, constitutional delay, primary gonadal failure or dysgenesis (i.e., Klinefelter syndrome in boys, Turner syndrome in girls), gonadotropin deficiency (i.e., Kallmann syndrome: typically boys with anosmia), hypopituitarism, CNS tumors, nutritional disturbances (i.e., anorexia nervosa in both sexes), GI disorders (i.e., inflammatory bowel disease)

List 4 ways delayed puberty is evaluated.

Careful history and physical examination, hand and wrist radiograph to determine bone age, thyroid tests, serum gonadotropins

In the evaluation of delayed puberty in boys, what 3 findings indicate the need for chromosome studies?

1. Evidence of poor testicular development
2. Dysmorphic features
3. Unexplained mental retardation or developmental delay

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In girls? (List 4)

Short stature, features of Turner syndrome, other dysmorphic features, and unexplained mental retardation or developmental delay

What are the features of Turner syndrome?

Short stature, low posterior hairline, webbed neck, widely spaced or inverted nipples, shield chest, cubitus valgus deformity, short fourth metacarpal, sensorineural hearing loss, renal malformations, cardiac malformations (bicuspid aortic valve, coarctation), lymphedema of hands and feet as infants, recurrent otitis media, high arched palate

What is the treatment of delayed puberty in boys?

Treatment of the underlying cause. If this is not possible, the physician may consider a brief course of **long-acting testosterone esters** by injection. A longer course may be indicated if puberty does not develop within 12 months, which may indicate an underlying pathology beyond constitutional delay.

What is the treatment of delayed puberty in girls?

Treatment of the underlying cause. If this is not possible, the physician may consider a **low-dose conjugated estrogen,** increasing the dosage gradually for about 1 year to mimic natural pubertal levels. This is most commonly used for Turner syndrome, other causes of primary ovarian failure, or for hypogonadotropic hypogonadism. Menarche can be achieved later by adding a progestational agent.

GH DEFICIENCY

What is GH?

It is an anterior pituitary hormone that causes growth of all tissues, especially bone and cartilage. It promotes mineralization of bones and has a role in carbohydrate and lipid metabolism.

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What are the common causes of GH deficiency?

1. Idiopathic or congenital
2. Histiocytosis, sarcoidosis, craniopharyngioma
3. Secondary to CNS trauma or infection
4. A sequela of surgery, chemotherapy, or irradiation for CNS tumors

Are newborns with GH deficiency usually of normal size?

Yes, because GH is not necessary for fetal growth

What are some potential presenting signs and symptoms of GH deficiency in childhood?

Short stature with growth velocity < 5 cm/yr, decreasing after 6–12 months of age; mild truncal obesity; frontal bossing; delayed dental development; sometimes hypoglycemia; microphallus in males; flat nasal bridge, high-pitched voice, central incisor, cleft palate, or other midline facial defects

List 3 diagnostic findings of GH deficiency.

1. Delayed skeletal development (shown by bone-age radiograph)
2. Low serum GH surrogates (IGF-1 and IGF-BP3)
3. Abnormal response to GH stimulation tests (random GH levels are not helpful because of pulsatile secretion)

List 2 potential associated conditions.

Panhypopituitarism; optic nerve hypoplasia (ONH; a.k.a. "septo-optic dysplasia" [SOD])

What is the treatment for GH deficiency?

Recombinant GH via daily subcutaneous injection

Other than GH deficiency, what are some other indications for the use of GH therapy?

Turner syndrome, Prader-Willi syndrome, chronic kidney disease, small-for-gestational age infants who have not caught up by age 2 years, and idiopathic short stature

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PANHYPOPITUITARISM

Which hormones are affected in panhypopituitarism?

Anterior pituitary hormones, in order of likelihood to be affected: GH, LH/FSH, thyroid-stimulating hormone (TSH), and adrenocorticotropic hormone (ACTH).

What are the causes?

Idiopathic; sequelae of the treatment of CNS tumors; trauma; congenital midline defects; familial causes, including single gene defects; ONH; hypothalamic disease can produce symptoms identical to primary pituitary disorders.

What is ONH or SOD?

Patients with ONH have small optic disc(s) with thin optic nerves and chiasm; in SOD, there is also absence of the septum pellucidum and agenesis of the corpus callosum. Hypopituitarism can develop in either ONH or SOD due to migrational defects of hypothalamic/pituitary neurons. Symptoms of hypopituitarism can develop gradually; frequent, regular endocrine follow-up is necessary.

List 6 complications of panhypopituitarism in infants.

Hypoglycemia; prolonged jaundice; apnea; hypotonia; microphallus in males; glucocorticoid insufficiency

How is panhypopituitarism diagnosed?

Low IGF-1, low GH concentration (<7–10 ng/dL) in response to GH stimulation tests; low TSH and thyroxine (T_4); and low morning cortisol level with abnormal response to ACTH stimulation

LH and FSH typically rise in the first months of life, then are low by 6–12 months of age until puberty; therefore, measurement after the newborn period is not helpful until adolescence.

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What is the treatment?

Replacement hormones: hydrocortisone, GH, L-thyroxine (T_4), and estrogen or testosterone at the appropriate time in adolescence

THYROID DISORDERS

HYPERTHYROIDISM

List 6 causes of hyperthyroidism.

Graves disease; autonomous thyroid nodules; subacute thyroiditis; McCune-Albright syndrome; chronic lymphocytic thyroiditis; in infants, neonatal thyrotoxicosis caused by maternal Graves disease

What is Graves disease?

The most common cause of hyperthyroidism, it is autoimmune hyperthyroidism secondary to diffuse thyroid hyperplasia ("diffuse toxic goiter").

What causes Graves disease?

Thyrotropin (TSH) receptor-stimulating antibodies that activate the TSH receptors

What are the findings on thyroid function tests?

Elevated free T_4 levels or total T_3; there is increased peripheral conversion of T_4 to T_3 in hyperthyroidism making T_3 testing useful in this setting; TSH is very low or undetectable as a result of feedback suppression by high T_4; positive TSH receptor antibody or TSH-stimulating immunoglobulin levels

Which sex is affected more often?

Females, approximately 5:1

At what age are children affected?

Two-thirds of childhood cases occur in patients 10–15 years of age.

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What are the signs and symptoms of Graves disease?

Increased appetite, weight loss, loose frequent stools; heat intolerance, diaphoresis; difficulty sleeping, emotional lability, inattention, hyperactivity, deterioration of school performance, anxiety; weakness and inability to participate in sports; tachycardia, increased systolic blood pressure with wide pulse pressure, tremor; proptosis/exophthalmos, lid lag, pain with eye movement; thyroid gland that is diffusely enlarged, smooth, nontender, and homogeneous with possible bruit; warm, moist, smooth skin; brisk deep tendon reflexes with rapid relaxation phase

What is Graves ophthalmopathy?

Autoimmune disease of the retro-orbital tissues, thought to be due to swollen extraocular muscles and retro-orbital connective tissue from inflammation and accumulation of glycosaminoglycans initiated by the TSH receptor antigens; may cause redness and edema of the conjunctiva, decreased mobility of the eye, or proptosis (exophthalmos)

What is thyroid storm?

A rare, **life-threatening complication** of Graves disease; uncontrolled exaggerated hyperthyroidism leads to marked hypertension, hyperthermia, tachycardia, vomiting, diarrhea, and CNS symptoms (apathy, confusion, coma); cardiac failure may occur.

List 4 factors that can cause thyroid storm.

Infection, surgery, trauma, or noncompliance with antithyroid medications

What are the therapies for thyroid storm?

β-Blockers, methimazole (PTU), glucocorticoids, Lugol solution (iodine), iodinated contrast agents

What is the natural history of Graves disease in children?

Waxing and waning hyperthyroidism with remission in up to 50% in 5 years, with possible relapses to follow

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What is presently the first-line treatment of Graves disease in children?

Antithyroid medications to block thyroid hormone production:

Methimazole (Propylthiouracil), which also blocks peripheral conversion of T_4 to T_3, now no longer recommended because of possible idiosyncratic fulminant liver toxicity

Propranolol or atenolol for relief of adrenergic symptoms

List 2 treatments that should be considered for failure or side effects of medical therapy, patient noncompliance, or recurrent hyperthyroidism.

Radioactive iodine (^{131}I) or subtotal thyroidectomy

What is the differential diagnosis of Graves disease?

Hyperthyroidism due to subacute or Hashimoto thyroiditis (early phases); autonomous thyroid nodule(s); factitious hyperthyroidism (excessive ingestion of thyroid hormone preparations); excessive TSH production (pituitary adenoma or pituitary resistance to thyroid hormone); McCune-Albright syndrome (constitutively activated G_s proteins in thyroid)

What is neonatal Graves disease?

Transient neonatal hyperthyroidism caused by transplacental passage of thyroid-stimulating immunoglobulins

List 8 presenting signs or symptoms of neonatal Graves disease.

Jitteriness, hyperactivity, stare, increased appetite, poor weight gain, tachycardia, possible cardiac failure, and thyroid enlargement in an infant born to a woman with history of Graves or autoimmune thyroid disease (even if mother has had thyroid ablation or surgery)

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List treatments of neonatal Graves disease.

Methimazole, iodide solution (Lugol solution), propranolol; steroids in extremely ill infants

What is the natural course of neonatal Graves disease?

It resolves during the first few months of life as maternal immunoglobulins are cleared from the infant's circulation; however, school-age IQ may be lower, even with adequate treatment.

HYPOTHYROIDISM

List 2 categories of hypothyroidism in children.

Congenital hypothyroidism caused by thyroid gland agenesis, dysgenesis, or enzymatic defects

Acquired hypothyroidism, which usually occurs after the first year of life, more commonly in adolescent girls

Which is more serious?

Congenital hypothyroidism

Why?

Thyroid hormone is required for normal brain growth and development during at least the first 2 years of life.

What may occur if diagnosis is delayed?

Disorder may be asymptomatic or mildly symptomatic in early neonatal period, but delay in diagnosis can lead to **mental retardation.** Diagnosis in the first week or two is optimal, after which time IQ points are lost. The longer the diagnosis and treatment are delayed, the lower the IQ.

How do infants with congenital hypothyroidism present?

Frequently, with abnormal newborn screening results; however, some hypothyroid infants will be missed! At birth, infants most often appear normal but may have prolonged jaundice. If infants are untreated, symptoms develop during 1–2 months that include poor feeding, lethargy, hypotonia, constipation, coarse facial features, large protruding tongue, large open fontanel, coarse cry, umbilical hernia, cool, dry, mottled skin, and developmental delay.

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What constitutes newborn screening for congenital hypothyroidism?

A battery of screening tests are performed which differ by state. Blood specimens are obtained by heel-stick after the first 24 hours of life. Thyroxine (T_4) or TSH is tested, depending on the state. **If there is an abnormal screening test in the newborn screen, a confirmatory venous sample for both free T_4 and TSH should be sent immediately and therapy initiated while awaiting results.**

What is the incidence of congenital hypothyroidism?

1:4,000 births

What is the treatment for congenital hypothyroidism in a newborn?

L-Thyroxine as soon as possible!

What is the differential diagnosis?

Transient hypothyroidism because of maternal blocking antibodies (low T_4, elevated TSH); thyroxine-binding globulin deficiency (low total T_4, normal free T_4, normal TSH); sick euthyroid syndrome, characterized by sick preterm infants (low T_4, normal TSH, high reverse T_3); central hypothyroidism (low T_4, normal TSH); drugs suppressing TSH release, including glucocorticoids and dopamine (low T_4, normal TSH)

What is the treatment strategy for congenital hypothyroidism?

Because of the risk of the infant developing CNS abnormalities, L-thyroxine administration should continue until 2–3 years of age, then it may be stopped in select patients (on low doses or with mild elevations in TSH at diagnosis) and free T_4 and TSH should be checked in **2–4 weeks.** If results are abnormal, the patient should be treated for life. If test results are normal, therapy can be discontinued, but follow-up tests in 2–3 months should be performed.

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What are the signs and symptoms of acquired hypothyroidism?

Slow growth; cold intolerance; decreased energy level; decreased appetite; constipation; irregular menses in adolescent girls; coarse puffy face; flattened nasal bridge; stocky habitus; dull, dry, thin hair; rough, dry, sallow skin; delayed relaxation phase of deep tendon reflexes; school performance is occasionally impaired.

List 4 etiologic factors.

Most commonly, autoimmune destruction secondary to chronic lymphocytic thyroiditis **(Hashimoto thyroiditis).** Other conditions include surgical or radioactive iodine ablation for the treatment of hyperthyroidism; goitrogens (iodides in cough syrups, seafood and kelp; soy products, cassava, millet; amiodarone, lithium, antithyroid drugs); ectopic thyroid dysgenesis

List 4 diagnostic findings.

Low free T_4; elevated TSH; thyroid antimicrosomal and antithyroglobulin antibodies often positive in Hashimoto thyroiditis; delayed bone age, which can indicate duration of hypothyroidism

What is the treatment for juvenile hypothyroidism?

Oral L-thyroxine

SIADH

What is it?

Syndrome of **i**nappropriate **a**nti**d**iuretic **h**ormone: excess antidiuretic hormone results in the expansion of vascular volume and hyponatremia.

What are the common causes?

CNS disease (trauma, tumors, meningitis, hydrocephalus), pulmonary disease (pneumonia, prolonged ventilatory support), severe nausea and emesis, and some chemotherapeutic and antiepileptic agents

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What are the signs and symptoms?

Water retention and **weight gain;** symptoms may progress to **lethargy, confusion,** and **seizures** secondary to **hyponatremia.**

List 2 components of treatment.

Fluid restriction because of inability to excrete free water; symptomatic hyponatremia may require *careful* infusion of hypertonic (3% NaCl) fluids or oral salt (if able).

ADRENOCORTICAL INSUFFICIENCY

What is it?

Adrenal insufficiency resulting in decreased cortisol and aldosterone production

What is Addison disease?

Primary adrenocortical insufficiency because of a destructive autoimmune process

List 9 other causes of adrenocortical insufficiency.

Congenital adrenal hyperplasia (CAH), Congenital adrenal hypoplasia (may also have ambiguous genitalia, pubertal disorders, or muscular dystrophy), bilateral adrenal hemorrhage (called "Waterhouse-Friderichsen syndrome" if from meningococcemia), trauma, thrombosis, infection (TB), tumors, drugs, adrenoleukodystrophy

List 11 signs and symptoms.

Weakness, fatigue, anorexia, abdominal pain, nausea, vomiting, weight loss, salt-craving, hypoglycemia, postural hypotension, and increased pigmentation (especially at pressure points, lips, nipples, buccal mucosa, palmar creases, under nails, and scarred areas of skin)

Why does "bronzing" occur?

Pigmentation due to primary adrenal insufficiency ("bronzing") occurs because of increased ACTH production, which leads to increased α-MSH (α-melanocyte-stimulating hormone) production. ACTH is produced from pro-opiomelanocortin (POMC), which makes both ACTH and α-MSH.

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What are the diagnostic findings?

Elevated ACTH and a low morning serum cortisol level that fails to rise with ACTH stimulation

There may be fasting hypoglycemia, hyponatremia, hyperkalemia, and elevated plasma renin activity.

What is the treatment?

Glucocorticoid and mineralocorticoid replacement

When must the glucocorticoid dosage be increased above normal replacement levels?

It must be increased at least 2- to 3-fold during physiologic stressors such as illness, trauma, and surgery.

What is an adrenal crisis?

A **life-threatening episode** that may be triggered by an illness or injury; it is characterized by fever, weakness, abdominal pain, vomiting, hypotension, dehydration, hypoglycemia, hyponatremia, and shock.

List 3 ways it is treated.

Rehydration; correction of hyponatremia, hypoglycemia, hyperkalemia, and metabolic acidosis; intravenous or intramuscular glucocorticoids in stress doses

CUSHING DISEASE AND SYNDROME

What is Cushing disease?

Bilateral adrenal hyperplasia secondary to increased ACTH production caused by pituitary adenoma, resulting in increased cortisol production

What is Cushing syndrome?

Increased cortisol production caused by adrenal tumors (including carcinomas), ectopic ACTH production by nonpituitary tumors, or exogenous glucocorticoids

List 7 signs or symptoms of glucocorticoid excess.

Signs and symptoms attributable to excess cortisol production or exogenous glucocorticoids are rounded face ("moon facies"), obesity, violaceous striae, large cervical fat pad ("buffalo hump"), impaired growth, hypertension, and hyperglycemia.

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What are the 2 laboratory findings in these conditions?

Elevated serum cortisol levels with loss of diurnal rhythm (may be normal in morning, but fails to decrease in evening; classically tested at midnight); elevated 24-hour urinary free cortisol

What is the treatment?

Cushing disease: transsphenoidal surgery; other options are radiation and medical or surgical adrenalectomy.

Cushing syndrome: therapy depends on the etiologic factors.

PHEOCHROMOCYTOMA

What is it?

A rare tumor of chromaffin cells, which are derived from neural crest tissue

Location of the tumor?

Most commonly in the adrenal medulla; most of the remainder occurs along the abdominal sympathetic chain, including the organ of Zuckerkandl and renal hilus. Extra-adrenal tumors may be referred to as "paragangliomas."

List 9 signs and symptoms.

Hypertension (sustained or paroxysmal), headache, vomiting, pallor, sweating, visual disturbances, weight loss, and sometimes tachycardia and tremor; hypertensive encephalopathy can be life-threatening.

What are the 2 laboratory findings?

Abnormally high serum levels of catecholamines (e.g., epinephrine and norepinephrine); high urine levels of their metabolites (e.g., metanephrine, normetanephrine, and vanillylmandelic acid)

What is the treatment?

Surgical excision of the tumor **(preoperative control of hypertension with α-blockers is mandatory)**

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What are the associated conditions?

Multiple endocrine neoplasia (MEN):

Type IIA: pheochromocytoma, medullary thyroid carcinoma, hyperparathyroidism

Type IIB: pheochromocytoma, medullary thyroid carcinoma, multiple mucosal neuromas

von Hippel-Lindau: variety of benign and malignant tumors, including pheochromocytoma

Neurofibromatosis type 1 (von Recklinghausen disease): pheochromocytomas occur in up to 5% of patients with NF type 1.

AMBIGUOUS GENITALIA

What does "ambiguous genitalia" refer to?

A constellation of congenital conditions, with a variety of causes, in which the genitalia are not phenotypically normal for either sex; the term "**disorders of sexual development**" (DSDs) is now used to describe this group of conditions.

How are DSDs classified?

1. Sex chromosome DSD, which includes Turner syndrome (45,X), Klinefelter syndrome (47,XXY), and the old term "true hermaphrodite," which is gonadal dysgenesis leading to ovotesticular DSD, where both testicular and ovarian tissue are present in a genetic male or female
2. 46,XX DSD, or virilized female, formerly called "female pseudohermaphrodite"
3. 46,XY DSD, or undervirilized male, formerly called "male pseudohermaphrodite"

What is the most common cause of 46,XX DSD, a virilized female infant?

CAH

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What causes CAH?

21-Hydroxylase deficiency is the most common cause. (11β-hydroxylase deficiency and 3β-hydroxysteroid dehydrogenase deficiency are the other steroid pathway enzyme deficiencies that can cause CAH.)

The enzyme deficiency blocks the production of aldosterone and cortisol and causes an excess formation of intermediate steroids (including **17-hydroxyprogesterone**) and adrenal androgens. The disorder has a spectrum of clinical phenotypes based on the amount of enzymatic activity. Essentially complete enzymatic deficiency of 21-hydroxylase results in deficiencies of glucocorticoids and mineralocorticoids, leading to **salt wasting** and **hypotension**, **masculinization** of external genitalia in girls, and possible bronzing.

What are the 3 causes of 46,XY DSD, an undervirilized male (phenotypic female)?

1. Inadequate testosterone production, caused by testicular dysgenesis or regression
2. Deficiencies or abnormalities of androgen receptors
3. Inadequate conversion of testosterone to dihydrotestosterone caused by 5α-reductase deficiency

List 3 ways 46,XY DSD may be discovered.

1. A female may present with virilization of external genitalia, typically during adolescence.
2. Testes may be discovered during repair of inguinal hernia in a female.
3. A female may present with lack of menses at puberty.

For what are these patients at risk?

The testes are at risk for seminoma or gonadoblastoma if left in place. Therefore, both testes should be removed after appropriate discussion with the family and the patient, with hormone replacement at puberty.

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In general, what do the genitalia in patients with ambiguous genitalia look like?

Phallus may be suggestive of a hypertrophic clitoris, with a small vaginal opening or urogenital sinus. The urethral meatus may be anywhere from the urogenital sinus to the base of the phallus to the tip of the phallus (described as Prader staging). When the phallus appears large enough to be a penis, it often has a chordee and hypospadias. The distance between the anus and the base of the urogenital sinus opening is increased compared with typical female (measured as an anogenital ratio), indicating the fusion of labioscrotal folds. There may be rugation or darkening of the labioscrotal folds.

What are the appropriate phallus sizes for a male infant?

As measured along the dorsum from the base (at the pubic symphysis) to the tip of the stretched glans:

At term: 3.5 ± 1 cm (mean ± 2.5 SD)

At 34 weeks' gestation: 3.0 ± 1 cm (mean ± 2.5 SD)

At 30 weeks' gestation: 2.7 ± 1.2 cm (mean ± 2.5 SD)

List 7 components that should be included in the general diagnostic approach for any patient with a DSD.

1. A history for any maternal drug ingestion that may suggest the presence of progestational agents or maternal virilization (i.e., excessive acne, hirsutism) that may suggest a luteoma of pregnancy.
2. A history of any genital abnormalities in relatives or unexplained infant deaths (which may have been attributable to electrolyte abnormalities caused by salt wasting)
3. Physical examination to assess gonadal location and symmetry, phallus size and shape, and evaluation of vaginal size
4. Karyotype

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5. Serum electrolytes
6. Abdominopelvic ultrasound
7. Determinations of hormonal levels: depends on history/exam, but should include 17-hydroxyprogesterone, cortisol, testosterone

As what gender should a child with a DSD be reared?

Traditional opinion has been that if there is a question about the ability to provide an adequate phallus, the patient should be reared as a female. However, some people believe that genetic status should determine gender assignment. Others advocate postponement of permanent reconstructive surgery until the individual patient can participate in the decision-making. This is a very controversial issue and careful discussion must be undertaken with the parents. Decisions must be made in close partnership with the parents.

What is the surgical approach to perineal reconstruction?

For infants who will be raised as females?

Cystoscopy is used to assess the urethral and vaginal openings because, in many of these patients, these 2 orifices join to form a common opening at the perineum (urogenital sinus) and need to be separated. Reconstruction involves clitoral recession and labial-scrotal reduction with vaginal repair. This may be staged in the infant and toddler years as necessary. If vaginal replacement is needed, this is usually done in early adolescence.

For infants who will be raised as males?

Reconstruction is performed in a staged manner. The first stage involves a release of the chordee; subsequent operations repair the hypospadias and, if needed, fix the testes in the scrotum (orchiopexy). Sometimes, a short course of testosterone therapy is given prior to repair to stimulate phallic growth to maximize surgical outcome.

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Chapter 25 Neurologic Diseases

SEIZURE

(Also see Ch 6, p. 43, for seizures and Ch 9, p. 81, for neonatal seizures.)

What is a seizure?

A paroxysmal event arising from synchronized electrical discharges of CNS neurons within cerebral gray matter that interferes with normal brain function

What features characterize a tonic-clonic seizure?

Rhythmic, generalized, jerking movements and loss of consciousness. Incontinence is common. Postictal lethargy is characteristic.

What is an absence seizure?

A brief episode (5–10 seconds) of staring and loss of consciousness, often easily induced by hyperventilation. There may be eye fluttering. These spells occur many times each day. There is no postictal state, and the patient is unaware of the seizure.

What are the characteristics of a partial complex seizure?

A sensory (smell, taste) hallucination and often an affective experience (e.g., fear, depersonalization) and is followed by loss of consciousness while the patient engages in repetitive, meaningless motor activity (automatism). A postictal state follows.

Are seizures dangerous?

Not necessarily. Prolonged seizures may cause damage through hypoxia, hypoglycemia, and other mechanisms, but the main danger is the underlying cause of the seizure or the accidents that may occur during the seizure.

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What causes seizures?

Seizures are symptoms. The physician must check for a CNS infection or metabolic problem, especially hypoglycemia. Seizures may also be triggered by fever. However, in children, the underlying cause is frequently never found; this constitutes an idiopathic seizure disorder, commonly called "epilepsy" (see Table 6–2).

Are brain tumors a cause of seizures?

Rarely

Is an EEG helpful?

It will help characterize a seizure, but does not diagnose or rule out seizure disorder. These objectives must be carried out clinically.

Should treatment be started after the first seizure?

It depends on the setting, but many clinicians do not treat a "single" seizure.

How are seizures treated acutely?

Severe seizures may require a benzodiazepine and respiratory support, but, if possible, the physician should treat the underlying cause (Ch 6, p. 46).

When should a child diagnosed with seizure disorder be treated with chronic anticonvulsants?

It is a matter of clinical judgment, but therapy is often started after a second seizure occurs, especially if it occurs soon after the first.

List 5 drugs commonly used to treat seizure disorder.

Phenytoin, carbamazepine, valproic acid, levetiracetam, and phenobarbital

What is the prognosis for seizure disorder?

Most seizure problems in children resolve spontaneously. If a child is seizure-free for 2 years on therapy, then gradual withdrawal of therapy can be considered.

ARNOLD-CHIARI MALFORMATION

What is it?

Elongation and downward displacement of the medulla and cerebellum into the spinal canal

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What are its consequences?	Obstruction of the fourth ventricle, resulting in obstructive hydrocephalus and possibly brainstem compression
What are the associated conditions?	Neural tube defects, especially myelomeningocele
List 2 ways it is treated.	Shunting, to relieve hydrocephalus; posterior fossa decompression if necessary

CEREBRAL PALSY

What is it?	A nonprogressive movement and posture disorder as a result of brain injury or malformation that occurs early in development. It is not an etiologic diagnosis but a clinical syndrome (a manifestation of static encephalopathy) that refers only to **motor** disability.
What are some etiologic factors?	
Prenatal? (list 4)	Genetic factors, toxins, placental factors, and infection
Perinatal? (list 2)	Prematurity and its sequelae, asphyxia
Postnatal? (list 3)	Infection, trauma, and asphyxia
What is the incidence?	Approximately 2 per 1,000 children
What are some signs and symptoms?	Delay in motor development with abnormalities in muscle tone, movement patterns, and reflexes
How is the diagnosis made?	**Clinical history** and **physical examination. Laboratory and imaging tests** are often needed to confirm suspected brain injury (e.g., porencephalic cyst), to rule out a progressive or degenerative neurologic process (e.g., astrocytoma, metachromatic leukodystrophy), or to define etiology (e.g., chromosome analysis).

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What are the 3 classifications of CP and their characteristics?

1. **Spastic:** subclassified topographically by the distribution of spasticity—diplegic, hemiplegic, triplegic, or quadriplegic
2. **Extrapyramidal:** subclassified by the quality of muscle tone or movement disorder—hypotonic, choreoathetoid, dystonic, or ataxic
3. **Mixed:** includes both spastic and extrapyramidal components

What are some associated disabilities?

Mental retardation, seizures, hearing or visual impairments, learning disabilities, attention deficits, dysphagia, malnutrition, poor growth, constipation, gastroesophageal reflux, and joint contractures and scoliosis

Does CP range in severity?

Yes, from minimal, with little or no functional disability, to severe, with total dependence for mobility, self-care, and feeding

What is the treatment?

Treatment is supportive and geared toward maximizing functional abilities, managing concurrent medical problems, and preventing secondary disabilities. Many disciplines are involved (pediatrics, neurology, orthopedics, speech pathology, physical and occupational therapy, special education, psychology, audiology, and orthotics).

INTRACRANIAL HEMORRHAGE

(Also see Periventricular-Intraventricular Hemorrhage, Ch 10, p. 92.)

List 5 causes of ICH in the newborn.

Trauma, asphyxia, primary hemorrhagic condition, congenital vascular anomaly, and prematurity

What commonly causes subdural hemorrhages in the infant?

A large-for-gestational-age (LGA) term infant with cephalopelvic disproportion relative to the mother

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What are the common predisposing factors for intraventricular hemorrhage in infants?

Mostly prematurity. Other risk factors include respiratory distress syndrome (Ch 10, p. 94), hypoxia or hypotension, reperfusion of ischemic tissue, pneumothorax (Ch 17, p. 230), extracorporeal membrane oxygenation, and hypertension.

When do most cases of intraventricular hemorrhage occur?

Between birth and day 3 of life

What are the common clinical manifestations?

A bulging fontanel, decreased muscle tone, lethargy, apnea, somnolence, seizures, hypotension, and bradycardia. In some cases, there may be no clinical manifestations.

How is the diagnosis made?

Head ultrasonography. CT or MRI may be used to further delineate the hemorrhage.

What are the 4 grades of intraventricular hemorrhage and their characteristics?

Please see Ch 10, p. 92.

What is the prognosis for intraventricular hemorrhage?

Risk for neurologic sequelae and fatal outcome increases significantly with increased grade.

What are the 3 long-term sequelae for survivors?

Hydrocephalus requiring ventriculoperitoneal shunt; long-term seizure disorder; significant development delay

GUILLAIN-BARRÉ SYNDROME

What is it?

An acute demyelination of peripheral nerves. It is an autoimmune syndrome and often follows a trivial viral infection.

What are the 4 signs and symptoms?

1. Extremity weakness usually begins distally and extends proximally, progressing for several days.
2. Painful sensory complaints
3. Areflexia
4. Autonomic involvement (e.g., hypotension, arrhythmias) may occur and is dangerous.

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Which diagnostic studies may be helpful?	CSF (elevated protein); electromyogram (EMG); nerve conduction velocity studies; pulmonary function tests (predict respiratory failure)
What is the differential diagnosis?	Diphtheria-associated polyneuropathy; tick paralysis. The physician must avoid misdiagnosing early Guillain-Barré as a conversion reaction.
What are the 4 treatments?	General supportive care; plasma exchange (if symptoms are severe or rapidly progressing); IVIG; mechanical ventilation if required
What is the prognosis?	Usually complete recovery

MYASTHENIA GRAVIS

What is it?	An autoimmune disorder with neuromuscular junction dysfunction that leads to weakness
What are the 3 symptoms?	**Rapidly fatigable weakness** (characteristic), double vision, upper airway weakness
How is it diagnosed?	Largely on a clinical basis, by eliciting a history of fluctuating weakness and by physical findings of rapidly fatigable weakness
What does EMG show?	Rapid loss of activity after repetitive stimulation of the same muscles
What is edrophonium chloride (Tensilon)?	A very short-acting acetylcholinesterase inhibitor administered intravenously
How is Tensilon used in assessing myasthenia gravis?	In myasthenia, Tensilon usually produces a rapid, dramatic increase in strength. A positive test is consistent with (but does not diagnose) myasthenia gravis.
What are the 3 treatments?	Acetylcholinesterase inhibitors (e.g., pyridostigmine); immunosuppressive drugs (e.g., steroids); thymectomy

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NEURAL TUBE DEFECTS

What are they?	Defects secondary to abnormal closure of the neural tube
List 4 examples of neural tube defects.	Anencephaly, encephalocele, meningocele, myelomeningocele
What causes neural tube defects?	Not known. Most are isolated (sporadic). There is an increased recurrence risk in families, suggesting multifactorial inheritance.
What is the incidence of neural tube defects?	Varies with geography, ethnicity, and other factors; possibly 1–4 of every 1,000 births
What is anencephaly?	Failure of the cranial portion of the neural tube to close, with associated cranial and brain malformations
What is the prognosis for anencephaly?	Most infants are stillborn or die shortly (within days) after birth.
What is encephalocele?	A defect in the cranium (usually posterior) with herniation of membranes (and sometimes brain tissue) through the opening
What is the prognosis?	Varies, depending on the amount of brain tissue involved in the process and any underlying brain abnormalities
How is it evaluated?	Imaging studies (e.g., ultrasound, MRI, CT scan) to assess the brain and plan for surgery, if indicated
What is a meningocele?	A defect involving vertebral arch malformation with protrusion of the meninges
Is the spinal cord usually normal?	Yes
How is a meningocele evaluated?	CT scan and MRI to rule out neural tissue involvement. A head CT should also be performed to rule out hydrocephalus.

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What is myelomeningocele?

A defect usually involving malformation of the vertebral arches with involvement of the spinal cord

What are the complications of myelomeningocele?

They depend on the location of the defect and the degree of spinal cord and nerve involvement. They include Arnold-Chiari malformation and hydrocephalus, neurogenic bladder, loss of motor function below the "neurologic level" of the lesion, lack of sphincter control, and clubfoot or contractures.

What is spina bifida occulta?

A general term referring to an interrupted vertebral column in the posterior midline (usually lower lumbar and sacral level) and intact skin. Tethering of the spinal cord may be a component of these lesions.

List 5 conditions that may be associated with spina bifida occulta.

Syringomyelia, diastematomyelia, tethered cord, dermoid cyst, dermal sinus tract

How can neural tube defects be prevented?

Women who take folic acid before pregnancy and during early pregnancy have a lower incidence of infants with neural tube defects. Any woman of childbearing age who is considering pregnancy should take folic acid.

MACROCEPHALY AND HYDROCEPHALUS

What is macrocephaly?

Large head size (i.e., greater than 95th percentile), regardless of the cause

What is hydrocephalus?

Increased CSF within the cranium

What are the 2 types of hydrocephalus?

Communicating and noncommunicating

What is the difference?

Communicating hydrocephalus is caused by decreased absorption or overproduction of CSF. **Noncommunicating** hydrocephalus is caused by obstruction of CSF flow.

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What is X-linked hydrocephalus?

A genetic form of hydrocephalus, usually affecting males, in which there is stenosis of the aqueduct of Sylvius. The gene is located on the X-chromosome.

What is the most common congenital cause of hydrocephalus?

Neural tube defects

COMMON MUSCULAR DYSTROPHIES

What is Duchenne muscular dystrophy?

An X-linked recessive disorder characterized by progressive muscle weakness, pseudohypertrophy of the calf muscles, and elevation of muscle enzymes, particularly creatine phosphokinase (CPK)

What is the molecular defect?

An abnormality (usually a partial deletion) of the dystrophin gene on the short arm of the X chromosome

When does it present?

Between 2 and 6 years of age

What is Becker muscular dystrophy?

Another disorder involving the dystrophin gene. It has a milder onset and rate of progression.

What are the 3 other types of muscular dystrophy?

Myotonic, limb-girdle, and fascioscapulohumeral

MISCELLANEOUS NEUROLOGIC CONDITIONS

What is spinal muscular atrophy (SMA)?

Disease of anterior horn cells, frequently progressive. Most are inherited as autosomal recessive traits.

What is Werdnig-Hoffmann disease?

Also known as "spinal muscular atrophy type I," it is an early-onset progressive disorder. The age of onset is usually before 6 months of age, with survival beyond 3 years uncommon. A late infantile and more slowly progressing disease is called "SMA type II."

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What is Kugelberg-Welander disease?

A juvenile-onset form of spinal muscular dystrophy. Age of onset is usually in the child's first decade, but it may be later; also known as "SMA type III"

What is Reye syndrome?

A mitochondrial metabolic encephalopathy, frequently associated with liver dysfunction and fatty changes in the liver. Many inborn errors of metabolism may present with features similar to Reye syndrome.

What is the cause?

Unknown; it is often seen after viral infections (e.g., varicella, influenza). A causal relationship to aspirin use has been suggested but never proven (however, use of aspirin is generally discouraged in children unless there is a specific indication). All suspected patients should be exhaustively evaluated for underlying metabolic disease.

What are the components of diagnosis?

1. Elevated hepatocellular enzymes in serum
2. Hyperammonemia
3. Exclusion of other diagnoses (such as medium-chain acyl-CoA dehydrogenase deficiency or other fatty acid oxidation defects)

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Chapter 26 Neoplastic Diseases

What is C.O.G. (a.k.a. "CureSearch")?

Children's **O**ncology **G**roup. It is an organization (study group) that compiles data from virtually all centers in the United States and Canada that care for children with cancer. It generates studies and protocols to treat childhood tumors in a systematic way. Goal is to optimize survival with minimal toxicity from therapy. COG encompasses the previous Pediatric Oncology Group (POG), Children's Cancer Study Group (CCSG), National Wilms Tumor Study Group (NWTS), and Intergroup Rhabdomyosarcoma Study (IRS).

NEUROBLASTOMA

What is it?

An embryonal tumor of neural crest cell origin

What is the incidence?

8.5 cases per 1 million children (about 500 new cases in the United States). It is the **second most common solid tumor of infancy and childhood** (brain tumors are the most common).

Where does neuroblastoma arise?

In the sympathetic nervous system: adrenal medulla (50%), para-aortic sympathetic ganglia (24%), mediastinum (20%), neck (3%), pelvis (3%)

List 5 groups of children who may be at increased risk for neuroblastoma.

Those with **other neural-crest conditions** (neurocristopathies), Beckwith-Wiedemann syndrome, adrenal hyperplasia, fetal alcohol syndrome, or those whose mothers took Dilantin during pregnancy

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List 4 associated neural-crest conditions.

Hirschsprung disease, Klippel-Feil syndrome, Waardenburg syndrome, and Ondine curse

What is the most common presenting symptom?

Abdominal mass, found in >50% of children with neuroblastoma

List 8 other presenting symptoms.

Respiratory distress, Horner syndrome, proptosis, bilateral orbital ecchymosis ("panda eyes"), paraplegia, cauda equina syndrome, bladder or vascular compression, or myoclonus with opsoclonus and nystagmus (dancing-eye syndrome)

What typical diagnostic studies are used?

CT or MRI to evaluate the primary tumor and detect metastases

Metastatic workup also includes bone scan, bone biopsy, and bone marrow aspiration.

Metaiodobenzylguanidine (MIBG) scanning may be helpful in identifying primary tumor and metastases if the origin is unknown; however, a biopsy of the primary tumor (or an obvious metastasis if more safely accessible) is usually needed for definitive diagnosis.

Tumor markers are also obtained for prognostic purposes.

What are the useful tumor markers?

Urine vanillylmandelic acid, homovanillic acid, and metanephrine levels

Other tumor markers include:

Serum neuron-specific enolase (NSE), ferritin, and lactate dehydrogenase (LDH)

Markers from the tumor itself include:

N-*myc* oncogene, TRKA proto-oncogene, DNA ploidy, integrity of chromosome 1p, CD44, VEGF-A, and a variety of genetic markers

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What are the stages and their characteristics?

The International Neuroblastoma Staging System:

Stage I: localized tumor with complete gross excision

Stage IIa: unilateral tumor with incomplete gross excision, microscopic residual, and negative lymph nodes

Stage IIb: unilateral tumor with or without complete gross excision, with positive local lymph nodes

Stage III: tumor infiltrating across the midline, or unilateral tumor with contralateral lymph node involvement, or midline tumor with bilateral extension by infiltration or by lymph node involvement

Stage IV: dissemination of tumor to distant lymph nodes, bone, bone marrow, liver, or other organs

Stage IV-S: localized primary tumor as defined for stage I, IIa, or IIb with dissemination limited to liver, skin, or bone marrow (limited to infants younger than 1 year)

What is unique about stage IV-S?

Most newborns and 30% of infants younger than 1 year present with this stage, which has an unusually good survival rate despite dissemination. Usually, no treatment is needed other than excision of the primary tumor. Chemotherapy or radiation therapy may be required if an enlarged liver compromises the infant's respiratory or nutritional status.

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List 8 favorable prognostic factors.

1. Low stage (I or II)
2. Patient age < 1 year
3. Fewer than 3 n-*myc* copies
4. Normal serum NSE, ferritin, and LDH levels
5. DNA aneuploidy or hyperploidy
6. High TRKA proto-oncogene expression
7. Intact heterogenous chromosome 1p
8. High expression of CD 44

What is the Shimada classification?

A method of determining favorable or unfavorable histology for neuroblastoma tumors (see Table 26–1)

What is mitotic karyorrhexis index (MKI)?

Refers to nuclear fragmentations and is the sum of the necrotic tumor cells, cells with mitoses, and cells with malformed, lobulated, or pyknotic nuclei per 5,000 cells examined

What is the treatment for stages I and II?

Primary surgical excision. Chemotherapy may be needed for stage II tumors with poor histology.

Small adrenal tumors that are found on prenatal ultrasound may be amenable to observation with serial blood and imaging studies to see whether they resolve spontaneously.

Table 26–1. Modified Shimada Pathologic Classification of Neuroblastoma Tumors

Appearance	Favorable Histology	Unfavorable Histology
Stroma-rich	Well-differentiated (ganglioneuroma) Ganglioneuroblastoma, nodular	Ganglioneuroblastoma, intermixed
Stroma-poor (neuroblastoma)		
Age < 18 mo	MKI < 4%	MKI > 4% or undifferentiated
Age 18–60 mo	MKI < 2% and differentiated	MKI > 2% or undifferentiated or poorly differentiated
Age > 5 years	None	All

MKI, mitotic karyorrhexis index.

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For stages III and IV?

Preoperative chemotherapy may be needed to shrink the tumor before resection. Radiation may be part of the postoperative regimen. Bone marrow transplantation (BMT) with prior total body irradiation may be used in stage IV tumors. Bone marrow is also used in some advanced-stage tumors.

What is the overall survival rate?

Overall survival rate for infants younger than 1 year is 72%, and for older than 1 year is 32%. However, survival is about 90% for children with stages I and II in both age groups, with significantly worse prognosis for stages III and IV. Stage for stage, children younger than 1 year have a better prognosis.

WILMS TUMOR

What is it?

An embryonal tumor of renal origin

What is the incidence?

500 new cases in the United States each year. Wilms tumor represents slightly more than 10% of all childhood cancer cases.

What is the age at diagnosis?

Usually 1–4 years of age

List 9 conditions with an increased risk of Wilms tumor.

1. Sporadic aniridia
2. Hemihypertrophy
3. Beckwith-Wiedemann syndrome
4. Neurofibromatosis
5. Genitourinary tract anomalies
6. Denys-Drash syndrome
7. Klippel-Trenaunay syndrome
8. Perlmann syndrome
9. WAGR syndrome

What are some key genetic markers for Wilms tumor?

1. *WT1* gene
2. Abnormal expressions of 11p15, 12p15, 16q, 1p, p53 genetic sites

What is Beckwith-Wiedemann syndrome?

Overgrowth syndrome that includes macroglossia, exomphalos, visceromegaly, hyperinsulinemic hypoglycemia

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What is WAGR syndrome?

Wilms tumor
Aniridia
Genitourinary malformations
Mental **r**etardation

What is Denys-Drash syndrome?

Pseudohermaphroditism, progressive glomerulopathy, Wilms tumor (associated with mutations on *WT1* gene)

List 3 signs and symptoms of Wilms tumor.

Large, palpable, painless abdominal mass; gross hematuria may be noted in 10–15% of cases; elevated BP may be noted in 20% of cases.

List 4 components of diagnosis of Wilms tumor.

1. The tumor is usually identified via CT scan.
2. Ultrasound to assess extension of tumor into the vena cava
3. Chest radiograph or CT scan rules out pulmonary metastases.
4. Diagnosis and staging are determined during surgical excision.

What are the stages and their characteristics?

Stage I: unilateral tumor without capsular involvement; it is completely resected.

Stage II: unilateral tumor with renal capsule or perivascular involvement; it is completely resected.

Stage III: unilateral tumor with incomplete resection, regional lymph node involvement, preoperative tumor rupture, or intraoperative tumor spill

Stage IV: metastasis to lung, bone, brain, liver, or distant lymph nodes

Stage V: bilateral renal tumors

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What is the treatment?

Surgical excision is followed by chemotherapy, depending on the stage. (Stage I tumors may not need chemotherapy.) Radiation is needed for advanced-stage tumors. Preoperative chemotherapy or radiation, or both, are sometimes used for very large tumors or tumors with extensive caval or atrial involvement.

List the 2 main pathologic categories, with examples of types included in each.

1. Favorable histology (89% of cases) includes blastema, epithelial, mixed, cystic, and glomerular types.
2. Unfavorable histology includes anaplastic types.

(Clear cell sarcoma and rhabdoid histology were earlier considered unfavorable histology but are now considered individual tumor types separate from Wilms tumor.)

What is the prognosis?

Overall survival rate for patients is 80% (90% for favorable histology). Survival rate approaches 95–100% for stage I and II tumors.

What is mesoblastic nephroma?

A renal tumor that usually presents in infants younger than 3–4 months. Presentation may be similar to that of Wilms tumor. Ninety-five percent are benign and surgical resection is the only treatment necessary.

What is nephroblastomatosis (nodular renal blastema)?

A capsular nest of primitive metanephric epithelial rests around the rim of the kidney. These may progress to Wilms tumor. Patients are treated with chemotherapy when these nests are found.

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HODGKIN DISEASE

What is it?	A malignant lymph node disorder of unknown etiology
What is the incidence?	5% of childhood malignancies; 6 cases per 1 million children
At what ages is it most common?	The first peak is 15–40 years of age (young adult form), and the later peak is 45–55 years of age (adult form); 15% of patients are younger than 16 years (childhood form).
What is the most frequent presenting finding?	Painless cervical or supraclavicular lymphadenopathy
List 3 other groups of presenting signs and symptoms.	1. Enlarged axillary or inguinal lymph nodes 2. Mediastinal involvement may cause respiratory distress, but this is more common in non-Hodgkin lymphoma. 3. Fever, night sweats, and weight loss (i.e., the "B" symptoms)
How is the diagnosis made?	By histologic examination of a lymph node biopsy. **Reed-Sternberg cells** are pathognomonic.
List the 4 histologic types.	Lymphocyte predominance, nodular sclerosing, mixed cellularity, lymphocyte depletion
Which is the most common histologic type in children?	Nodular sclerosing (>65%)
Which histologic type has the best prognosis?	Lymphocyte predominance
Which histologic type has the worst prognosis?	Lymphocyte depletion

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What are the stages and their characteristics?

The Ann Arbor classification:

Stage I: involvement of a single lymph node region or a single extralymphatic organ

Stage II: involvement of 2 or more lymph node regions on the same side of the diaphragm, or localized involvement of an extralymphatic organ or site and its regional lymph nodes with involvement of 1 or more lymph node regions on the same side of the diaphragm

Stage III: involvement of lymph node regions on both sides of the diaphragm; other lymphatic organs may be involved

Stage IV: diffuse or disseminated disease

(Stages are further classified as "A" or "B" depending on whether or not "B" symptoms are present. A new substage E denotes minimal extralymphatic disease.)

What are the components of staging?

Staging involves clinical assessment, chest radiograph, abdominal CT, chest CT, and bone marrow biopsy. MRI may provide better imaging than CT.

Is there still a role for staging laparotomy?

With improved imaging and greater emphasis on systemic chemotherapy and less emphasis on radiation therapy, outcome is not greatly affected by staging laparotomy. If only radiation therapy is being considered for localized HD in an adolescent male, staging laparotomy may be considered, but this is uncommon.

List 3 steps involved in a staging laparotomy.

1. Splenectomy
2. Core liver biopsies of each lobe
3. Lymph node biopsies from the celiac region, splenic hilum, porta hepatis, para-aortic region, and bilateral iliac regions

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How is HD treated?

Treatment depends on the disease stage. Chemotherapy is the primary treatment. Radiation is avoided, if possible, for children still undergoing growth, and for females because of an increased risk of breast cancer. In some older children with stage I or II disease, radiation therapy alone may be sufficient.

What are the complications of therapy?

Most complications are attributable to the specific agents used in treatment and include:

Myelosuppression and cardiac toxicity (Adriamycin)

Pulmonary fibrosis (bleomycin)

Gonadal dysfunction or sterility (alkylating agents)

Neurologic impairment (vincristine, vinblastine)

Complications from radiation therapy include growth impairment, solid tumors, gonadal dysfunction, and toxicity to lungs, heart, intestine, and other organs

List 9 possible secondary neoplasms.

1. Acute nonlymphoblastic leukemia
2. Non-Hodgkin lymphoma
3. Thyroid carcinoma
4. Parathyroid adenoma
5. Soft tissue sarcoma
6. Osteogenic sarcoma
7. Breast carcinoma
8. Basal cell carcinoma
9. Melanoma

What is the prognosis for HD?

Overall survival of children with HD reaches 98%. The youngest children have the best prognosis. Even children and adolescents with stages III and IV disease can expect a 60–85% 5-year survival rate.

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NON-HODGKIN LYMPHOMA

What is it?

It is a heterogeneous group of lymphoid tumors.

What is the incidence?

7–10% of all pediatric malignancies; it is the third most common pediatric malignancy (after leukemia and brain tumors).

List the 3 most common types in childhood.

1. Lymphoblastic lymphoma
2. Small noncleaved cell (Burkitt and Burkitt-like lymphoma)
3. Large-cell lymphoma (diffuse large B cell and anaplastic)

What are the 2 possible causes of non-Hodgkin lymphoma?

Viral infections and **immunodeficiency** have been implicated. Burkitt lymphoma of the endemic type, normally found in Africa, is usually associated with Epstein-Barr virus. In the United States, where sporadic Burkitt lymphoma occurs, the Epstein-Barr virus is involved in only 10–20% of cases.

List 7 of the associated immunodeficiency conditions.

1. HIV
2. Wiskott-Aldrich syndrome
3. Bloom syndrome
4. Ataxia-telangiectasia
5. Severe combined immunodeficiency disease
6. X-linked lymphoproliferative syndrome
7. Patients who are immunosuppressed for organ transplantation

What are presenting signs and symptoms:

In lymphoblastic lymphoma?

Usually presents as an anterior mediastinal mass with respiratory symptoms or superior vena caval syndrome

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In Burkitt lymphoma or Burkitt-like lymphoma of the sporadic type?

Usually presents with abdominal symptoms, which represents tumor involvement of the bowel, manifesting as a palpable abdominal mass, intussusception, or obstruction; the endemic type of Burkitt lymphoma presents with involvement of the eye or the jaw.

In large-cell lymphomas?

They are usually extranodal, and patients present with widely disseminated disease.

How is the diagnosis made?

By biopsy and evaluation of an involved lymph node, bone marrow, or pleural fluid or ascites

List 5 ways in which non-Hodgkin lymphomas are classified.

Morphology, immunophenotype, histochemical staining, cytogenetic markers, and molecular analysis

List 7 tests that are needed for a complete workup.

1. CBC with differential
2. Liver and renal function tests
3. Serum uric acid, calcium, phosphorus, LDH, and electrolytes
4. Chest radiograph
5. Chest or abdominal CT (or both)
6. Bone scan
7. Lumbar puncture

How is non-Hodgkin lymphoma treated?

It depends on the type of lymphoma.

1. Generally chemotherapy is used.
2. Radiation therapy or steroids may be needed to reduce large mediastinal tumors when respiratory distress is present but otherwise is not typically used.
3. Bone marrow transplant may ultimately be needed.
4. If disease is intra-abdominal and localized to a bowel segment, resection may be appropriate.

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What is tumor lysis syndrome?

This can result from an overload of lysed tumor material into the bloodstream when the tumor is destroyed during treatment. Hyperuricemia may result, which can compromise renal function. This syndrome is particularly characteristic during the treatment of lymphoma.

How is tumor lysis treated?

During treatment, **hydration** is very important. If tumor lysis occurs, allopurinol is given and $NaHCO_3$ is added to the IV fluid to alkalinize the urine and increase the solubility of uric acid to facilitate renal clearance. If hyperphosphatemia occurs, alkalinization must be halted because calcium phosphate may precipitate. Diuretics must be used with caution; they may lower the urine pH, enhancing hyperuricemia.

LEUKEMIA

What is the incidence of leukemia in childhood?

3,000 new cases per year in children younger than 15 years in the United States

List 12 clinical features that may exist on presentation.

Fatigue, fever, pallor, petechiae, purpura, lymphadenopathy, hepatosplenomegaly, bone pain, joint pain, weight loss, anorexia, headache

List 3 laboratory findings on presentation.

Thrombocytopenia, anemia, low (or high) total WBC count

What are the 4 predisposing conditions?

Down syndrome, Fanconi anemia, Bloom syndrome, Wiskott-Aldrich syndrome

List 5 ways leukemias are classified.

According to cell morphology, chromosome abnormalities, staining properties, surface antigens, clinical behavior (rapidity of onset)

What is the most common leukemia in childhood?

Acute lymphoblastic leukemia (ALL)

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ACUTE LYMPHOBLASTIC LEUKEMIA

How common is ALL?	ALL accounts for 80–85% of childhood leukemias.
At what age is the peak incidence of ALL?	4 years of age
List 3 good prognostic features.	Child is 1–10 years of age; WBC count $< 50{,}000/\mu L$; certain chromosomal trisomies such as 4 and 10
List 3 poor prognostic features.	Child $<$ 1 year or $>$ 10 years of age; WBC count $> 50{,}000/\mu L$; certain chromosomal abnormalities such as t9:22
List 3 ways leukemia is diagnosed.	By examination of bone marrow aspirate; cell surface marker studies; karyotype
List the 4 types of ALL.	Precursor B-lineage ALL, infant ALL, T-ALL, and mature B-ALL
What is induction?	4- to 6-week initial treatment phase for rapid reduction of leukemic burden
List 4 usual medications for induction in ALL.	Usual medications include a corticosteroid, vincristine, and asparaginase with or without anthracycline.
How successful is induction in ALL?	98% of patients achieve remission.
What is consolidation (intensification)?	Multiagent chemotherapy regimens in some protocols that lead to further reduction in malignant cells
What constitutes remission?	Decrease of blast cells in bone marrow to $<5\%$, with normalization of peripheral blood counts and, in newer classifications, low or absent levels of minimal residual disease
What is maintenance therapy?	Longer-term treatment with multiagent chemotherapy designed to further reduce the chance of recurrence of the leukemia

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List 4 commonly used maintenance drugs.

Methotrexate, 6-mercaptopurine, corticosteroid, vincristine

What is CNS prophylaxis?

Treatment to prevent leukemia relapse in the CNS

Why is this necessary?

The CNS is a sanctuary for leukemia cells, and systemic medications may not adequately penetrate the CNS.

List 3 methods of CNS prophylaxis.

Intrathecal medications (e.g., methotrexate, hydrocortisone, ARA-C), radiation, higher dosage of systemic methotrexate

What is the overall cure rate for ALL?

About 80%

ACUTE MYELOCYTIC LEUKEMIA

What is the incidence of AML?

850 new cases per year in the United States in children 15 years old or younger. It accounts for 15–20% of childhood acute leukemias.

List 9 presenting signs and symptoms.

Fever, anemia, pallor, pain (particularly bone pain), bleeding, bruising, hepatosplenomegaly, DIC, skin nodules

List 4 poor prognostic features at presentation.

Organomegaly, high WBC count, DIC; certain chromosome abnormalities indicate a poor prognosis.

How many subtypes of AML are there?

At least 8 (based on French-American-British [FAB] classification system)

List 4 conditions that predispose a child to AML.

Fanconi anemia, Down syndrome, Bloom syndrome, Kostmann syndrome

What is the significance of chromosome abnormalities in AML?

Chromosome abnormalities are common in AML, and some may be associated with an improved prognosis. Chromosome 7 abnormalities are associated with a relatively poor prognosis.

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What is the treatment for AML?

Treatment is usually more intensive than that for ALL. Induction medications may include ARA-C, daunorubicin, and other more experimental drugs. BMT should be considered if there is a suitable donor.

What is the outcome of AML?

Most patients achieve an initial remission, but long-term survival is worse than that for ALL. Survival approaches 70% if there is a suitable bone marrow donor. Survival is about 50% if only chemotherapy is used.

RETINOBLASTOMA

What is it?

The most common childhood eye tumor. It arises from primitive cells of the retina before differentiation.

What is the incidence?

About 1 in 20,000 children

List 2 symptoms with which patients may present.

Strabismus; abnormal red reflex (the reflex actually appears white [leukocoria] because of reflection of light off the tumor surface)

Is retinoblastoma hereditary?

About 40% of cases are familial; the remainder are sporadic.

What causes retinoblastoma?

Loss of function of both allelic copies of the retinoblastoma gene (*RB1*), a tumor-suppressor gene on chromosome 13

What are the 2 goals of treatment?

Eradication of the tumor and retention of vision

What studies may be included in the workup of retinoblastoma?

CT scan, ultrasound, MRI. Bone marrow aspirate and lumbar puncture may be indicated to assess for metastases.

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List 3 treatments of unilateral retinoblastoma.

1. Enucleation if no potential for vision exists
2. If tumor is small, laser or cryotherapy is preferred.
3. If tumor is large, chemotherapy is used with possible radiation or brachytherapy as required.

How is bilateral retinoblastoma treated?

Chemotherapy and subsequent local control with laser, cryotherapy, or hyperthermia

What is the prognosis?

A 90% survival rate may be expected when enucleation of a unilateral tumor can be performed. Good survival is also expected with vision-sparing strategies. Bilateral disease has a poorer outcome. Potential side effects of therapy include adverse radiation effects (cataracts, impaired orbital growth, lacrimal dysfunction, retinal vascular injuries) and secondary malignancies induced by chemotherapy.

For what other type of tumors are patients with retinoblastoma at risk?

Osteosarcomas, particularly in patients with hereditary retinoblastoma

RHABDOMYOSARCOMA

What is it?

A soft tissue tumor of skeletal muscle origin

What is the incidence?

250 new cases per year in the United States. It is the most common soft tissue sarcoma and the third most common solid tumor in infants and children. Seventy percent are younger than 10 years at diagnosis. There is a slight male predominance.

List 5 conditions that predispose a child to rhabdomyosarcoma.

Li-Fraumeni syndrome (familial cancer syndrome associated with *p53* gene mutation), Werner syndrome, basal cell nevus syndrome, tuberous sclerosis, neurofibromatosis

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Is there family clustering of cases?	There may be familial occurrences. Also, female relatives of children with rhabdomyosarcoma may have an increased risk of breast cancer.
What are the most common ages at presentation?	Two peak age spans: 2–5 years and 15–19 years
List 3 important prognostic criteria.	Site, histology, stage
List 12 of the primary sites of rhabdomyosarcoma in children.	Orbit, paratesticular, vagina, uterus, extremity, bladder, prostate, perianal, retroperitoneal, chest wall, head, and neck
Which 2 sites are the most common?	Head and neck
List 4 sites for which prognosis is relatively favorable.	Orbit, vagina, vulva, and paratesticular sites
What are the 6 histology types and their relative prognoses?	**Favorable:** botryoid, spindle cell **Intermediate:** embryonal, pleomorphic **Poor:** alveolar, undifferentiated
What is the most common histologic type?	Embryonal (~60% of rhabdomyosarcoma cases)
List the groups (i.e., stages) of rhabdomyosarcoma, with their characteristics.	**Group I:** completely resected localized disease **Group II:** grossly resected tumor with residual microscopic disease or positive lymph nodes (removed) **Group III:** gross residual disease **Group IV:** metastatic disease

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What are the signs and symptoms?

They vary according to the site of tumor.

List 2 ways in which it is usually diagnosed.

By biopsy or at excision of the tumor after primary workup

How is further tumor evaluation carried out?

This also depends on the site of the tumor.

1. Usually MRI or CT imaging is required.
2. Chest radiographs and bone scan are used to rule out metastases.

What are the treatment options and when are they used?

Surgical excision is desired, but this may not be possible if the tumor involves vital structures. In these cases, chemotherapy and radiation may be required before tumor resection. Overall, the trend has been away from radical surgery. When the tumor can be primarily resected, chemotherapy and often radiation therapy are needed as adjuvant treatment. A significant exception is when the primary tumor arises in the orbit. In these cases, chemotherapy and radiation, without surgery, will result in a 90% survival rate.

What is the prognosis?

Overall survival rate during the third IRS trial was 70% for 5 years (90% for group I, 80% for group II, 70% for group III, 30% for group IV).

OSTEOGENIC SARCOMA

What is it?

A bone tumor characterized by spindle cells

List 6 cytologic forms in which it may occur.

It may occur in various cytologic forms, including osteoblastic, chondroblastic, fibroblastic, telangiectatic, giant-cell type, and malignant fibrous histiocytoma-like.

How common is osteogenic sarcoma?

Fewer than 500 new cases yearly, but it is the most common malignant bone tumor in children.

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List 9 risk factors for osteogenic sarcoma.	Loss of retinoblastoma (*RB1*) gene; Li-Fraumeni syndrome; Rothmund-Thomson syndrome; radiation for other malignancies; enchondromatosis; Paget disease; fibrous dysplasia; hereditary exostoses; previous radiation therapy for other malignancies
List 3 sites that are most commonly affected.	Distal femur, proximal tibia, proximal humerus
Is there a sex difference in incidence?	Males outnumber females by as much as 2:1.
Which 2 portions of the bone are most commonly affected?	Medullary cavity, metaphysis
How do patients with these tumors typically present?	Persistent pain after minor trauma is the typical history. A mass may be palpable.
What is the characteristic radiographic finding?	Periosteal elevation with a **"sunburst"** pattern of soft tissue calcifications
What is the most common mode of spread for these tumors?	To the lung via hematogenous route
What is the tumor marker for osteogenic sarcoma?	Alkaline phosphatase
What are the components of the most common treatment strategy?	Although surgical resection was often the initial treatment, preoperative chemotherapy and resection using limb salvage techniques are now the most common treatment strategy.
What is the outcome?	Stage I tumors have a 90% survival. Survival declines to 20–50% in advanced stages.

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EWING SARCOMA

What is it?	A sarcoma that normally develops in the bone marrow and consists of small blue round cells. It was once believed to be separate from primitive (a.k.a. "peripheral") neuroectodermal tumors (PNET) but is now known to be genetically identical. PNET and Ewing sarcoma are now known as Ewing's family tumors.
What causes Ewing tumors?	A variety of genetic translocations
List 6 bones that are commonly affected.	The femur, tibia, humerus, pelvis, ribs, and flat bones (e.g., the scapula); however, any bone may be affected.
What part of the bone is most commonly affected?	The midshaft
In what age group and sex does Ewing sarcoma most commonly appear?	Male adolescents
What are the typical presenting symptoms?	Bone pain followed by swelling is usually the initial symptom. The pain characteristically occurs at rest. Bone necrosis can ensue, causing fever, and thus Ewing sarcoma is often misdiagnosed as osteomyelitis.
What are the typical radiographic findings?	Disruption of the bony cortex with layers of new periosteal bone formation resulting in an "onionskin" appearance
What is the typical treatment strategy?	Usually, a combination of chemotherapy and radiation is undertaken before surgical resection of the involved bony region.
What is the outcome?	Usually, limb salvage can be achieved. The overall survival rate is approximately 75% for localized tumors. Prognosis is worse for pelvic tumors and metastatic disease.
To what site does the tumor most commonly metastasize?	The lungs

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MEDULLOBLASTOMA

What is it?	It is a tumor of the **posterior fossa.** It is characterized by "small blue round cells" of the PNET type.
Where does it originate?	From the **roof of the fourth ventricle**
What are the typical symptoms?	Signs and symptoms of **increased intracranial pressure (ICP)** including headache, vomiting, diplopia, and papilledema; **in infants, a bulging fontanel** may be present.
What is the most useful diagnostic study?	MRI
What is the age of onset?	Generally, younger than 7 years of age
What is the typical treatment strategy?	Surgical removal with associated radiation therapy; if there is residual tumor after surgical removal, chemotherapy may also be needed.
What is the outcome?	In favorable risk groups, 70–90% 5-year survival. Survival ranges to <30% in higher-risk groups.

ASTROCYTOMA

What is it?	It is a tumor of glial origin that tends to be cystic in nature.
List 2 regions in which it occurs.	In the cerebellar and intracerebral regions
What are the symptoms and signs?	They depend on the location of the tumor.
Cerebellar tumors? (list 5 symptoms)	Headache, vomiting, diplopia, papilledema, or hydrocephalus
Tumors of the cerebral tissue? (list 3)	Epilepsy, upper motor neuron signs, or even arrested growth of the opposite extremity

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What is the most useful diagnostic study?

MRI

What is the treatment?

Surgical removal of the tumor. Follow-up chemotherapy and radiation therapy may be required for high-grade astrocytoma or tumors that show residual progression postoperatively.

What is the outcome?

Cerebellar tumors have a much better outcome than cerebral tumors. Five-year survival for cerebellar tumors is 90%; for cerebral tumors 30–80%. The worse the grade, the poorer the prognosis.

HEPATOBLASTOMA

What is it?

A malignant liver tumor of embryonal origin

List the 5 types.

1. Fetal or well-differentiated
2. Embryonal (immature and poorly differentiated)
3. Mixed epithelial and mesenchymal
4. Microtrabecular
5. Anaplastic

At what age is hepatoblastoma most commonly seen?

Usually before 4 years of age. Median age is 18 months.

List 12 potential risk factors.

Hemihypertrophy, Beckwith-Wiedemann syndrome, familial polyposis, Gardner syndrome, Fanconi anemia, fetal alcohol syndrome, cirrhosis, tyrosinemia, TPN-cholestasis, type I glycogen storage disease, a variety of chromosomal aberrations, very low birth weight

What are the typical signs and symptoms?

A large right upper quadrant mass

Nausea and vomiting may also be present.

What are the pertinent laboratory values?

Serum bilirubin and alpha-fetoprotein (AFP) are commonly elevated. Human chorionic gonadotropin (HCG) is elevated in rare cases.

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What are the components of the diagnostic workup?

Serum tumor markers, a plain chest and abdominal radiograph, ultrasound examination to rule out involvement of surrounding structures or the vena cava, a CT scan of the abdomen, bone marrow aspirate, and bone scan. MRI may further delineate anatomy and tumor involvement of vascular and biliary structures.

How is hepatoblastoma treated?

Tumor resection is the primary treatment. Chemotherapy may be the initial treatment for exceptionally large tumors to reduce the tumor to a resectable size. **Complete surgical resection of the primary tumor, either before or after adjuvant therapy, is required for survival.** If a tumor is confined to the liver, but not to a resectable area, liver transplantation is considered.

What percentage of children with hepatoblastoma have surgically resectable tumors?

<50%

List the stages of hepatocellular tumors and their characteristics. This staging is based on surgical findings. (Note: This system is the same for hepatoma.)

Stage I: total resection of the specimen with clean margins

Stage II: total gross resection with microscopic residual disease

Stage III: unresectable tumor or gross residual disease

Stage IV: metastatic disease

What is the PRETEXT staging system?

This was introduced by SIOP (International Society of Pediatric Oncology). This is based on radiographic findings and assesses for number and location of involved liver segments, invasion of hepatic or portal veins, and extrahepatic and metastatic disease. It may be useful in assessing surgical resectability at presentation and tumor response to different neoadjuvant therapies.

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What is the outcome?

Children with stage 1 disease who undergo a complete resection and chemotherapy may have an 85–90% survival rate. Survival rate may be slightly higher if histology is purely fetal. Overall survival rate for all cases of hepatoblastoma is 50%.

HEPATOMA

What is it?

Hepatocellular carcinoma is an epithelial malignancy similar to that seen in adults. It accounts for about 25% of pediatric liver tumors but is rare in infants and young children.

List 16 risk factors for hepatoma.

Chronic hepatitis from hepatitis B and C viruses; cirrhosis; hemihypertrophy; Beckwith-Wiedemann syndrome; Fanconi anemia; fetal alcohol syndrome; type I glycogen storage disease; tyrosinemia; aflatoxin ingestion; hemochromatosis; hepatic venous obstruction; androgen and estrogen exposure; Alagille syndrome; α_1-antitrypsin deficiency; neonatal hepatitis; biliary atresia

In which lobe of the liver is hepatoma commonly found?

The right lobe

How and to what sites does the tumor commonly spread?

It first spreads intrahepatically via lymphatic and vascular channels. It may then extend along the hepatic veins and vena cava. Hematogenous spread is to the lung, brain, and bone marrow.

What are the typical manifestations of hepatoma?

Right upper quadrant mass, nausea, vomiting, abdominal pain, weight loss, anemia

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List 7 important laboratory tests.

1. CBC
2. Serum alanine aminotransferase (ALT)
3. Serum aspartate aminotransferase (AST)
4. Alkaline phosphatase
5. Bilirubin, which is almost always normal except in advanced cases
6. Serum AFP, which is elevated in 50% of childhood cases
7. Serum ferritin, which is elevated in virtually all cases

What are the key diagnostic tests?

1. Ultrasound determines that the mass is solid.
2. CT delineates extent of the tumor and vascular involvement.
3. Bone marrow aspirate
4. Bone scan
5. A hepatic angiogram or MRI may help determine liver and tumor anatomy and vascular variations.

What is the preferred treatment?

Complete resection is required for cure. If the tumor is too large for resection initially, chemotherapy may be used to try to shrink the tumor. Postoperative chemotherapy is always needed.

List 3 major metabolic concerns following hepatic resection.

Hypoalbuminemia, hypoglycemia, hypothrombinemia

What is the overall survival rate for children with hepatoma?

<15%

TERATOMA

What is it?

A teratoma is a tumor consisting of tissue from some or all of 3 primitive germ-cell layers (i.e., endoderm, mesoderm, ectoderm). Tissue may reveal itself in varying degrees of maturity. The sacrococcygeal area is the most common site for teratomas.

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List 2 occasions when a sacrococcygeal teratoma (SCT) is usually noted.

Prenatal ultrasound examination or at birth

List 6 other sites teratomas may be found.

Ovary, testicle, head, neck, mediastinum, retroperitoneum

List 3 signs and symptoms.

1. In sacrococcygeal area, tumor protrudes from presacral space and pushes the rectum forward. The tumor may weigh as much as the infant!
2. Tumors in other sites may manifest as physical deformities.
3. Tumors may manifest as a result of compression of surrounding structures, such as the lung or trachea.

What are the characteristic radiologic findings?

Calcifications in 50% of cases

What is the malignant potential?

Low in infants, but the potential increases with age

What are the 2 serum tumor markers for SCT?

β-HCG (choriocarcinoma) and AFP (yolk sac carcinoma)

These markers are monitored in follow-up to monitor for recurrence.

What is the treatment?

Surgical removal

What is the vascular source of an SCT?

Presacral vessels; these must be removed with the coccyx to minimize the potential for recurrence and malignancy.

What is the outcome?

If the tumor is benign, outcome is excellent (but it is necessary to monitor for recurrence of malignant tissue). If malignancy is present, outcome is poor. If the primary tumor has malignant components, recurrence is common, even if original tumor is thought to be completely excised.

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MELANOMA

What is the incidence of melanoma in children?

It accounts for 1–3% of all pediatric malignancies and 1–4% of all melanomas.

List 10 risk factors for melanoma.

1. Fair skin
2. Familial atypical mole melanoma (FAMM) syndrome
3. Xeroderma pigmentosum
4. Increased numbers of melanocytic nevi
5. Acquired nevi, especially in areas of chronic irritation or trauma
6. Giant congenital nevus
7. Atypical nevi
8. Excessive (especially intense and intermittent) sun exposure
9. Family history **(Note: Melanoma may arise in an infant as a metastasis from a mother with melanoma.)**
10. Immunosuppression

Note: In 30–50% of cases, melanoma will occur at a site without a previous nevus.

List Clark's levels of tumor invasion, with their characteristics.

Level I: tumor cells above basement membrane (i.e., in situ)

Level II: invasion of papillary dermis

Level III: tumor cells at the junction of papillary and reticular dermis

Level IV: invasion of reticular dermis

Level V: invasion of subcutaneous fat

List Breslow's classifications of tumor thickness.

In situ

<0.76 mm

0.76–1.5 mm

1.5–4 mm

>4 mm

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What determines prognosis?

Thickness of tumor and evidence of metastases. Workup should include chest radiograph and possibly whole-body CT scan in children.

What is the overall mortality rate?

40%

What is the treatment?

1. Local excision with an appropriate margin and sentinel node excision. If the sentinel node is negative, excision of the primary site may be all that is needed. If positive, completion lymphadenectomy in the draining basin is probably advisable.
2. For extensive tumors, adjuvant chemotherapy is likely to be used, but its effectiveness is not well understood in children.
3. Immunotherapy is used more frequently in adults, but its usefulness in children is not determined. It may also be very toxic in children.
4. Long-term follow-up with periodic imaging is important.

List 2 preventive measures.

Avoidance of intense sun and use of protective clothing and sunscreen; regular surveillance of questionable nevi by a dermatologist

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Chapter 27 Skin, Soft Tissue, Nail, and Hair Disorders

DEFINITIONS

Macule	Flat, circumscribed lesion with color change ≤ 1 cm
Patch	Flat, circumscribed lesion with color change > 1 cm
Papule	Elevated, circumscribed lesion ≤ 1 cm
Plaque	Elevated, circumscribed lesion > 1 cm
Nodule	Solid, circumscribed, elevated lesion
Wheal	Elevated lesion characterized by local, superficial, transient edema
Vesicle	Elevated, circumscribed, fluid-containing lesion ≤ 1 cm
Bulla	Elevated, circumscribed, fluid-containing lesion > 1 cm
Pustule	Elevated, circumscribed lesion ≤ 1 cm that contains purulent exudate
Abscess	Elevated, circumscribed lesion > 1 cm filled with purulent material
Lichenification	Thickening of the epidermis with associated exaggeration of skin markings

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INFLAMMATORY DISORDERS

SEBORRHEIC DERMATITIS

What is it?	An oily, yellow, scaly eruption, usually involving the scalp ("cradle cap" in infants), but it may also involve face, trunk, and diaper area
What is the etiology?	Unknown, possibly related to overproduction of sebum and overgrowth of *Malassezia* yeast
What is the treatment?	Gentle shampooing with a mild shampoo resolves symptoms in most infants. In refractory cases and older patients, treatment options include medicated shampoos, topical antifungal agents, and topical corticosteroids.

ATOPIC DERMATITIS (ECZEMA)

What is it?	A common chronic or recurrent inflammatory skin disorder of infancy and childhood, characterized by dry skin and an "itch-scratch" cycle
How does the patient usually present?	With erythematous, exudative, crusted patches and plaques that can progress to a scaly, lichenified dermatitis over time
What is the etiology?	Unknown, possibly an immune dysfunction leading to IgE sensitization and activation of type 2 helper T cells
What is the typical distribution?	
In infancy?	Cheeks, scalp, trunk, and extensor surface of the extremities
In childhood?	Flexor surfaces, especially the antecubital and popliteal fossae, wrists, and ankles
In adulthood?	Face, hands, feet, and flexor surfaces

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Individuals with atopic dermatitis are often prone to what 5 types of skin infections?	*Staphylococcus aureus*, β-hemolytic streptococci, herpes simplex, *Molluscum contagiosum*, fungal infections
With what is atopic dermatitis associated?	High incidence of allergies, asthma, or both
What is the differential diagnosis?	Seborrheic dermatitis, psoriasis, scabies, contact dermatitis, drug reactions, zinc deficiency, severe combined immunodeficiency
What is the natural history?	90% of patients present before 5 years of age; 60–90% of patients outgrow it by puberty.
What is the treatment?	Therapy is directed at controlling dryness (bath oils, mild soap, moisturizers, emollients), inflammation (topical corticosteroids), and itching (antihistamines). Environmental control is important. Antibiotics or antifungals are used for infections as needed.
What precautions should be used when using topical corticosteroids?	Because of potential side effects of prolonged therapy (e.g., atrophy, hypo- or hyperpigmentation, striae), use the lowest potency steroid for the shortest duration possible that effectively achieves symptom control.

CONTACT DERMATITIS

What is it?	A localized inflammation of the skin due to contact with an irritating or allergy-causing foreign substance
What are some common triggers?	Diapers; poison ivy, oak, or sumac; detergents or skin products containing dyes or perfumes; nickel-containing jewelry and belt buckles; topical medications

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What are the clinical features?	Multiple types of lesions may occur depending on the patient and the offending agent (e.g., erythema, papules, vesicles, bullae); often associated with pruritus
What is the treatment?	Identification and avoidance of triggers, topical corticosteroids

ACNE VULGARIS

See Ch 12, p. 125.

PSORIASIS

What is it?	A chronic inflammatory skin disorder, thought to be due to immune dysregulation causing epidermal proliferation
What are the clinical features?	Discrete erythematous plaques which are sometimes pruritic and often have a silvery scale; nail involvement is common and may include pits and nail destruction
What is the treatment?	Multiple options are available and include tar preparations, topical corticosteroids, ultraviolet (UV) light therapy, salicylates, and anthralin

PITYRIASIS ROSEA

What is it?	A self-limited erythematous scaly eruption of unknown etiology common in adolescents
What are the clinical features?	An erythematous annular "herald patch" on the trunk, followed by secondary eruption of erythematous scaly macules in a "Christmas tree" distribution on the trunk 1–2 weeks later
What other clinical entity should always be considered in the differential diagnosis?	Secondary syphilis

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BACTERIAL INFECTIONS

IMPETIGO

What is it?	A superficial cutaneous infection, common in children during warm weather, particularly on the face
What are the 2 types?	Bullous (30%) and nonbullous (70%)
What is the etiology?	*S. aureus* (80%) and *Streptococcus pyogenes* (20%). Presence of bullae implies *S. aureus* as the cause.
What are the characteristics of bullous impetigo?	Transparent, flaccid bullae develop on the affected skin.
What are the characteristics of nonbullous impetigo?	A small vesicle or pustule forms on a predisposing lesion and may spread to a crusted honey-colored lesion
List 5 predisposing lesions.	Chickenpox, insect bites, abrasions, lacerations, burns
What is the treatment?	Topical mupirocin may be considered for localized nonbullous lesions, but oral antibiotics are indicated for extensive lesions or bullous impetigo.
List 5 potential complications.	Cellulitis, osteomyelitis, septic arthritis, pneumonia, bacteremia
What is a rare renal complication?	Poststreptococcal glomerulonephritis, not preventable with treatment of impetigo (Ch 21, p. 334)

CELLULITIS

What is it?	Acute inflammation of the dermis and subcutaneous fat, usually caused by bacterial invasion of the skin
What are some etiologic agents?	*S. pyogenes* (group A) and *S. aureus* are most common. *Streptococcus pneumoniae*, *Haemophilus influenzae*, group B streptococcus, and *Escherichia coli* also cause cellulitis.

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List 3 features of the clinical picture.

1. Skin that is red, tender, edematous, warm, and may be indurated; borders of the lesions are indistinct and not elevated.
2. Enlarged, tender regional lymph nodes, lymphangitis
3. Fever, chills, malaise, poor appetite

List 3 mechanisms of spread.

1. Local spread: break in skin (e.g., wound, bite, excoriation, impetigo, folliculitis, carbuncle, varicella)
2. Hematogenous spread
3. Direct extension from deeper infection

List 4 groups of patients in which blood cultures are useful for determining the offending organism.

Those with systemic illness or facial cellulitis, newborns, immunocompromised patients. In most others, blood cultures are usually unrevealing.

What is the treatment?

Antibiotic therapy is aimed at the most likely causative organisms. Adjunctive therapies include warm compresses, bed rest, elevation, and pain control.

Which patients require IV antibiotics?

Newborns; immunocompromised patients; patients with high fever, systemic toxicity, vomiting, or periorbital or orbital cellulitis; or patients who show no improvement after 2 days of oral therapy. Outpatient therapy with IM ceftriaxone is an alternative for patients with capacity for reliable follow-up.

What is erysipelas?

A more superficial skin infection, frequently caused by group A β-hemolytic streptococci. Clinical characteristics include erythema and a well-defined border.

What is periorbital cellulitis?

Inflammation of soft tissues of the eye superficial to the orbital septum

List 6 causes.

Trauma, insect bites, severe conjunctivitis with spread of infection to the surrounding area, bacteremia, sinusitis, and hematogenous spread

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List 4 causative organisms.

S. aureus: usually when trauma, insect bite, or other skin infection (e.g., impetigo) is present

H. influenzae: with bacteremia

S. pneumoniae: with bacteremia

S. pyogenes (group A β-hemolytic streptococcus)

What is the clinical presentation?

Eyelid, conjunctiva, and the surrounding area are swollen, red, warm, and indurated. A purple hue to the skin may be associated with *H. influenzae*. Fever and systemic toxicity may be present.

What is involved in the evaluation of periorbital cellulitis?

CBC, blood culture, and culture of the wound or lesion (if appropriate). The physician must rule out true "orbital" cellulitis clinically or by formal ophthalmologic evaluation, a CT scan, or both, looking for orbital involvement (e.g., extraocular muscle entrapment, proptosis, swelling of the optic nerve). Lumbar puncture should be performed if meningitis is suspected.

What is the treatment?

IV antibiotic therapy

What are the 3 possible complications of true orbital cellulitis?

Compression or stretching of the optic nerve and loss of vision, cavernous sinus thrombosis, meningitis (Ch 28, p. 462)

FOLLICULITIS

What is it?

A common superficial infection of the hair follicle

What is the etiology?

S. aureus (MRSA has increased the threat of this), also some gram-negative bacteria, particularly pseudomonas

What are the risk factors?

Shaving, hot tub use, poor personal hygiene

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What are the clinical features? Small erythematous pustules in hair-bearing areas

What is a furuncle? Deep folliculitis of a single hair follicle, usually forming a tender erythematous nodule

What is a carbuncle? Confluent infection of multiple contiguous hair follicles, usually forming an abscess

What is the treatment? Depends on the extent of infection. Superficial folliculitis can often be treated with warm compresses and/or topical antibiotics, but deeper infection often requires systemic antibiotics and in some cases incision and drainage.

SCARLET FEVER

What is it? A rash associated with streptococcal pharyngitis due to toxin production by *S. pyogenes*

Describe the rash. Usually diffuse and erythematous, with a sandpaper quality. It may have darker lines in skinfold areas (Pastia's lines), and desquamate, particularly on the face.

STAPHYLOCOCCAL SCALDED SKIN SYNDROME

What is it? An exfoliative dermatitis caused by toxigenic strains of *S. aureus*

What are the clinical features? Illness often begins after an upper respiratory infection with fever, irritability, and faint macular erythema that progresses to generalized, painful, beefy red skin, often with bullae formation, diffuse desquamation, and crusting

What is the treatment? Antistaphylococcal antibiotics, typically IV

TOXIC SHOCK SYNDROME

What is it? A toxin-mediated illness characterized by fever, multiorgan system dysfunction, shock, and characteristic skin findings

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What is the etiology?	*S. aureus* or *S. pyogenes*
What are the risk factors?	Menstruation with prolonged tampon use, other foreign bodies, surgical wound infections, other soft tissue infections
Describe the rash.	Diffuse nontender macular erythroderma, sometimes with erythema of the mucus membranes, typically followed by desquamation 1–2 weeks after the onset of rash, particularly of the palms and soles
What is the treatment?	IV antibiotics, removal of potentially infected foreign bodies, meticulous critical care for multiorgan system involvement

VIRAL INFECTIONS

EXANTHEMS

Describe the following rashes:	
Measles	Erythematous maculopapular rash that generally starts on the face and spreads to the extremities usually lasting 7–10 days; Koplik spots may be seen in the mouth (Ch 28, p. 489).
Rubella	Erythematous maculopapular rash that spreads from face to extremities; shorter in duration than the measles rash (Ch 28, p. 491)
Roseola	Erythematous macular rash, typically appears at the end of the febrile illness (Ch 28, p. 492)
"Fifth disease" (erythema infectiosum)	"Slapped cheek" appearance on face, mottled or reticular rash on trunk and extremities (Ch 28, p. 492)
Varicella	Erythematous papules and vesicles, often described as "dewdrops on a rose petal," with lesions at multiple stages of development (Ch 28, p. 493)

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HAND-FOOT-MOUTH DISEASE

What is it?	A viral illness characterized by fever and a typical rash
What is the etiology?	Enteroviruses, most commonly Coxsackie A16
Describe the rash.	Erythematous papules and vesicles on the hands, feet, bottom, and posterior oropharynx
What is herpangina?	A similar illness in which only oral lesions are present

HERPES GINGIVOSTOMATITIS

See Ch 28, p. 475.

MOLLUSCUM CONTAGIOSUM

What is it?	A benign superficial eruption of dome-shaped, flesh-colored papules with central umbilication
What is the etiology?	Poxvirus
What is the treatment?	Lesions can be removed using a number of techniques if desired for cosmetic reasons or due to problematic locations

WARTS

Describe the clinical appearance.	Irregularly shaped discrete flesh-colored papules that may be smooth or rough and can occur on any skin surface
What is the etiology?	Human papillomavirus
What is the treatment?	Many warts resolve spontaneously over time, but treatment options include liquid nitrogen, salicylic acid, cantharidin, podophyllin, laser ablation, and surgical removal.

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FUNGAL INFECTIONS

CANDIDAL DIAPER DERMATITIS

Describe the rash.	Well-demarcated, raw-appearing erythematous plaques concentrated in intertriginous areas, often with satellite lesions
How is the clinical appearance different from irritant contact dermatitis due to diapers?	The erythema of irritant contact dermatitis is more ill defined, tends to spare the intertriginous areas, and is not associated with satellite lesions.
What is the treatment?	Topical antifungal therapy

TINEA (DERMATOPHYTOSIS)

What is it?	A superficial fungal infection caused by *Trichophyton*, *Microsporum*, and *Epidermophyton*, often referred to as "ringworm"
What are some common locations?	Scalp (tinea capitis), body (tinea corporis), groin (tinea cruris or "jock itch"), feet (tinea pedis or "athlete's foot"), nails (tinea unguium or onychomycosis)
What is the typical clinical appearance?	Discrete annular scaly plaques with raised edges that are often pruritic. Onychomycosis leads to thickened, discolored, dystrophic nails. Tinea capitis often presents with focal areas of alopecia due to hairs breaking at the scalp surface giving a characteristic "black dot" appearance.
What is a kerion?	A boggy inflammatory mass that develops as an immune reaction to a dermatophyte infection, typically found on the scalp in association with tinea capitis
List 4 ways tinea can be diagnosed.	1. Clinical appearance 2. Fluorescence under a Wood's lamp 3. Identification of hyphal elements under microscopic examination using potassium hydroxide (KOH) 4. Fungal culture

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What is the treatment? Topical antifungal agents for most superficial lesions. A prolonged course of oral griseofulvin is indicated for tinea capitis.

TINEA VERSICOLOR

What is it? A superficial fungal infection caused by *Malassezia furfur*

What are the characteristic lesions? Round-to-oval lesions, sometimes with a fine scale, that may be either hyperpigmented or hypopigmented and are usually located on the upper trunk and back

What are the treatment options? Selenium sulfide shampoo, topical or oral antifungal agents

PARASITIC INFECTIONS

SCABIES

What is it? An infestation of the burrowing mite *Sarcoptes scabiei*

What is the typical clinical presentation? Intensely pruritic papules, vesicles, and linear burrows with secondary excoriations

What is the typical distribution? Interdigital spaces, dorsal hands, groin, axilla, and abdomen

What is the treatment? Topical permethrin (or lindane for adolescent patients). All household members should be treated, and all linens and clothing should be washed in hot water.

LICE (PEDICULOSIS)

What is it? An infestation of the louse *Pediculosis humanus capitis* (head lice), *Pediculosis humanus corporis* (body lice), or *Pediculosis pubis* (pubic lice or "crabs")

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What is the typical clinical presentation? Intense pruritus of the affected area. In hair-bearing areas, lice and nits (eggs) can be visualized attached to hairs near the scalp. On the body, papules with secondary excoriations are often found on the trunk.

What is the treatment? Permethrin shampoo and combing to remove nits. All linens and clothing should be washed in hot water.

HYPERPIGMENTATION AND HYPOPIGMENTATION

What are hyperpigmentation and hypopigmentation? An excess or deficiency in skin pigmentation, respectively

What are the causes of pigmentation changes? Pigmentation changes may be local or generalized and are attributable to a wide variety of defects, including absence of melanocytes, defective melanocytes, overproduction of melanin, pigmentation changes induced by hormones, focal developmental defects, and postinflammatory sequelae

DERMAL MELANOSIS

What is it? Benign hyperpigmented areas frequently found on the lumbosacral and gluteal regions of newborns, previously referred to as "mongolian spots"

In which infants are they commonly found? In infants whose natural skin has medium-to-dark pigmentation

Why should this always be carefully documented? They can be mistaken for bruises in cases of child abuse or neglect.

FRECKLES (EPHELIDES)

What are freckles? Tiny light or dark brown macules with poorly defined edges on sun-exposed areas that represent areas with increased melanin due to larger melanosomes

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What patients are most prone to freckling?

Fair-skinned, light-haired patients with family history of freckling

In what genetic syndrome is axillary and inguinal freckling a diagnostic criterion?

Neurofibromatosis type 1

MOLES (NEVI)

What are they?

Small circumscribed brown macules or papules that can occur anywhere on the skin and represent areas with increased melanin due to increased numbers of melanocytes in the basal layer of the epidermis

How are they broadly categorized?

Congenital or acquired

Which ones have significant risk of malignancy?

1. Large congenital nevi
2. Any nevus with ABCDE features (asymmetry, irregular borders, atypical coloration, large diameter, elevation, and evolving features)

CAFÉ AU LAIT SPOTS

What are "café au lait" spots?

Uniformly hyperpigmented macular lesions with sharp demarcation that may be quite large in size. The true hue (not always "café au lait") is determined by the natural skin tone; the deeper the color of the background skin tone, the deeper the color of the spot.

Are café au lait spots found in otherwise healthy children?

Yes. Ten percent of healthy children have café au lait spots. A child in this group typically has 1–3 spots.

With what conditions may café au lait spots be associated?

Neurofibromatosis (von Recklinghausen disease; Ch 30, p. 516), McCune-Albright syndrome, Russell-Silver syndrome, multiple lentigines syndrome, ataxia telangiectasia, Fanconi anemia (Ch 15, p. 180), tuberous sclerosis (Ch 30, p. 516), Bloom syndrome, Chédiak-Higashi syndrome

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VITILIGO

What is it?	Acquired, sharply circumscribed depigmented macules of varying sizes and shapes
What is the etiology?	Unknown, possibly due to an autoimmune mechanism
List 5 associated conditions.	Hypothyroidism, hyperthyroidism, adrenal insufficiency, pernicious anemia, diabetes
What is the natural history?	Progression of depigmentation occurs in most patients, although spontaneous repigmentation occurs rarely.
What are the treatment options?	Repigmentation therapy options include oral or topical psoralen compounds administered together with exposure to sunlight or UV light sources, topical steroids, and laser therapy. Repigmentation may be partial and may take many months to occur. It is important to protect depigmented areas from excessive sunlight, because there is no protection provided by melanocytes. Depigmentation therapy may be attempted in selected patients.

ALBINISM

What is it?	A group of conditions in which there is failure or deficiency of melanin production in the skin, hair, and eyes
What disorders are included in this group?	Oculocutaneous albinism, ocular albinism, Chédiak-Higashi syndrome, Hermansky-Pudlak syndrome, Griscelli syndrome
What is partial albinism?	Also called "piebaldism," it is characterized by sharply demarcated amelanotic patches on the forehead, anterior scalp, ventral trunk, elbows, and knees

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What is the inheritance pattern of partial albinism?	Autosomal dominant
What is the underlying genetic defect in partial albinism?	Mutation in the KIT proto-oncogene or in the gene for *SNAI2*
What is the histology of the depigmented regions in partial albinism?	Absence of melanocytes and melanosomes

HYPERSENSITIVITY REACTIONS

HIVES (URTICARIA)

What is it?	A hypersensitivity reaction to a variety of stimuli characterized by pruritic, raised wheals caused by degranulation of mast cells
What are some common triggers?	Infections, medications, foods, skin care products, irritating substances, exercise, sun or cold exposure
What is the treatment?	Antihistamines, identification and avoidance of offending agents

ERYTHEMA MULTIFORME

What is it?	A hypersensitivity reaction to a variety of stimuli characterized by an eruption of erythematous annular lesions, often associated with nonspecific systemic symptoms
What are some common triggers?	HSV-1 infection is the most common cause overall, and *Mycoplasma pneumoniae* is the most common bacterial cause. Many other triggers exist, including infections, medications, malignancies, immunologic disorders, and mechanical forces.

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What is the distribution of the lesions?	Erythema multiforme (EM) minor: often localized to the extremities EM major: acral distribution as above plus involvement of at least 1 mucus membrane
What is the treatment?	Identification and avoidance of triggers, cessation of any offending drug, supportive care
What is the prognosis?	Most cases are self-limited and resolve without complication over 2–4 weeks, but some cases may progress to Stevens-Johnson syndrome (SJS) or toxic epidermal necrolysis (TEN)

STEVENS-JOHNSON SYNDROME

What is it?	A hypersensitivity reaction predominantly to drugs; characterized by extensive mucosal involvement and cutaneous lesions including atypical target lesions and inflammatory vesiculobullous lesions
What are the most common triggers?	NSAIDs, antibiotics (particularly penicillins and sulfonamides), certain anticonvulsants
What is the prognosis?	Mortality rate 5%

TOXIS EPIDERMAL NECROLYSIS

What is it?	A hypersensitivity reaction almost exclusively to drugs; characterized by severe, extensive mucosal involvement and full epidermal thickness necrosis
What is the prognosis?	Mortality rate 30%

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What is the relationship among EM, SJS, and TEN?

Controversy exists in the literature. Most experts view EM and SJS as distinct clinical entities but support the idea that SJS and TEN exist along a continuum of a single disease process. Percentage of epidermal detachment based on total body surface area is often used as a diagnostic tool, where <10% is consistent with SJS, 10–30% is consistent with an SJS/TEN overlap, and >30% is consistent with TEN.

BENIGN MASS LESIONS

HEMANGIOMA

What is it?

Abnormal proliferation of vascular endothelial cells, resulting in tumors of varying sizes and types composed of abnormal blood vessels

How do people with hemangiomas present?

Although most are visible on the skin, hemangiomas may involve any organ. Presentation may relate to the effect on the involved organ. In addition, large hemangiomas (particularly of the liver) may cause heart failure because of arteriovenous shunting, or purpura because of consumption of platelets (Kasabach-Merritt syndrome).

How are they diagnosed?

Cutaneous lesions are easily diagnosed by physical examination. Ultrasound or CT may be required for intracorporeal lesions.

May hemangiomas be multiple?

Yes. Discovery of 1 hemangioma should prompt a search for others. The examiner should ask about any evidence of airway obstruction.

What is the natural history of a hemangioma?

Growth of the hemangioma for the first 1–1.5 years, followed by involution; 80% are gone by 5 years of age

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List 2 treatments.

If functional difficulties should arise, oral or injected steroids may be used initially; alpha interferon (IFN-α) has also been effective.

List 7 indications for medical therapy.

1. Obstruction of vision
2. Thrombocytopenia
3. Obstruction of luminal organs
4. Uncontrollable hemorrhage or ulceration
5. Repeated infection
6. Cardiac compromise because of arteriovenous shunting
7. Increasing size causing symptoms (e.g., enlarging liver hemangioma impeding diaphragmatic excursion)

List 2 indications for surgical resection.

Any indication listed in the preceding text that does not respond to medical therapy; symptoms too severe to wait for medical results

LYMPHANGIOMA

What is it?

A benign tumor of the lymphatic system that consists of large or small saccules of lymph fluid

What is "cystic hygroma"?

This term is usually used to describe a large, primarily cystic lymphangioma of the neck.

When do patients usually present with lymphangiomas?

At or soon after birth

Where are lymphangiomas located?

Anywhere in the body, but most commonly the neck, axilla, mediastinum, groin, and lower abdomen

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What are the symptoms? Usually presents as a painless mass, but lymphangiomas of the neck, tongue, or glottic regions may cause respiratory distress. Occasionally, a lymphangioma may present as an acutely enlarging mass as a result of infection or inflammation.

Do lymphangiomas regress? No

List 2 treatments. Surgical excision (re-excision may be necessary in 10–15% of cases); sclerosing agents (mixed results)

SYNDROMES WITH PROMINENT CUTANEOUS FEATURES

NEUROFIBROMATOSIS

See Ch 30, p. 516.

TUBEROUS SCLEROSIS

See Ch 30, p. 516.

STURGE-WEBER SYNDROME

What is it? A condition characterized by a constellation of symptoms, the most obvious of which is a facial hemangioma (**port wine stain**). Seizures, hemiparesis, intracranial calcifications, and mental retardation may be components of this condition. (**Note:** Not all patients with a facial port wine stain have Sturge-Weber syndrome.)

What is the etiology? Thought to be caused by anomalous development of the vascular bed during cerebral vascularization

What are the 4 clinical manifestations, and their characteristics?

1. Facial hemangioma is present at birth. It may extend to the lower face, the trunk, and the mucosa of the mouth and pharynx and frequently occurs in the distribution of the trigeminal nerve.

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2. Seizures, if present, usually occur within the first year of life and become increasingly refractory to therapy.
3. Mental retardation may be present; more common in patients with seizures
4. Ocular manifestations include buphthalmos and glaucoma.

How is the diagnosis made?

1. The constellation of symptoms
2. Radiograph of the skull: the presence of intracranial calcifications on radiograph, together with the symptoms, suggest the diagnosis
3. CT scan of the head: may show unilateral cortical atrophy and ipsilateral dilation of the lateral ventricle

What are the 4 components of treatment?

1. Control of seizures
2. Hemispherectomy or lobectomy (for intractable seizures)
3 Laser therapy (for facial hemangioma)
4. Monitor ocular pressure for glaucoma

EHLERS-DANLOS SYNDROME

What is it?

A group of inherited connective tissue disorders characterized by skin hyperextensibility, abnormal wound healing, and joint hypermobility

What is the etiology?

Thought to be a genetic defect in 1 of the collagen genes

What are the characteristic skin findings?

Skin is smooth, velvety, hyperelastic, and fragile. Scars are typically widened and atrophic ("papyrus scars").

What is the inheritance pattern?

Depends on the type. Some types are autosomal dominant, some are autosomal recessive, and some are X-linked.

What are the common complications?

Joint dislocations, hernias, organ prolapse, cervical insufficiency, chronic pain

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Which type of Ehlers-Danlos syndrome (EDS) is characteristically associated with decreased life expectancy?

Vascular type (type IV), due to a defect in type III collagen. Patients with EDS type IV often have arterial, intestinal, and/or uterine fragility that can cause premature death due to vascular rupture or dissection, gastrointestinal perforation, or uterine rupture.

WAARDENBURG SYNDROME

What is it?

An inherited auditory-pigmentary disorder characterized by a constellation of findings

List 7 characteristics.

Lateral displacement of the medial canthi with dystopia canthorum, broad nasal root, heterochromic irises, congenital deafness, a white forelock, and cutaneous hypopigmentation

What is the usual inheritance pattern?

Autosomal dominant with variable penetrance and expression

PROTEUS SYNDROME

What is it?

A disturbance of ectodermal and mesodermal growth of unknown etiology

List 7 clinical manifestations.

Asymmetric overgrowth of the extremities, verrucous skin lesions, angiomas, lipomas, bone thickening, macrocephaly, excessive muscle growth

CUTANEOUS MANIFESTATIONS OF SYSTEMIC DISEASE

Describe the characteristic skin findings in:

Cushing disease?

Facial plethora, skin atrophy, acne, striae, excess hair growth

Dermatomyositis?

Periorbital heliotrope rash, shawl rash, Gottron's papules, periungual telangiectasias

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Henoch-Schönlein purpura?	Palpable purpura, predominantly on the buttocks and lower extremities
Hyperthyroidism?	Warm moist skin, hyperpigmentation, periorbital swelling with proptosis, facial flushing, soft loose nails, pretibial myxedema
Hypothyroidism?	Coarse dry skin and hair, brittle nails, myxedema
Infective endocarditis?	Janeway lesions (nontender erythematous lesions on the palms and soles, thought to be due to septic emboli), Osler's nodes (tender palpable erythematous nodules on the palms and soles, thought to be due to local immunologic inflammation), splinter hemorrhages
Inflammatory bowel disease?	Erythema nodosum (painful erythematous nodules, typically on the anterior lower extremities), pyoderma gangrenosum (cutaneous ulceration with violaceous borders)
Insulin resistance?	Acanthosis nigricans (hyperpigmentation and velvety thickening of intertriginous areas, particularly the back of the neck and axial)
Lyme disease?	Erythema migrans (erythematous macule or papule that expands to form large, erythematous annular lesions with central clearing)
Meningococcemia?	Petechiae and purpura on the trunk and lower extremities
Rheumatic fever?	Erythema marginatum (nonpruritic, evanescent, erythematous serpiginous lesions on the trunk and extremities)

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Rocky Mountain spotted fever?	Erythematous macules and petechiae that begin on the wrists and ankles that spread to the trunk, palms, and soles
Still disease (systemic juvenile idiopathic arthritis)?	Diffuse evanescent salmon-colored macules on the trunk and upper extremities
Systemic lupus erythematosus?	Butterfly malar facial rash, discoid scaly erythematous macules, psoriasiform lesions in sun-exposed areas

NAIL DISORDERS

HANGNAIL

What is it?	Growth of the nail material along the lateral aspect of the nail where it joins into the skin. It is commonly deep seated and tends to curl away from the normal nail. Discomfort occurs when this area is rubbed or caught on clothes or fabric.
How is it treated?	By pulling it out. In some cases, freezing or providing a local anesthetic beforehand can help alleviate discomfort from the procedure.

INGROWN TOENAIL

What is it?	The side of the toenail burrows into adjoining skin, resulting in swelling, granulation tissue, erythema, and sometimes infection
Which toe is usually affected?	The large toe
List 3 components of treatment.	1. Soak toe in warm, soapy water to clean and provide symptomatic relief 2. Antibiotics (if infected) 3. Removal of one-third to one-half of the toenail on the affected side is ultimately needed. Removal of the entire nail with disruption of the matrix of the nail bed will keep the nail from regrowing (if desired by the patient).

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How is recurrence prevented?

If the nail regrows, recurrence is common. Chances of ingrowth may be reduced by cutting nails straight across and by teasing the skin away from the nail with a cotton swab as the nail regrows.

SUBUNGUAL HEMATOMA

What is it?

Blood clot collected under the nail bed secondary to trauma. It appears as a bluish collection under the nail and can be very painful.

What is the treatment?

Small painless subungual hematomas do not require treatment. Larger, more painful lesions can be evacuated by melting the nail over the hematoma using electric cautery or by heating the end of a paper clip.

What condition may exist if a lesion resembling a hematoma exists without a history of trauma?

Melanoma

PARONYCHIA

What is it?

An area of inflammation or abscess formation involving the folds of tissue at the base (or at the lateral base) of the fingernail

What are the 3 treatments?

1. Often, warm soaks induce drainage, leading to resolution
2. Antibiotics for the surrounding cellulitis
3. Occasionally, a small incision is needed for drainage

FELON

What is it?

Infected collection (essentially a small abscess) in pulp of distal finger pad

List 4 signs and symptoms.

Swelling of finger pad with tenseness and sometimes erythema, extreme tenderness to touch

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What is the treatment?

1. Surgical drainage with incision in the midportion of finger pad in the direction of finger

2. Antibiotics may be needed for cellulitis

Note: Incisions in lateral aspects of finger are to be avoided despite description of this in older sources. These incisions may damage digital nerves and vessels.

HAIR DISORDERS

ALOPECIA

What is alopecia?

Partial or complete hair loss (distinguished from hypotrichosis, which is deficient hair growth)

What are the causes of true alopecia?

Inflammatory dermatoses, mechanical trauma, drugs, infection, endocrine disorders, nutritional imbalance, disturbance of the hair

List 5 causes of alopecia in children.

Alopecia areata, traction alopecia, telogen effluvium, trichotillomania, tinea capitis

What is the natural history?

Usually alopecia will resolve when the underlying cause is treated. It is rarely primary or congenital.

What is alopecia areata?

Focal hair loss, believed to be immune mediated

What is traction alopecia?

Localized hair loss due to excessive tensional forces on the hair follicle (e.g., certain hairstyles and processes)

What is telogen effluvium?

Diffuse hair loss due to disruption of the hair growth cycle, usually triggered by metabolic or hormonal stresses or medications

What is trichotillomania?

Irregular areas of incomplete hair loss as a result of compulsive pulling, twisting, or breaking of the hair

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HYPERTRICHOSIS

What is it?	Excessive hair growth in inappropriate areas. It may be localized, generalized, permanent, or transient. The pattern of hair growth is not in a sexual distribution.
What are the etiologic factors?	It may have racial or familial forms. It is also associated with a variety of conditions, including local trauma, malnutrition, anorexia nervosa, chronic inflammatory dermatoses, hamartomas or nevi, endocrine disorders (e.g., hypercortisolism), and congenital and genetic disorders (e.g., Cornelia de Lange syndrome), as well as a wide variety of drugs, including phenytoin, steroids, cyclosporin, minoxidil, and streptomycin.

HIRSUTISM

What is it?	Excessive hair growth in appropriate areas (i.e., in a sexual pattern)
List 7 conditions or factors that are common causes.	Hyperprolactinemia; gonadal tumors (Ch 22, p. 347); endocrine insensitivity; adrenal conditions (e.g., enzyme deficiencies, neoplasms, Cushing syndrome); drugs (e.g., minoxidil, phenytoin, cyclosporin, steroids, oral contraceptives); congenital anomalies or syndromes (e.g., trisomy 18 [Ch 30, p. 514], Cornelia de Lange syndrome, Hurler syndrome, juvenile hypothyroidism [Ch 24, p. 386]); true precocious puberty (Ch 24, p. 375)
What is the treatment?	Treatment of the underlying cause

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Chapter 28 Infectious Diseases

MENINGITIS

What is it?	Inflammation of the meninges
What are the common clinical findings?	Fever, headache, stiff neck, changes in mental status, CSF leukocytosis
What are the 2 major classes of meningitis?	Bacterial and aseptic (usually viral)

BACTERIAL MENINGITIS

(See Ch 10, p. 102, for a discussion of neonatal bacterial meningitis.)

How does a patient with bacterial meningitis present?	Presentation is highly variable and age dependent.
Neonates and infants?	Nonspecific signs of serious illness include **lethargy, irritability, poor feeding, tachypnea, jaundice, hypoglycemia,** and **vomiting.** Child may be febrile, afebrile, or hypothermic. Later signs: **bulging fontanel, seizures,** and **poor muscle tone**
Older children?	May have more classic meningeal signs, including **Kernig** or **Brudzinski signs** (or both), **headache, photophobia, vomiting, mental status changes** (e.g., lethargy, disorientation). **Petechiae** and **purpura** may be signs of a poor prognosis.
What are the 3 most common causative organisms from birth to 1 month of age?	Group B streptococcus, *Escherichia coli, Listeria monocytogenes*

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1–3 months of age? (list 3)

Streptococcus pneumoniae, group B streptococcus, *Haemophilus influenzae* type b

3 months to 3 years of age?

S. pneumoniae, Neisseria meningitidis, H. influenzae type b

Children older than 3 years? (list 3)

N. meningitidis, S. pneumoniae, H. influenzae type b

How is the diagnosis of bacterial meningitis made?

Lumbar puncture (see Ch 2, p. 11)

List 5 tests that should be conducted on the CSF.

Gram stain, culture, cell count, glucose, and protein (see Ch 2, p. 12, for normal values of RBC, WBC, and protein concentration. Gram stain and CSF culture should show nothing normally. Glucose should reflect serum levels.)

List 5 CSF findings that suggest bacterial meningitis.

1. Gram stain: may show bacteria
2. Culture: will reveal specific organisms
3. Cell count: CSF leukocytosis (usually $>1{,}000/mm^3$) with **predominance of polymorphonuclear neutrophil leukocytes (PMNs).** Note, however, that the presence of any PMNs in CSF is abnormal.
4. Glucose: relative **hypoglycemia** (<60–70% of serum glucose)
5. Protein: **elevated total protein**

List 3 issues that may cause difficulties in interpreting CSF findings.

1. "Bloody" spinal taps may confound both protein and WBC levels.
2. Previous treatment with antibiotics (e.g., amoxicillin for otitis) renders culture results inaccurate and may decrease WBC count.
3. Viral meningitis can have CSF profile similar to that of bacterial meningitis early in its course. A second LP may be necessary.

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List 3 other findings that are suggestive of meningitis.

1. **High peripheral WBC** with left shift (Caution: WBC may be low.)
2. **Thrombocytopenia** with **decrease in hematocrit** (Hct) is suggestive of disseminated intravascular coagulation (see Ch 15, p. 185).
3. **Blood cultures** may be positive.

What should precede LP if high ICP is suspected?

If elevated ICP is suspected (papilledema, focal neurologic signs), CT scan should precede LP. **Do not delay treatment in a seriously ill patient.**

What is the common treatment for bacterial meningitis?

Birth to 4 weeks of age? Ampicillin and gentamicin

1–3 months of age? Ampicillin and third-generation cephalosporin, consider vancomycin if suspicious for *S. pneumoniae* meningitis

3 months of age or older? Third-generation cephalosporin plus vancomycin (**Note:** Determination of antibiotic sensitivities is essential. Treatment before culture should cover likely organisms in the patient's age group and clinical setting.)

What is the duration of treatment?

It depends on the patient's age, the causative organism, and the patient's response to treatment. General guidelines:

H. influenzae type b and *S. pneumoniae:* 10–14 days

N. meningitidis: 7 days

Group B streptococcus: 14 days

E. coli: 21 days after negative CSF culture

For meningitis caused by gram-negative organisms, an LP is often recommended at the end of treatment to determine that the CSF is free of the organisms.

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What are some other components in the management of meningitis?

1. **Fluid restriction** to two-thirds maintenance (when intravascular volume is restored) may help prevent cerebral edema.
2. **Monitor head circumference** in infants.
3. Close monitoring of **glucose, acid-base,** and **volume status** and **tissue oxygenation** is essential.

Are steroids indicated?

Steroids may decrease hearing loss in *H. influenzae* meningitis and morbidity with *S. pneumoniae*. Use varies among institutions. ONLY shown useful if given prior to initial antibiotic administration

List 16 complications of meningitis.

Syndrome of inappropriate anti-diuretic hormone secretion, cerebral edema, toxic encephalopathy, brainstem herniation, cranial nerve palsies, deafness, seizures, subdural effusion, cerebral infarct, cortical vein thrombosis, disseminated intravascular coagulation, paresis, mental retardation, hydrocephalus, visual impairment, mental and motor delays

In what 2 groups of patients are complications most common?

1. **Newborns with gram-negative infection**
2. **Patients with pneumococcal** meningitis (up to 50% of patients experience complications)

What is the most common complication?

Sensorineural hearing loss (up to 20% of patients with *H. influenzae* meningitis)

What 2 bacteria cause the highest mortality rate?

S. pneumoniae and *N. meningitidis*

ASEPTIC MENINGITIS

What are the signs and symptoms of aseptic meningitis?

Similar to those of bacterial meningitis: **headache, vomiting, stiff neck, photophobia, fever, malaise, myalgia, gastrointestinal (GI) symptoms, rash, tachypnea**

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What is the clinical course?

Usually more indolent than in bacterial meningitis. Classic meningeal signs may be absent. Mental status is usually unaffected, unless meningoencephalitis or increased ICP has developed. **It is prudent to treat as if bacterial until culture results are available.** Some viruses (e.g., herpesvirus, rabies, arbovirus) also cause encephalitis and its accompanying complications.

List 4 findings of the LP.

1. **CSF pleocytosis** is the hallmark, but usually at levels lower than those in bacterial meningitis (i.e., WBC $> 100/mm^3$ and $< 1{,}000/mm^3$).
2. **CSF lymphocytosis,** as shown by the differential, may show a higher percentage of neutrophils early in the course.
3. Glucose levels are normal or slightly decreased.
4. Protein levels are normal or slightly elevated.

Other specific findings vary with etiologic factors.

List 9 viral causes.

Enteroviruses (e.g., coxsackievirus, echovirus) are the most common, especially in summer and early fall. Others include **Epstein-Barr, mumps, influenza, herpesvirus, and adenoviruses,** rarely **rabies** and **arboviruses,** and **poliovirus** in endemic areas or unimmunized populations.

How is it diagnosed?

Many viruses can be cultured from the CSF. Enteroviruses can be cultured from stool, and PCR testing for enterovirus in the CSF is available. Influenza, mumps, and adenovirus may be cultured from the nasopharynx. Serum titers (acute and convalescent) may be helpful. Herpesvirus can be difficult to verify. CT, MRI, and EEG may be useful.

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How is viral meningitis treated?

Primarily supportive. Dehydration and pain sometimes necessitate hospitalization. Acyclovir is used for herpes.

What is the clinical course?

Symptoms usually last 1–3 weeks. Headache may be severe.

List 6 categories, with examples, of nonviral causes of aseptic meningitis.

1. **Mycobacteria:** *Mycobacterium tuberculosis*
2. **Fungal:** *Cryptococcus neoformans* and *Coccidioides immitis* are most common (should be considered in immunocompromised patients).
3. **Rickettsia:** Rocky Mountain spotted fever, Q fever, typhus, and *Ehrlichia* (this should be considered when tick bite or farm animal exposure is in a child's history)
4. **Spirochetes:** leptospirosis, Lyme disease, syphilis
5. **Parasites** (very uncommon): *Naegleria fowleri* and *Acanthamoeba* (amebic meningitis); *Toxoplasma gondii*, *cysticercosis*, and *trichinosis*
6. **Drugs: IV immunoglobulin (IVIG), nonsteroidal anti-inflammatory drugs (NSAIDs), trimethoprim-sulfamethoxazole (TMP-SMZ), tacrolimus**

CONJUNCTIVITIS

What is it?

Inflammation of the conjunctiva

What are the typical signs and symptoms of infectious conjunctivitis?

Erythema (injection) of sclera or inner surface of eyelids, or both; increased tearing, discharge, or both; eyelids may stick together. Pain is uncommon, although child may complain of roughness or itching.

List 3 causes of conjunctivitis.

Infection (e.g., bacteria, viruses), allergy, chemicals

Do viruses or bacteria more commonly cause conjunctivitis?

Bacteria

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Which bacteria are the most common in young children?

Nontypeable *H. influenzae* and ***S. pneumoniae***

Moraxella catarrhalis, *Staphylococcus aureus*, and α-hemolytic streptococcus are possible pathogens but are also found in uninfected eyes.

List 2 infectious agents that particularly need to be ruled out in newborns.

Chlamydia trachomatis, *Neisseria gonorrhoeae*

Which is the most common virus isolated?

Adenovirus; herpesvirus and enteroviruses uncommonly cause conjunctivitis.

What other condition is often associated with conjunctivitis?

25–33% of patients have **otitis media;** 75% of these infections are bacterial.

How is it diagnosed?

Based on **culture;** in the newborn period, a rapid test is available for *N. gonorrhoeae* and *C. trachomatis.*

How is it treated?

It will usually resolve without treatment in 7–10 days. Infections with *N. gonorrhoeae* and *C. trachomatis* need to be treated aggressively with topical and systemic antibiotics.

Topical antibiotics include trimethoprim-sulfa-polymyxin B, erythromycin, bacitracin, gentamicin, and sulfacetamide. *S. pneumoniae* and *H. influenzae* are often resistant to aminoglycosides.

In what 3 instances are systemic antibiotics indicated?

1. When otitis media is simultaneously present (see Otitis Media, p. 469)
2. When the patient cannot tolerate topical therapy and treatment is indicated
3. When infection with *N. gonorrhoeae* or *C. trachomatis* is suspected or confirmed

What is EKC?

Epidemic keratoconjunctivitis

What causes EKC?

Adenovirus type 8; EKC **is very contagious.**

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List 3 symptoms.

It is associated with a **preauricular node** and presents with **eye pain** and **photophobia.**

List 4 signs and symptoms of allergic conjunctivitis.

Itching, redness, tearing, and photophobia—usually bilateral. Seasonal exacerbations and recurrent disease are common.

What is a characteristic physical finding?

Papillary hyperplasia with edema, leading to "cobblestoning" of conjunctiva

What are the 2 associated features?

1. There may be history or presence of other atopic disease.
2. Child may have angioedema of eyelids.

What is vernal conjunctivitis? (list 4 characteristics)

A **chronic** form of conjunctivitis, characterized by severe itching, photophobia, blurry vision, and lacrimation

What is a characteristic physical finding?

Large papillae on the upper eyelids

OTITIS MEDIA

What are the 3 types of otitis media?

1. Acute otitis media (AOM)
2. Otitis media with effusion (OME)
3. Chronic otitis media (COM)

ACUTE OTITIS MEDIA

What is it?

Infection of fluid in middle ear space

Why is otitis media more common in infants than in older children?

Anatomy: The eustachian tube drains fluid from the middle ear to the nasopharynx and protects against the reflux of nasopharyngeal pathogens. Infants have relatively horizontal eustachian tubes that become more vertical and widen as they grow.

Infections: Babies have frequent colds, causing obstruction of the eustachian tubes. Viruses also damage the ciliated epithelium of the tube. These factors inhibit the protective function of the eustachian tube.

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List 7 clinical signs of otitis media.

Prior or current upper respiratory infection (URI), fever, fussiness, sleeplessness, ear pain, decreased hearing, poor appetite

List 5 physical findings.

1. URI is often present.
2. Tympanic membrane (TM) is swollen, opaque, and discolored (red or yellow).
3. Normal TM landmarks are obscured.
4. Mobility of the TM is decreased or absent on pneumatic otoscopy or tympanogram.
5. Fluid may or may not be visible through the TM.

What finding is the most conclusive?

Obvious purulent effusion

List the 4 most common bacterial pathogens.

S. pneumoniae is the most common (30–40% of cases), followed by nontypeable ***H. influenzae*, *M. catarrhalis*,** and ***Streptococcus pyogenes*.**

S. aureus is an uncommon cause.

Viral pathogens? (list 4)

Believed to cause up to 30% of AOM cases; include RSV, influenza, adenovirus, and coxsackievirus

List 2 reasons to treat otitis media.

Treatment is thought to:
1. Prevent rare complications, such as mastoiditis, meningitis, and cholesteatoma
2. May accelerate the time to improvement of the child

List 5 considerations in choosing whether to treat with antibiotics.

1. **Etiology:** Many infections are minor or viral and resolve without therapy. In many countries, AOM is rarely treated with antibiotics.
2. **Organism resistance:** Nearly 100% of *M. catarrhalis*, 20% of *H. influenzae*, and 25% of *S. pneumonia* cases are β-lactamase-producing and resistant to penicillins.

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3. **Compliance:** It is not easy to give medicine to a baby. Dosing 3 or more times a day requires medication to be given at day care or at school.
4. **Cost:** New cephalosporins are effective but expensive.
5. **Resistance:** Third-generation cephalosporins have an unnecessarily wide spectrum and contribute to emerging drug resistance.

What are the 2 usual choices of antibiotics?

Amoxicillin or amoxicillin with clavulanic acid (Augmentin)

List 3 reasons to change antibiotics.

1. **Treatment failure**—no improvement in 2–3 days on initial antibiotic with good compliance
2. **Recurrence**—another episode of AOM within 6 weeks
3. **Side effects**—diarrhea, GI upset, allergic reactions

What are some second-choice antibiotics?

1. Amoxicillin (40 mg/kg per day) *plus* amoxicillin/clavulanic acid
2. Cefuroxime, cefpodoxime
3. Erythromycin with sulfa (Pediazole)

What is recurrent otitis media?

Three or more episodes of AOM in 6 months, or 4 or more episodes in 12 months, with documented clinical resolution in between episodes

How can it be prevented?

Avoidance of pacifiers, bottle feeding, day care attendance, and smoking exposures

OTITIS MEDIA WITH EFFUSION

What is it?

Fluid (effusion) in the middle ear space without infection; it occurs alone, secondary to URI, or as a sequela of AOM.

List 4 possible symptoms.

OME is often asymptomatic, but can manifest as hearing loss, "plugged ears," or otalgia.

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What are the signs?	Fluid seen behind TM; decreased mobility of the TM
What is the clinical course?	Spontaneous resolution in majority of cases (>50% by 3 months and 75% by 6 months); more rapid resolution after AOM (90% by 3 months)
What are the complications?	1. OME predisposes the patient to AOM and subsequent COM. 2. Hearing impairment may cause modest delay in concurrent language development but overall language development is unchanged.
What is the treatment?	
Duration < 3 months?	Observation only; assess hearing if a condition lasts 3 months or longer.
Duration up to 6 months without hearing loss?	Antibiotics may cause more rapid resolution but are not necessary because of high rate of spontaneous resolution.
Duration of 3 months with bilateral hearing loss?	Antibiotics may be given, with close follow-up for resolution of effusion and restoration of hearing.
Duration > 3 months with bilateral hearing loss?	Myringotomy with tympanostomy tube placement may be indicated.
Are steroids beneficial?	Benefits have not been conclusively shown, and steroids may have adverse effects.
Do decongestants or antihistamines help?	Not usually
Is tonsillectomy or adenoidectomy helpful?	No

CHRONIC OTITIS MEDIA

What is it?	Inflammation of the middle ear, mastoid, or both, with otorrhea through the TM for >3 months
List 3 complications.	Mastoiditis, labyrinthitis, cholesteatoma

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What is a tympanocentesis?

Also called **myringotomy with aspiration,** it involves puncturing through the TM to collect and drain fluid from the middle ear space.

In which patients is it indicated?

Critically ill or immunocompromised patients, neonates, and patients in whom AOM is unresponsive to 2 or more full courses of antibiotics

What are tympanostomy tubes?

Also called pressure equalization tubes (**PE tubes**), these small plastic or metal tubes are surgically placed in the TM to drain and ventilate middle ear space.

List 4 indications.

1. Frequent, recurrent AOM
2. OME with bilateral hearing impairment (>3 months' duration)
3. Severe retraction or atelectasis of TM
4. Chronic suppurative complications

List 8 complications of tympanostomy tubes.

Tympanosclerosis or atrophy, dislocation of the tube into the middle ear, cholesteatoma, extrusion of the tube, chronic TM perforation, prolonged otorrhea, and complications of general anesthesia

List 6 risk factors for otitis media.

Passive smoke exposure, day care attendance, horizontal bottle feeding, anatomic defects of oral pharynx, being a twin, experiencing first episode of otitis at <2 months of age

THRUSH

What is it?

Overgrowth of *Candida albicans* in the oral cavity

Who gets it? (list 4 groups of children)

Infants, children on antibiotics, immunosuppressed children, and those with chronic systemic disorders

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What are the clinical features?

Soft, creamy white plaques on buccal mucosa, tongue, palate, and lip commissures; lesions do not scrape off easily and leave an ulcerated red base when removed.

What are the treatments?

For infants, nystatin suspension. If it fails, gentian violet may be effective. For older children not at risk for aspiration, clotrimazole topical preparation for the mouth or systemic oral fluconazole is effective.

What are the 2 prevention and control measures?

For **formula-fed infants,** nipples and pacifiers should be consistently sterilized by boiling them for 5 minutes or placing them in a sterilizer; this prevents reinfection.

In **breast-fed infants,** the mother's nipples can be a source of infection. The mother should be asked about sore, red, cracked nipples; she may need concurrent treatment as well.

PHARYNGITIS OR STREPTOCOCCAL PHARYNGITIS

What is pharyngitis?

Sore throat (inflammation of the pharynx)

What is the most common cause?

90% of cases are **viral.**

What is streptococcal pharyngitis?

Pharyngitis caused by group A β-hemolytic streptococcus

List 2 complications of streptococcal pharyngitis.

1. Acute rheumatic fever
2. Local complications (e.g., peritonsillar abscess)

List 2 ways the diagnosis is made.

Throat culture (best) or rapid antigen tests

Can streptococcal pharyngitis be diagnosed purely on clinical grounds?

Not consistently. Clinical features overlap with the more common viral causes.

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List 2 ways streptococcal pharyngitis is treated.

1. Treatment of choice is penicillin, for 10 days; it may be oral or injectable (1 dose IM).
2. Erythromycin for patients allergic to penicillin

GINGIVOSTOMATITIS

What is it?

Inflammation of the gingiva and oral mucosa

What is the usual cause?

A primary infection with **HSV type 1.** HSV type 2 is a less common cause.

At what age is it commonly seen?

6 months to 3 years of age

What are the typical signs and symptoms?

A prodrome of headache, fever, malaise, and local lymphadenopathy, followed by erythema, swelling, and pain of gingiva and palatal mucosa. Grouped vesicles and ulcerations occur on oral mucosa. Bleeding and crusting may occur. It most frequently involves anterior gingiva and palate. Dehydration can follow when pain prevents adequate fluid intake. Secondary infection with a second organism, often bacterial, may occur.

List 3 ways the diagnosis may be made.

1. It is **usually made clinically.**
2. A **Tzanck prep** of the base of the oral lesions may show multinucleated giant cells and intranuclear inclusions.
3. Fluid from vesicles may be cultured to confirm HSV.

How can HSV gingivostomatitis be differentiated from hand-foot-mouth disease or aphthous stomatitis?

1. Hand-foot-mouth disease (**coxsackievirus**) lesions typically involve the **posterior palate and pharynx.**
2. **Aphthous stomatitis** lesions are found on **buccal, lingual,** and **inner lip mucosa.**

What is the clinical course?

Lesions heal spontaneously in 1–2 weeks.

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Can it recur?

Reactivation of HSV, leading to recurrent infections, is common. Recurrent infections tend to be less severe with fewer and more localized lesions.

What 3 types of treatments are helpful?

1. **Pain control:** Either acetaminophen, ibuprofen, or both are usually sufficient. Diphenhydramine-antacid (Benadryl-Maalox) suspension (1:1 mix) may provide local pain relief. Viscous lidocaine can be added for children older than 6 years.
2. **Oral hygiene:** Rinsing with chlorhexidine or glycerin-peroxide mix (for younger children) should replace tooth brushing, which may be too painful.
3. **Hydration:** Cold liquids are best tolerated. Gelatin, flavored ice-pops, and ice cream are useful. Citrus and carbonated beverages are painful to drink. Occasionally, IV fluids are needed.

When is antiviral therapy indicated?

Systemic acyclovir is indicated **only in immunocompromised patients.**

LYMPHADENITIS

What is lymphadenitis (also called "adenitis")?

Swelling and inflammation of lymph nodes. It may or may not be painful depending on the etiologic factors.

List 8 common causes of lymphadenitis in children.

Reactive lymph node, staphylococcal infection, atypical mycobacterium, cat-scratch disease, mononucleosis, toxoplasmosis, brucellosis, tularemia

How is it managed?

Usually by treating the primary disease process

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GASTROENTERITIS

List 5 signs and symptoms of acute gastroenteritis.

Signs are variable, but may include nausea, vomiting, diarrhea, abdominal pain, and excess flatulence.

What causes most cases of acute gastroenteritis?

Most cases in the United States are viral.

VIRAL GASTROENTERITIS

List the 3 viruses that are most common in children.

Rotavirus, adenovirus, "Norwalk" virus

List 2 ways the viruses are spread.

Fecal-oral route and respiratory route. Good hygiene and hand washing help reduce the risk of infection.

List 3 ways viral gastroenteritis is diagnosed.

1. Usually is a clinical diagnosis
2. Detection of viral antigens in stool
3. Viral culture of stool

How is viral gastroenteritis treated?

Usually supportive; **prevent dehydration** with IV or oral fluid and electrolyte management, depending on the severity

Do most children with acute viral gastroenteritis need IV fluids?

No

What may distinguish viral from bacterial gastroenteritis?

Bacteria more commonly cause bloody diarrhea, stool leukocytes, and tenesmus. Children with **viral** gastroenteritis also may have non-GI symptoms such as cough, nasal discharge, and myalgia.

BACTERIAL GASTROENTERITIS

Name 8 bacteria that cause acute gastroenteritis.

Salmonella, *Shigella*, *Campylobacter jejuni*, *E. coli*, *Yersinia enterocolitica*, *Clostridium difficile*, *Clostridium perfringens*, and *S. aureus* (toxin)

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List 4 ways acute bacterial gastroenteritis is diagnosed.

1. Stool culture (*Salmonella*, *Shigella*, *Campylobacter*, *E. coli*, *Yersinia*, *C. difficile*, *C. perfringens*)
2. Serologic testing (*Yersinia*)
3. Toxin assay (*C. difficile*)
4. Toxin assay in food (*S. aureus*)

How is it treated?

Supportive treatment (IV or orally) for fluid and electrolyte loss. Antibiotic therapy is usually not indicated because illnesses are often self-limited.

Exception: *Shigella* is usually treated because of public health concerns. Extraintestinal infections (including sepsis) are indications for antibiotic treatment. Infants may be given antibiotics more readily than older children.

Why not treat the usual *Salmonella* infection?

It is usually self-limited. Treatment may prolong the "carrier state" and may not significantly alter the clinical course.

When should *Salmonella* infection be treated?

Patient is an infant (younger than 4 months), has an immune deficiency, has a systemic disease (e.g., sepsis, osteomyelitis), or has typhoid fever.

What oral rehydration regimen is recommended for gastroenteritis?

The World Health Organization (WHO) oral rehydration solution, which includes:

Glucose: 90 mmol/L

Sodium: 80 mmol/L

Potassium: 20 mmol/L

Chloride: 80 mmol/L

Base (citrate): 30 mmol/L

Final total osmolarity: 300 mOsm/L

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List 4 commercially available oral fluids that are useful.

Several are useful, including Rehydralyte, Pedialyte, Ricelyte, and Infalyte.

List 6 indications for IV fluid.

Inability to drink liquids, severe vomiting, significant decrease in urination, significant tachycardia, shock or impending shock, coma

Which IV fluid should be used?

It depends on the situation. Lactated Ringer's solution or normal saline for volume expansion is used in severely dehydrated patients, followed by calculated replacement of electrolytes and maintenance fluids. (See Ch 3, p. 17, for electrolyte concentrations of GI fluids; see Table 3–1.)

Should one feed a child with acute gastroenteritis?

In general, yes, but judiciously. In most children, careful feeding may promote healing and help prevent malnutrition.

URINARY TRACT INFECTION

What does the term UTI refer to?

Infection of the urethra and bladder; the term grossly encompasses ascending infections up to the kidney as well.

Are female or male children more likely to have UTIs?

In infants, males and females are affected equally. Uncircumcised male infants may have a slightly higher risk. In older children, females are affected more commonly.

What are the most common etiologic agents?

E. coli accounts for 70–90% of infections. Other agents include *Klebsiella*, *Proteus*, and enterobacteria species.

What is the pathophysiology in infants versus children?

Infection in infants usually results from either bacteremia or migration of bacteria from the urethra. In older children, UTIs more commonly occur because of ascending bacteria from the lower urinary tract.

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What is acute bacterial cystitis?

Infection of the bladder itself

List 2 characteristics.

1. Hyperactivity of the detrusor muscle
2. Decreased functional capacity of the bladder

What are the potential symptoms of a UTI?

In infants? (list 8 symptoms)

Fever, weight loss, fussiness, failure to thrive, nausea, vomiting, diarrhea, and jaundice

In older children? (list 8 symptoms)

Fever, urinary frequency, pain during urination, incontinence, bed-wetting, abdominal pain, foul-smelling urine, and hematuria

How is urine obtained for analysis and culture?

In infants and small children? (list 2 methods)

Catheterization or **suprapubic puncture** (see Ch 2, p. 13, for a description of the technique for suprapubic puncture)

In older children?

A true "midstream" urine specimen

List 4 findings that suggest infection.

1. Pyuria; however, infection can occur in the absence of pyuria. Conversely, pyuria can be present without infection.
2. Microscopic hematuria
3. Presence of nitrites on urinalysis
4. A urine culture result of 100,000 colony-forming units/mL is diagnostic of infection. Occasionally, infection may be present with a slightly lower colony-forming unit count of a single organism.

How are UTIs treated?

Antibiotics of choice: Oral: TMP-SMZ, nitrofurantoin, or cefixime. If symptoms are not severe, it is preferable to wait for culture results before starting antibiotics. However, if symptoms are bothersome, antibiotic therapy should be started **after** the specimen is collected.

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If UTI is diagnosed, what 2 further workups are required?

1. A renal ultrasound for young children and for girls with recurrent UTIs. Consideration of a VCUG to assess for reflux is indicated for young children and for girls with recurrent UTIs. Reflux may predispose to ascending infection and pyelonephritis. If VUR is present, antibiotic prophylaxis may be needed for as long as the reflux persists or until surgical correction is undertaken. (See Ch 21, p. 335, for a discussion of VUR.)
2. A follow-up urine culture should be obtained after treatment to confirm that the UTI is cleared.

PYELONEPHRITIS

What is it?

An infection of the renal parenchyma that is usually caused by ascending infection from the lower urinary tract

What 2 conditions predispose a child to pyelonephritis?

Recurrent UTI and VUR

What are the clinical manifestations?

In infants?

Signs typical of systemic infection, including fever, weight loss, failure to thrive, and irritability

In older children?

Fever, chills, and flank or abdominal pain are typical.

List 3 laboratory tests that are valuable for diagnosis.

1. Urine culture
2. Urinalysis showing WBC casts, positive leukocyte esterase and/or positive nitrites
3. Peripheral WBC count may be elevated.

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What imaging studies may be helpful?

1. **Ultrasound** may show hydronephrosis, a perirenal abscess, or pyonephrosis. The latter 2 conditions may require prompt drainage of purulence.
2. Renal **scanning** may confirm pyelonephritis by revealing filling defects in the renal parenchyma.

What is the treatment?

IV antibiotics

What are the potential complications of pyelonephritis?

Arterial hypertension, renal insufficiency, or both, secondary to chronic renal damage

List the 2 components of treatment for renal or perirenal abscesses or infections associated with obstructed urinary tract.

1. IV antibiotic therapy
2. Drainage of the infected and obstructed areas using image-guided (ultrasound or CT) or surgical technique

DACTYLITIS

What is it?

An infection of the volar fat pad of the distal portion of the finger or thumb. It is usually blistering in nature.

What are the etiologic factors?

Usually, this condition occurs spontaneously. In rare cases, dactylitis may be the first manifestation of **sickle cell disease** in an infant.

List the 3 most common bacteria that cause it.

Group A β-hemolytic streptococcus, *S. aureus*, group B streptococcus

What are the 2 components of treatment?

1. Incision and drainage of the blistering lesion
2. Penicillin, clindamycin, or erythromycin therapy

SEXUALLY TRANSMITTED DISEASES (STD)

Which age group has the highest rate of STDs?

Adolescents

(Also see Ch 14, p. 159)

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List 8 common STDs.

1. Gonorrhea (*N. gonorrhoeae*)
2. Syphilis (*Treponema pallidum*)
3. Chlamydia (*C. trachomatis*)
4. Chancroid (*Haemophilus ducreyi*)
5. Genital herpes
6. Human papillomavirus (HPV)
7. Trichomoniasis (*Trichomonas vaginalis*)
8. *Gardnerella* infection (*Gardnerella vaginalis*)

If organisms that are usually sexually transmitted are found in younger children, what should be suspected?

Child sexual abuse

GONORRHEA

What is the causative pathogen in gonorrhea?

N. gonorrhoeae

What are the typical symptoms?

In males?

A purulent discharge that causes burning on urination (dysuria); occasionally infected males are asymptomatic.

In females? (list 3 symptoms)

Purulent vaginal discharge, vulvar vaginitis, dysuria; some females are asymptomatic.

What is the most common complication?

Pelvic inflammatory disease (PID) (p. 489)

How is the diagnosis made?

Culture of urethral or cervical discharge

What is the treatment?

Ceftriaxone

What is the duration of treatment?

Depends on whether the disease is localized or whether complications, such as PID or disseminated disease, have occurred

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SYPHILIS

What is the causative pathogen in syphilis?

T. pallidum

What are the symptoms of syphilis?

Primary syphilis?

A painless chancre appears at the site of inoculation approximately 2–6 weeks after the infection. There may be associated **adenitis.** The chancre heals spontaneously within 4–6 weeks.

Secondary syphilis?

2–10 weeks after the chancre heals, a nonpruritic maculopapular **rash** occurs. Pustules may develop. **Condylomata** may occur around the anus and vagina. There may be an associated **flu-like illness** with lymphadenopathy. Thirty percent of people infected with secondary syphilis develop meningitis. After 1–2 months, the infection becomes latent but may recur up to the first year.

Tertiary syphilis?

This late stage manifests with **neurologic and cardiovascular involvement along with granulomatous lesions.**

What are the diagnostic tests?

1. Demonstration of *T. pallidum* on dark-field microscopy as direct immunofluorescence on specimens from skin lesions, placenta, or umbilicus
2. The Venereal Disease Research Laboratory (VDRL) and rapid plasma reagin (RPR) tests detect antibodies against a cardiolipin-cholesterol-lecithin complex. They are not specific for syphilis but do tend to correlate with disease activity and are therefore useful in screening.

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3. The fluorescent treponemal antibody absorption (FTA-ABS) test, the microhemagglutination assay for antibodies to *T. pallidum* (MHA-TP), and the *T. pallidum* immobilization (TPI) test are tests that measure antibodies specific to *T. pallidum.* They are usually used to confirm positive findings on nonspecific screening tests.

How is syphilis treated?

A single dose of **penicillin** is adequate for primary, secondary, and latent secondary disease. Treatment must be adjusted for congenital syphilis, tertiary disease, and neurosyphilis.

What is the likelihood of transmission of syphilis from an infected mother to her infant?

Virtually 100%

What is the fetal or perinatal death rate of infected infants?

40%

When should infants be treated for syphilis?

When there is evidence of the infection in the mother, or when mother had inadequate treatment

CHLAMYDIA

Which of the *Chlamydia* species is most commonly involved in sexually transmitted infection?

C. trachomatis

What are the clinical manifestations?

A urethritis that can be similar to that of gonorrhea and includes **burning** during urination as well as a urethral **discharge** (traditionally called "nongonococcal urethritis"). **Perihepatitis, conjunctivitis, PID, and subsequent sterility** are symptoms of chlamydia that has spread beyond its local site.

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List 5 ways the diagnosis may be made.

1. Isolation of the organism in tissue culture from the urethra in men and from the endocervix in women and girls
2. Fluorescent antibody tests
3. Enzyme-linked immunosorbent assay (ELISA)
4. DNA probe
5. Polymerase chain reaction (PCR) assay on a urine sample

How is *Chlamydia* infection treated?

Doxycycline, cefoxitin, erythromycin, and azithromycin may be adequate medications. For pregnant women, erythromycin or amoxicillin is recommended.

What symptoms may occur in infants of infected mothers?

Primarily conjunctivitis and secondary pneumonia

CHANCROID

What is chancroid?

A lesion characterized by a painful, purulent, sharply delineated ulcer. There is no induration and this helps to distinguish it from a syphilis chancre. Lymphadenopathy may be associated with this condition.

What is the treatment?

Ceftriaxone

GENITAL HERPES

List the 2 viral causes in genital herpes.

HSV type 2 and HSV type 1 (may cause 10–35% of cases of genital herpes)

What are the clinical manifestations?

In females?

The **vulva and vagina may have vesicles and ulcers. The cervix is the primary site of infection, and therefore, the disease may be subclinical.** However, virus may still be shed, thus infecting a partner. There may be associated pain along affected nerve roots in the perineal region.

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In males?	**Vesicles or ulcers occur on the penis.** The scrotum is less frequently involved. There may be associated pain along affected nerve roots in the perineal region.
What is the treatment?	There is no cure. Symptomatic and shedding phases may be shortened by the use of acyclovir or valacyclovir.
For what other disease are women with HSV at risk?	**Cancer of the cervix** and potential transmission to newborns through infected birth canal

HUMAN PAPILLOMAVIRUS (HPV)

What is it?	This term encompasses at least 75 different types of viruses that contain DNA.
What are the manifestations of sexually transmitted HPV?	Genital warts (also called "condylomata acuminata")
How is the diagnosis made?	Usually by **physical examination;** however, application of **3% acetic acid** to a lesion may show a characteristic whitening that provides diagnosis.
What are some potential treatments?	Primary prevention with 3-shot immunization recommended for all healthy adolescent girls aged 11–12 years, before initiation of intercourse. Topical treatments include trichloroacetic acid, liquid nitrogen, podophyllin, and topical 5-fluorouracil. Laser ablation may be required for lesions that do not respond to medical therapy.
What are the 3 complications of infection with HPV?	Cervical dysplasia, cervical cancer, development of respiratory papillomas, which, in infants, may become malignant or may spread to a point of being uncontrollable, resulting in airway obstruction and death

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TRICHOMONAS INFECTION (TRICHOMONIASIS)

What is the causative pathogen in this infection?	*T. vaginalis*
What are the clinical manifestations of *Trichomonas* infection?	
In females?	There is a frothy, malodorous, yellow **vaginal discharge,** which may be accompanied by **vulvar or vaginal irritation, dysuria,** and **dyspareunia.** Punctate cervical hemorrhages ("strawberry cervix") may be present.
In males?	Usually asymptomatic, but about 10% may experience urethritis.
How is the diagnosis made?	**Wet mount** examination of vaginal or urethral secretions will show the *Trichomonas.* This test is successful in about 70% of cases. Cultures may be needed to obtain a definitive diagnosis.
What is the treatment?	**Metronidazole—however, it should not be used in pregnant women. Clotrimazole** should be used in the first trimester of pregnancy if infection is suspected.

GARDNERELLA INFECTION

What is the principal manifestation of *Gardnerella* infection?	Usually a foul-smelling vaginal discharge
How is it diagnosed?	**10% potassium hydroxide** is added to a wet preparation of the discharge. This results in the emission of a fishy odor. **Clue cells,** which are epithelial cells ringed with the rod-shaped organism, are also evident on the "wet prep."
What is the treatment?	Metronidazole, which is contraindicated in pregnant women

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PELVIC INFLAMMATORY DISEASE (PID)

What is it?

A condition that may be caused by a variety of sexually transmitted organisms that have ascended through the vaginal tract into the cervix and uterus. Subsequent migration may occur toward the fallopian tubes.

List 2 common clinical manifestations.

Lower abdominal pain, which can be severe, and often fever

What are the 2 physical findings?

1. Extreme tenderness on motion of the uterus and adnexa (i.e., the **"chandelier sign"**)
2. A **purulent discharge** from the cervical region may be present.

What are the common conditions in the differential diagnosis?

Appendicitis, ovarian cyst, ovarian tumor, ovarian torsion, ectopic pregnancy, UTI, inflammatory bowel disease

How is PID treated?

IV antibiotics are usually necessary and should include coverage for *N. gonorrhoeae* and *Chlamydia* species. Consideration should be given to the treatment of anaerobes.

List 5 complications of PID.

Sterility, increased risk of ectopic pregnancy, chronic pain, dyspareunia, increased risk of recurring PID

COMMON VIRAL SYNDROMES

MEASLES

What is another name for measles?

Rubeola

List 6 signs and symptoms of measles.

Fever, cough, coryza, conjunctivitis, maculopapular rash, Koplik's spots (enanthem)

What are the 2 ways in which measles is spread?

Usually by direct contact with infectious secretions, but sometimes via airborne route

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What is the incubation period?	8–12 days from exposure to onset of symptoms, and 14 days from exposure to appearance of rash
List 5 complications of measles.	Pneumonia, croup, diarrhea, encephalitis, SSPE
What is SSPE?	Subacute sclerosing panencephalitis
When can SSPE occur?	Long after the illness; average incubation period is 10.8 years after the identified measles infection.
What is the treatment for measles?	Supportive; vitamin A may be useful.
Is isolation of the patient necessary?	Respiratory isolation for 4 days after the onset of rash; longer for immunocompromised patients

MUMPS

What is it?	It is a systemic viral disease, most notable for **swelling of the parotid salivary glands.** The mumps virus is a member of the **paramyxovirus** group, which includes **measles and parainfluenza.**
How is mumps spread?	Direct contact via respiratory exposure
What is the incubation period?	12–25 days, although it is usually 16–18 days
List 6 complications.	**Orchitis,** arthritis, pancreatitis, hearing loss, mastitis, renal involvement
Is isolation of the patient necessary?	Respiratory isolation for 9 days after the onset of parotid swelling
What is the treatment?	Supportive

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RUBELLA

What is another name for rubella?	German measles
List 2 clinical features.	1. Generalized lymphadenopathy (usually suboccipital, postauricular, cervical nodes) 2. A pink maculopapular erythematous rash appears first on the face then spreads downward.
How is rubella spread?	Direct or droplet contact with nasopharyngeal secretions
What is the incubation period?	14–21 days, but it is usually 16–18 days
Is isolation of the patient necessary?	Contact isolation for 7 days after onset of the rash
What is the treatment?	Supportive
What are the potential complications?	Polyarthralgia, arthritis, thrombocytopenia, encephalitis; **the major concern is congenital rubella.**
What is congenital rubella?	Rubella infection in a fetus, acquired as a consequence of maternal infection during pregnancy
List 4 ophthalmologic complications of congenital rubella.	Cataracts, microphthalmia, glaucoma, chorioretinitis
List 4 cardiac complications of congenital rubella.	PDA, peripheral pulmonic stenosis, ASD, VSD
What are the other complications?	Sensorineural deafness, microcephaly, mental retardation, growth retardation, thrombocytopenia, ecchymoses or purpura (sometimes called "blueberry muffin" baby)

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Is isolation of a child with congenital rubella necessary?	Contact isolation until 1 year of age, or until nasopharyngeal and urine viral cultures are consistently negative for rubella after 3 months of age

FIFTH DISEASE

What is another name for fifth disease?	Erythema infectiosum
What is its cause?	Parvovirus B19
List 4 clinical features.	Fever, systemic illness (usually mild), a "slapped cheek" rash on the face, and a lacy or reticular rash on the extremities
What are the potential complications?	Arthralgia, arthritis, bone marrow suppression; **it may cause hydrops fetalis in the fetus of a woman infected in the first half of pregnancy.**
How is it spread?	Respiratory secretions and blood
What is the incubation period?	28 days; 4–14 days on average
What is the treatment?	Supportive; immunoglobulin may be helpful in chronic infections in immunocompromised patients.

ROSEOLA

What is it?	A systemic viral infection, characterized by high fever for 3–7 days, followed by a maculopapular rash; it may include respiratory or GI signs.
List 2 complications.	Febrile seizures and rarely encephalitis
What causes roseola?	Human herpesvirus type 6 and type 7
Give 2 other names for roseola.	Exanthem subitum and sixth disease
How is it spread?	Unknown—likely respiratory secretions

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What is the incubation period?	5–15 days
Is isolation of an infected child necessary?	No
What is the treatment?	Supportive

VARICELLA

What is another name for varicella?	Chicken pox
List 2 clinical features.	Systemic illness with fever and generalized vesicular rash
What is the cause?	Varicella-zoster virus
List 4 potential complications.	Secondary bacterial infection of skin lesions, thrombocytopenia, arthritis, pneumonia
In what 3 ways is the varicella virus spread?	Direct contact, airborne spread, contact with zoster lesions
What is the incubation period?	10–21 days, but it is usually 14–16 days
When is a child infectious?	1–4 days before lesions erupt, and for 7–10 days afterward
What is the treatment?	Supportive and symptomatic; **salicylates should be avoided.** Antiviral agents (acyclovir) can modify the course of the disease if administered early.
List 2 ways chicken pox can be prevented.	1. Varicella vaccine is recommended for universal use. 2. Varicella-zoster immune globulin (VZIG), given after exposure, can modify the course of the disease or prevent it. It is used mainly with immunocompromised patients.

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What is zoster?	A painful vesicular eruption in a dermatomal distribution
What causes zoster?	Latent varicella virus that becomes active after primary systemic infection

HUMAN IMMUNODEFICIENCY VIRUS (HIV)

How do children acquire HIV infection?	The vast majority of children acquire it through vertical transmission from their infected mother.
List 3 other modes of transmission.	Contaminated blood products, sexual transmission, shared needles
Has screening of blood products affected the risk of HIV infection?	Yes—the risk from blood products has been greatly reduced, but not completely eliminated.
What proportion of children born to mothers with HIV infection will become infected?	About **20–30%** of babies born to infected, untreated mothers
When does vertical transmission of HIV infection to infants take place?	25% prepartum, 65% intrapartum, and 10% postpartum (via breast-feeding)
List the 4 major determinants of perinatal HIV transmission.	1. High maternal viral load 2. Lack of maternal antiretroviral therapy during pregnancy 3. Vaginal delivery 4. Breast-feeding
List 3 effective measures to prevent perinatal HIV transmission from an infected mother to her infant.	1. Provide aggressive antiretroviral therapy during pregnancy to reduce the maternal viral load. 2. Offer elective cesarean section at 38 weeks' gestation for mothers with elevated viral loads at the time of delivery. 3. Strongly discourage breast-feeding in United States.

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How effective are these measures in preventing perinatal HIV infection?

These interventions reduce the risk of perinatal transmission to <2%.

What is the reason that perinatal HIV infection in children has not been eliminated?

Failure to identify all pregnant women with HIV infection and offer them effective therapy. Prevention of perinatal HIV infection is predicated on universal testing of all pregnant women for HIV infection.

Can perinatal HIV infection be prevented if the mother does not receive testing and treatment during pregnancy?

Yes. Treatment of the mother during labor and delivery, and treatment of the infant shortly after birth will still decrease the risk of HIV transmission. This requires the use of rapid HIV tests for previously untested women and their babies.

List 2 components of treatment that should be given to babies born to HIV-infected women.

1. Beginning at 8–12 hours after birth, these infants should receive **oral zidovudine,** continuing until 6 weeks of age.
2. Beginning at 4–6 weeks of age, these infants should receive **TMP-SMZ prophylaxis** against *Pneumocystis jiroveci* pneumonia.

How is perinatally acquired HIV infection diagnosed?

HIV DNA PCR or highly sensitive HIV RNA detection performed at birth, 1–2 months of age, and again at 4–6 months of age. Positive tests should be confirmed by repeat testing before a definitive diagnosis is made. HIV infection is excluded if all three tests are negative.

Is testing for HIV antibody useful?

Only in children older than 2 years. Transplacentally acquired maternal antibody to HIV can persist in children for up to 18 months after birth. Thus, antibody is not useful to definitively diagnose or exclude HIV infection in young infants.

Are newborns with HIV infection clinically ill?

Not necessarily; most are well at birth.

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List 7 common clinical features of HIV infection.

Prolonged or unexplained fever, lymphadenopathy, hepatosplenomegaly, chronic diarrhea, poor weight gain, poor linear growth, parotitis

What are the potential infectious complications of HIV infection in children?

1. Increased susceptibility to infection with common bacterial and viral pathogens (otitis media, sinusitis, pneumonia, sepsis, and meningitis)
2. Increased susceptibility to infection with opportunistic organisms (*P. jiroveci*, *C. albicans*, *Mycobacterium avium*, *T. gondii*, and cytomegalovirus)

List 4 common organ-specific complications of HIV infection.

Encephalopathy, lymphoid interstitial pneumonitis, cardiomyopathy, nephropathy

What information does measuring CD4 counts provide?

CD4 counts provide a rough measure of the **extent of destruction of the immune system by HIV.** Severe depletion of CD4 cells renders the patient susceptible to opportunistic infections. **Exception: Susceptibility to *P. jiroveci* pneumonia in young infants does not correlate with CD4 counts. Most practitioners follow viral loads now rather than CD4 counts.**

How is pediatric HIV infection treated?

With a combination of antiretroviral agents. The usual treatment consists of 2 drugs that inhibit HIV reverse transcriptase combined with a drug that inhibits HIV protease.

What is the prognosis for pediatric HIV infection?

Before the availability of effective therapy, the prognosis was grim, with a median survival of about 8 years. Potent antiretroviral therapy has dramatically decreased morbidity and mortality. The full extent of the benefit of combination therapy is not yet known.

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TUBERCULOSIS

What are the 3 infecting agents for tuberculosis?	*M. tuberculosis*, *Mycobacterium bovis*, *Mycobacterium africanum*
Which infecting agent is the most prevalent?	*M. tuberculosis*
What percent of the world's population is infected with *M. tuberculosis*?	33%
How many people in the United States are infected with *M. tuberculosis*?	10–20 million
Which age group has the lowest rate of tuberculosis?	5–14 years of age
How is tuberculosis transmitted?	By mucous droplets that become airborne from person to person
Is it common for young children to infect others?	No, because often children do not have cough symptoms with tuberculosis, and if they do, the cough is not forceful enough to suspend infectious particles.
What is the primary portal of entry of tuberculosis?	The lung
What percentage of patients who are infected with tuberculosis develop clinical disease?	Approximately 5–10%; however, 40% of infected infants develop disease.
List 5 categories of children at highest risk for developing tuberculosis.	1. Children born in countries with high incidence of the disease 2. Poor and indigent children 3. Homeless children 4. Abusers of injected drugs 5. Children exposed to high-risk adults

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List 4 conditions that predispose children to become symptomatic with tuberculosis once they are infected.

1. Infection with HIV
2. Immunocompromising diseases, especially malignancy
3. Immunosuppressive drug treatments
4. Age of 3 years or younger

List 2 features of the "primary complex" of tuberculosis.

1. Local infection at the portal of entry, usually the lung
2. Subsequent infection of regional lymph nodes in that area

What is the Mantoux tuberculin skin test?

This is an intradermal injection containing purified protein derivative (PPD). Usually, 0.1 mL of solution containing 5 tuberculin units is used for initial testing.

What defines a positive test?

1. In children with a high risk of infection, an area of induration $\geq$ 5 mm is positive.
2. For other high-risk adults and children older than 4 years, an area of induration $\geq$ 10 mm is positive.
3. For low-risk persons, an area of induration $\geq$ 15 mm is positive.

List 5 factors that can cause a false-negative result on the tuberculin test.

Young age, malnutrition, immunosuppression, viral diseases (measles, mumps, varicella, influenza), overwhelming tuberculosis

What 2 factors can cause a false-positive response on the tuberculin test?

1. Cross-sensitization to nontuberculous mycobacteria
2. Exposure to the bacille Calmette-Guérin (BCG) vaccine

What are some clinical manifestations of tuberculosis?

Initially, there is a lung parenchymal focus with involvement of regional lymph nodes. This may result in bronchial obstruction in small children and infants. The clinical manifestations are fairly mild, however. Infants are the most prone to showing signs and symptoms, and these are usually nonproductive cough and mild dyspnea. Other generalized systemic symptoms such as fever, night sweats, anorexia, or decreased activity may occur.

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List 2 radiographic findings that may be present in children with pulmonary tuberculosis.

Collapse or consolidation of a lung segment as a result of bronchial obstruction, signs of bacterial pneumonia

List 3 other possible pulmonary manifestations.

Pleural effusion; extension of infection to the pericardium (pericarditis); upper respiratory tract disease

What other organs may tuberculosis affect?

Any organ

What are the characteristics of CNS involvement?

Meningitis with subsequent **caseous lesion** development. They affect the **brainstem** most commonly. Early symptoms resemble meningitis but may progress to coma, decerebrate posturing, and death.

CSF fluid usually shows 10–500 WBCs/mm^3, glucose level of 20–40 mg/dL, and elevated protein.

What is typical of bone involvement?

Bone involvement is usually in the **lower vertebrae.** Spondylitis (**Pott disease**) results in kyphosis.

What are the manifestations of abdominal disease?

1. Abdominal lymph nodes may become infected, causing localized peritonitis or even generalized peritonitis if caseous lymph nodes rupture.
2. In the intestine, ulcers may form, resulting in pain, diarrhea, or constipation.

What are the characteristics of genitourinary disease?

Early symptoms may be subtle. However, late symptoms may include dysuria, flank or abdominal pain, and gross hematuria. Superinfection may occur.

Hydronephrosis and urethral strictures may develop. In males, epididymitis or orchitis may occur.

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What are the symptoms of perinatal disease?

These usually occur after 2 or 3 weeks of life and include respiratory distress, fever, enlargement of the spleen or liver, poor feeding, lethargy, irritability, lymphadenopathy, abdominal distension, failure to thrive, ear drainage, and skin lesions. Chest radiograph may reveal a miliary pattern.

How is tuberculosis diagnosed?

By **isolation of the bacteria;** typically seen as **acid-fast bacteria** on staining with aryl methane. However, it may take 1–6 weeks to confirm growth in culture.

How is tuberculosis typically treated?

The most common drugs used are **isoniazid (INH)** and **rifampin.** Coverage with 2–4 drugs is necessary for patients with clinically active disease. This implies a large bacterial load and is meant to cover that population of bacteria that is resistant to a single drug. Patients with infection but no clinically active disease have a smaller bacterial load and may be treated with 1 drug, typically INH.

List 2 side effects of isoniazid.

Peripheral neuritis and **hepatotoxicity**

What is the side effect of rifampin?

Hepatotoxicity

What is the typical treatment strategy for tuberculosis?

For asymptomatic infection?

Asymptomatic infection is treated with a single drug (typically INH) for at least 9 months.

For active disease?

A typical **3-drug regimen, INH, rifampin,** and **pyrazinamide,** is prescribed for 2 months and then INH and rifampin for at least another 4 months.

In an area of multidrug-resistant tuberculosis, treatment must be based on susceptibility patterns of isolates, usually from adult contacts.

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List 3 instances in which children who do not exhibit tuberculosis disease should be treated.

1. Children with a positive PPD test
2. Children younger than 6 years who have been exposed to infected adults
3. Infants born to mothers who have tuberculosis

If exposed children are PPD negative 3 months after the treatment, the treatment may be discontinued.

What is the BCG vaccination?

It is a vaccine for Bacille Calmette-Guérin.

What is its use?

It is best used in infants and children to reduce the risk of disseminated tuberculosis. This is especially true for infants who are at high risk of exposure because of their living environment.

Has this vaccine resulted in overall decrease in tuberculosis?

No

PERTUSSIS

What is another name for pertussis?

Whooping cough, 100-day cough

What causes pertussis and how is it transmitted?

Bordetella pertussis; spread via respiratory secretions

What is the incubation period?

7–14 days

What are the 3 stages?

Catarrhal, paroxysmal, convalescent

What are characteristics of the cough?

A quick cough, such that the child may not be able to catch his or her breath until the end of the cough. The deep breath is the "whoop." The cough is most obvious in the catarrhal stage.

What are the complications of pertussis?

Pneumonia is seen, as well as CNS and GI complications, related to infection and to the consequences of violent coughing and pressure.

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List 4 ways it is diagnosed.

1. Clinical suspicion with possible presence of lymphocytosis
2. Positive culture for *B. pertussis*
3. Demonstration of the organism using a fluorescent antibody test of nasopharyngeal secretions
4. PCR identification of the organism in nasopharyngeal secretions

List 3 elements of treatment.

1. Hospitalization during period of severe coughing paroxysms; the child may need suction, supplemental oxygen, nutritional support, and respiratory support.
2. Antibiotic—usually erythromycin
3. Isolation until 5 days of antibiotic treatment are completed

What antibiotic prophylaxis is given, and for how long, for people exposed to pertussis?

14 days of erythromycin or 5 days of azithromycin

Does treatment of infected persons prevent the cough?

Probably not, but those treated early may have a shorter course

How is pertussis prevented?

Vaccination

What are the 2 possible complications of pertussis vaccination?

Some individuals have a febrile reaction, and there are reports of rare CNS complications. The relationship of these conditions to the vaccine is controversial.

Is protection by pertussis vaccination lifelong?

No

When are pertussis vaccines given?

In early childhood (DTaP) and then with diphtheria and tetanus booster (Tdap) every 10 years in adolescents and adults

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PARASITIC INFECTIONS AND INFESTATIONS

ROUNDWORM

What is the formal name for roundworm?	*Ascaris lumbricoides*
How big are the organisms?	Adults can be quite large (15–40 cm).
How is roundworm contracted?	Fecal-oral route (i.e., oral ingestion of eggs). The eggs usually hatch in the duodenum and the larvae penetrate the intestinal mucosa and migrate to the lungs and up the trachea, to be swallowed.
Where do the adults live?	Usually in the jejunum
What are the 2 categories of symptoms of ascariasis?	1. Possible **pulmonary symptoms,** including **Löffler's pneumonia** 2. **GI signs,** including abdominal pain, loss of appetite, nausea, and vomiting
What are the 2 possible serious complications?	Intestinal obstruction and aberrant migration (to liver, eyes, brain) with inflammatory responses
How is it usually diagnosed?	By finding the eggs or the worm in stool
What is the life span of *Ascaris*?	Usually 2 years
List 3 treatments.	Mebendazole, pyrantel pamoate, albendazole
How can it be prevented?	Good hand washing and sanitation

VISCERAL LARVA MIGRANS (TOXOCARIASIS)

What 2 agents cause visceral larva migrans?	*Toxocara canis*, *T. cati*
What are they?	Dog and cat roundworms (intestinal parasites)
How do humans become infected?	Ingestion of eggs (from animal feces, perhaps in dirt)

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What happens when humans ingest these eggs?

The eggs hatch in the intestines and migrate to organs (usually the liver).

What are the symptoms?

Symptoms may vary with the tissues involved. Fever, hepatomegaly, or other organ-specific findings may be present.

List 4 laboratory findings.

1. Eosinophilia, as a result of the tissue invasiveness of the parasite
2. Elevation of isohemagglutinin A and B antibodies
3. Elevated ESR
4. Positive ELISA

What is ocular larva migrans?

Eye involvement of visceral larva migrans

List 2 agents for treating visceral and ocular larva migrans.

Mebendazole or **albendazole** have been used. Dying organisms may cause an allergic or inflammatory response. **Ocular larva migrans** may also need **steroid** treatment, as antiparasitic treatment may not be solely effective for the inflammatory response.

PINWORMS

What is the formal name for pinworms?

Enterobius vermicularis

How is it transmitted?

Hand to mouth

What are the symptoms?

Perianal itching, sometimes leading to insomnia

How is it diagnosed?

Demonstration of pinworms or their eggs in the perianal region

What is the tape test?

Use of a clear adhesive tape to pick up eggs or worms from the perianal region; this pickup can be performed by the parents and the eggs or worms examined by the physician.

What is the treatment for pinworm infection?

Mebendazole

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List 3 ways it can be prevented.	Practice good hygiene. Cut nails. Wash sheets, underwear, and bedclothes daily for several days to prevent reinfection.

WHIPWORM

What is the formal name for whipworm?	*Trichuris trichiura*
How is it spread?	Fecal-oral route with ingestion of infective eggs
What are the potential symptoms?	The patient may be asymptomatic. Symptoms may range from abdominal pain and flatulence to rectal bleeding and prolapse, depending on the severity of the infestation.
How is it diagnosed?	Demonstration of characteristic eggs or worms in stool
What is the treatment?	Mebendazole
List 2 ways it can be prevented.	Good hygiene, sanitary disposal of human waste

HOOKWORM

What causes hookworm?	*Necator americanus* (in the United States) and *Ancylostoma duodenale*
What is the life cycle of the hookworm?	The larvae usually **burrow through the skin of the feet,** enter the bloodstream, and migrate to the lungs, where they ascend and are swallowed. They then reside in the intestines.
List 4 complications.	Irritation at the site of skin entry (**"ground itch"**), anemia, hypoproteinemia, nutritional deficiency
How is it diagnosed?	By finding ova in stools
List 2 treatments.	Mebendazole, pyrantel pamoate
List 2 ways it can be prevented.	By wearing shoes and improving sanitation

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ATYPICAL MYCOBACTERIA

What are atypical mycobacteria?	Mycobacteria that are nontuberculous
How are they generally acquired?	From the environment, as opposed to person-to-person spread
List 2 categories of atypical mycobacteria.	"Rapid-growing" mycobacteria (grown within 3–7 days) include *M. fortuitum*, *M. chelonae*, and *M. abscessus*, whereas "slow-growing" mycobacteria often require weeks before sufficient growth occurs, including *M. tuberculosis* and other nontuberculous mycobacteria.
What are the most typical infectious manifestations in children?	**Cervical lymphadenitis;** however, children with AIDS are commonly infected systemically with *M. avium* complex.
Can nontuberculous mycobacteria infect other regions of the body?	Yes—particularly the skin, lungs, bones, and joints
Which mycobacterium accounts for most cases of cervical lymphadenitis?	***M. avium*** is responsible in 80% of cases. Most other cases are caused by either ***M. intracellulare*, *M. fortuitum*,** or ***M. kansasii*.**
List 2 typical symptoms of cervical lymphadenitis.	1. Enlargement of an isolated lymph node or group of nodes 2. With progressive disease, caseation may occur, resulting in drainage to the skin.
How is the definitive diagnosis made?	By isolation of the organism from a tissue sample
What is the treatment?	Surgical excision of the involved nodes
Can HIV-positive children who are infected with disseminated *M. avium* be cured of this bacterial infection?	Usually not, but multiple-drug therapy may diminish the effects of the disease.

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Chapter 29 Allergic Diseases

ATOPY

What is it?	A genetic tendency to produce IgE antibodies in response to common environmental proteins. This can result in symptoms of hay fever, asthma, or eczematoid dermatitis, or in allergic reactions to food, drugs, and insect bites.
What is the pathophysiologic process in an atopic individual?	Selected synthesis of IgE antibodies to common environmental antigens
How is this process demonstrated in atopic individuals?	A "wheal-and-flare" reaction occurs when their skin is tested with allergenic extracts or elevated allergen-specific IgE antibodies in their serum.
Can nonatopic individuals form IgE antibodies?	Yes—but not to common environmental substances in the same manner as atopic individuals
What is the definition of allergy?	A hypersensitivity reaction mediated by an immunologic mechanism, causing an undesired physiologic response Individuals can have allergic reactions to foods or drugs. However, a true allergy should be differentiated from an adverse reaction (commonly nausea or vomiting) that does not have a true immunologic basis. Adverse reactions without an immunologic basis are food or drug intolerances.

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Are antigens and allergens the same?

Not necessarily. Allergens are antigens that provoke a specific immunologic allergic response. All allergens are antigens, although not all are good antigens. All antigens are not necessarily allergens.

List 8 laboratory values that help confirm allergic diseases.

1. Peripheral blood eosinophilia (>500 cells/mm^3)
2. Elevated serum IgE
3. Respiratory or gastrointestinal secretions containing >10% eosinophils
4. Positive allergy skin testing (evidenced by wheals) using prick or intradermal techniques
5. Allergen-specific IgE
6. Positive food and drug challenges
7. Positive bronchial provocation tests to histamine or methacholine challenge
8. Measurement of exhaled nitric oxide (NO) as a surrogate marker for eosinophilic airway inflammation

What are the 3 treatment strategies?

Avoidance of irritant; pharmacotherapy; immunotherapy

List 8 choices in pharmacotherapy.

α-Agonists (reduce edema of mucous membranes); β-agonists (dilate airways); theophylline (treat asthma); cromolyn (smooth-muscle relaxant); topical and systemic steroids; antihistamines; anticholinergics; antileukotrienes

FOOD ALLERGIES

What are food allergies?

IgE-mediated reactions that usually occur up to 1–4 hours after ingestion

List typical symptoms of food allergy.

Nausea, vomiting, diarrhea, anaphylaxis, asthma, eczema, urticaria, or angioedema

What are nonallergic adverse food reactions?

These reactions (more common than allergic ones) can be secondary to toxic substances in food, chemical, or bacterial contaminants, endogenous pharmacologic agents, or metabolic diseases in the individual.

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What is the most common target organ in IgE-mediated food hypersensitivity?	The skin—may exhibit urticaria, angioedema, pruritic rash, or eczema
List 8 of the most common foods to which children are allergic.	Milk, eggs, peanuts, soybean, tree nuts, wheat, shellfish, and fish cause >90% of food allergy reactions.
How is food sensitivity evaluated?	**History** and **physical examination.** (Skin tests and serum-specific IgE antibodies to specific foods are only helpful in assessing food sensitization.)
List 2 methods by which a food allergy can be confirmed.	Food challenges; elimination diets (occasionally)

ALLERGIC RHINITIS

List 4 common causes of seasonal allergies.	Outdoor inhalant allergens: tree, grass, and weed pollens (e.g., ragweed), outdoor mold spores
List 3 common causes of perennial allergies.	Indoor inhalant allergens: pet dander, dust mites, molds (e.g., *Aspergillus*, *Penicillium*)
What are the symptoms of allergic rhinitis?	Profuse, watery nasal discharge; itchy nose; postnasal drip; sneezing; cough. If the eyes are also involved, redness, tearing, and itching are observed (**rhinoconjunctivitis**).
What are "allergic shiners"?	Dark discoloration of the infraorbital area caused by venous stasis secondary to nasal congestion. Also seen in individuals who do not have allergies

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What is the pathophysiology of allergic rhinitis?

An immediate hypersensitivity response occurs in the nasal mucosa of a sensitized individual. Specific IgE, which is stimulated by allergens, binds to mast cells. On re-exposure to the allergen, an allergen IgE-antibody reaction occurs, causing mast cell degranulation and release of mediators (e.g., **histamine, metabolites of the arachidonic acid pathway,** and **inflammatory cytokines**), which increase vascular permeability, smooth-muscle contraction, mucus secretion, and pruritus.

What is the differential diagnosis?

Upper respiratory tract infection, sinusitis, nonallergic rhinitis with eosinophilia, vasomotor rhinitis, rhinitis medicamentosa

What is a Hansel stain?

An eosin methylene blue stain that shows eosinophils well. Stained cell preparations with >10% eosinophils are highly suggestive of allergic rhinitis.

How is allergic rhinitis managed?

1. Avoidance of allergens
2. Treatment options include antihistamines, systemic and topical decongestants, intranasal cromolyn or corticosteroids, antileukotrienes, and immunotherapy.

INSECT STINGS AND BITES

List 7 common stinging insects.

Apidae family—bumble bee, honey bee

Vespidae family—yellow jacket, white-faced hornet, yellow hornet, wasp

Formicoidea family—fire ant

All of the above belong to the Hymenoptera order of insects.

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List 3 classifications of reactions and the characteristics of each.

Local: Swelling < **2 cm** and lasts <**24 hours,** often with local erythema and pruritus

Large local: Swelling **2 cm** and lasts up to **48–72 hours**

Systemic: Diffuse urticaria and pruritus, laryngeal edema, bronchospasm, hypotension, abdominal cramping, nausea, and vomiting

Which classifications are involved in most typical allergic reactions?

Local or large local—most children do not have systemic reactions, and those that do occur are rarely life-threatening. Of children who have life-threatening reactions, <50% have a second life-threatening event after another sting.

List 4 components of the long-term management of insect allergy.

Education; avoidance; kits containing epinephrine for acute treatment after a sting; immunotherapy for children who have experienced a significant systemic reaction

How effective is immunotherapy?

At least 95% effective if a maintenance dose of 100 μg of venom is achieved, the amount in an average sting

How long should the injection immunotherapy be continued?

At least 3–5 years in childhood, and possibly longer for adults

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Chapter 30 Genetics

DEFINITIONS

What is the difference between the terms *congenital* and *genetic*?

Congenital means appearing at birth, without regard to the cause, whereas *genetic* implies that the basis of a disease or defect, at least in part, is determined by the genetic makeup of the individual. By definition, all birth defects are congenital, but many are not genetic.

What is a malformation?

A primary defect in the formation or development of a body part or organ

What is a deformation?

A change in the shape, form, or position of a normally formed body part or organ by extrinsic or mechanical forces

What is a disruption?

A defect caused by breakdown in a previously normal body part or organ

What is a malformation syndrome?

A pattern of multiple primary malformations in an individual from a single underlying cause

What is a sequence?

A primary malformation and 1 or more secondary malformations or deformations

What is an association?

The simultaneous occurrence of 2 or more traits or abnormalities that cannot be explained by chance. The VACTERL (formerly VATER) association is the best-known pediatric association.

What is an autosome?

A non-sex chromosome (i.e., a chromosome other than X or Y)

What is an autosomal condition?

A condition caused by an abnormality involving a gene on an autosome

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What is a mendelian trait or condition?

A genetic condition that is inherited as a single gene trait, the occurrence and recurrence of which conform to Mendel's laws

What is an autosomal recessive trait?

A trait or condition found when the affected person has a pair of mutant genes (i.e., is homozygous) for that condition. With true autosomal recessive traits, heterozygotes are free of clinical disease.

What is an autosomal dominant trait?

A condition caused by the presence of a single mutant gene, rather than a pair of mutant genes

What is a multifactorial trait?

A trait or condition caused by the interaction of multiple genes as well as additional nongenetic factors; these traits account for many common birth defects.

What is anticipation?

A phenomenon in which a genetic condition becomes more severe or has an earlier age of onset in succeeding generations. Most appear to be caused by expansions of trinucleotide repeats, such as fragile X syndrome.

What is mosaicism?

The presence of 2 or more genetically different cell lines in the same individual. One of these cell lines is usually normal.

COMMON GENETIC SYNDROMES

What is Down syndrome?

A recognizable pattern of malformations caused by the presence of extra chromosome 21 material. Also called "trisomy 21" if caused by nondisjunction

What are the features of Down syndrome?

In infants:

Hypotonia, flattened occiput (brachycephaly), redundant skin (especially on the posterior neck), flattened midface, epicanthal folds, upslanted palpebral fissures, small ears, prominent or protruding tongue, single transverse palmar creases, congenital heart disease (particularly atrioventricular canal defect; Ch 16)

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Older children:	Same as those for infants with associated developmental delay, mental retardation, or both
List 5 other complications.	Congenital duodenal obstruction; Hirschsprung disease (Ch 19, p. 293); hypothyroidism (congenital or acquired; Ch 24, p. 386); increased incidence of respiratory infections; increased risk of leukemia
What causes Down syndrome?	Extra material from chromosome 21, either through trisomy or a translocation. Ninety-five percent of cases are caused by trisomy 21, and 5% are caused by unbalanced translocations resulting in the presence of extra chromosome 21 material or by mosaicism.
What is the most common risk factor for Down syndrome?	Advanced maternal age (older than 35 years at delivery)
List 6 features of trisomy 13.	Oral or facial clefts (or both), microphthalmia, postaxial polydactyly, apical scalp defects, intrauterine growth retardation, congenital heart disease
What is the prognosis for trisomy 13?	Most children die in the first year of life. Survivors are usually profoundly retarded.
List 6 features of trisomy 18.	Intrauterine growth retardation, small ears with flattened helices, small mouth, congenital heart disease, omphalocele (Ch 23, p. 360), unusual hand positioning (second and fifth fingers overlapping the third and fourth)
What is the prognosis for trisomy 18?	Most children die in the first year of life. Survivors are retarded, although some learn communication skills.

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What is Turner syndrome?

Classic Turner syndrome is caused by the absence of 1 X chromosome in a female. About 50% of patients with the Turner syndrome phenotype have a 45,X karyotype, and about 20–30% are mosaic. About 10–20% have a structural rearrangement involving a deletion of part or all of the short arm of 1 of the 2 X chromosomes.

What are the features of Turner syndrome?

Girls with Turner syndrome may have short stature, delayed puberty with primary amenorrhea (caused by gonadal dysgenesis), lymphedema of the hands and feet, coarctation of the aorta (29%; Ch 16), kidney malformations (50%), webbed neck, shield (broad) chest, and prominent, posteriorly rotated ears.

Are girls with Turner syndrome retarded?

Not usually, although they may have problems in spatial perceptual ability.

What is Klinefelter syndrome?

A syndrome seen in males who have an extra X chromosome (47,XXY)

What are the features of Klinefelter syndrome?

In young boys?

Generally few physical abnormalities

Older patients?

Above-average height, small testes, gynecomastia, fat distribution on hips and chest, below-average IQ

What is fragile X syndrome?

An X-linked condition caused by the expansion of a trinucleotide repeat in the *FMR1* gene

What are the features of the fragile X syndrome?

Young boys with the full mutation may have developmental delay, large ears, and a long face. Postpubertal boys usually have enlarged testes. Boys with the full mutation are usually cognitively impaired. Girls with the full mutation may also be cognitively impaired, but usually are less severely affected. They generally have few physical findings.

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What is a premutation for the fragile X syndrome?

An increase in the size of the trinucleotide repeat from the normal size (usually <50 repeats) to about 52–200 repeats. Individuals with the premutation are usually asymptomatic, but females risk having children with a full mutation.

What is neurofibromatosis type 1 (von Recklinghausen disease)?

An autosomal dominant disorder characterized by hyperpigmented macules (café au lait spots; Ch 27, p. 448) and cutaneous neurofibromata

List 4 common findings.

Multiple (usually >6) hyperpigmented macules, Lisch nodules of the iris, axillary freckling, cutaneous neurofibromata

What are some complications?

Pseudoarthrosis, scoliosis, meningioma, optic glioma, seizures, learning disabilities, mental retardation, pheochromocytoma, hypertension, malignant degeneration of a neurofibroma, leukemia

What is tuberous sclerosis?

An autosomal dominant disorder characterized by hamartomas, hypopigmented skin lesions, and an increased risk of seizures and mental retardation. The kidney, heart, and eyes may also be affected by this condition.

What is the classic triad of symptoms?

Hypopigmented skin lesions, with epilepsy and mental retardation

What are 5 physical findings?

"Ash leaf" hypopigmented macules (found most commonly on the trunk), shagreen patches, adenoma sebaceum, periungual fibromas, and intracranial lesions, which are sometimes calcified

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What are the signs and symptoms with which pediatric patients present?

Seizures; developmental delay, mental retardation, or both; skin lesions. Skin lesions may not be present or obvious in early infancy.

Hypopigmented skin lesions may be easier to see in light-skinned patients if viewed with a Wood's (ultraviolet) lamp.

What is the prognosis for tuberous sclerosis?

It varies. Some patients are healthy, whereas others may have severe mental retardation, seizures, or cardiac tumors.

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Index

Page numbers in *italic* designate figures.

Abbreviations, common medical, 3–8
ABC (airway, breathing, circulation), 30
  in cardiac arrest, 34, 39
  in coma, 39
  in head trauma, 41
  in major trauma, 50
  in near drowning, 48
  in respiratory distress, 34
Abdominal pain, 124
Abdominal wall defects (*see also* Hernia)
  gastroschisis, 362–363
  omphalocele, 360–362
ABGs (arterial blood gases), in neonatal cyanosis, 88
Abscess
  perianal/perirectal, 309–310
  peritonsillar, 250–251
  renal or perirenal, 482
Acid(s)
  essential amino, 24
  essential fatty, 24, 25
  retinoic, 126
Acidosis
  metabolic, 54
  renal tubular (RTA), 336–337
Acne, 125–126
Acrocyanosis, 73
Acute cardiovascular collapse, 33–40 (*see also under* Emergencies)
Acute chest syndrome, 170
Acute lymphoblastic leukemia (ALL), 419–420
Acute myelocytic leukemia (AML), 420–421
Acute renal failure, 328–330
Acute rheumatic fever, 206–207
Acute threatening life events (ALTEs), 104
Addison disease, 389 (*see also* Adrenocortical insufficiency)
Admission notes, 2
Adolescent medicine, 155–162
  accidental and nonaccidental death, 161–162
  contraception, 160–161
  emancipation and consent, 157
  hygiene issues, 157–158
  immunizations, 157, 160
  legal issues, 157
  menstruation, 158–159
  pregnancy, 160–161
  puberty and growth, 155–157
  sexually transmitted diseases (STDs), 159–160
Adrenal crisis, 390
Adrenal hyperplasia, 390
Adrenal hypoplasia, congenital, 38
Adrenocortical insufficiency, 389–390
Age
  bone, 377
  gestational, 67
  for sports participation, 134
Airway, pediatric differences, 30
Airway disease, reactive (*see* Asthma)
Airway foreign body, 227–228
Airway obstruction, intra- vs. extrathoracic, 31
Alagille syndrome, 325–327
Albendazole, in ocular larva migrans, 504
Albinism, 449–450
Alcohol abuse, 152
Alkaline phosphatase test, 312
Allergens, vs. antigens, 508
Allergic conjunctivitis, 469
Allergic disorders, 507–511
  allergic rhinitis, 509–510
  atopy, 507–508
  food allergies, 508–509
  insect stings and bites, 510–511
  sports participation and, 148
Allergic rhinitis, 509–510
Allergic shiner, 509
Alopecia, 460
Alpha-fetoprotein, 348

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ALTEs (acute threatening life events), 104
Amino acids, essential, 24
Amniocentesis, 95
AMPLE history, 31
Anaphylaxis, exercise-induced, 148
Androgens, acne and, 125
Anemia (*see also* Blood and blood products)
  anemias, 168
  aplastic, 173–174
  in chronic renal failure, 332
  cyanosis and, 87
  Fanconi, 173–174, 180
  hemolytic, 171–173
    glucose-6-phosphate dehydrogenase deficiency, 171–172
    spherocytosis, 172–173
  nutritional, 163–166
    folate-deficiency anemia, 164–165
    iron-deficiency anemia, 163–164
    vitamin B12 deficiency (pernicious anemia), 165–166
  screening for, 117
  treatment, 332
Anencephaly, 402
Anesthetic
  for circumcision, 77
  for IVs, 11
Ann Arbor classification, of Hodgkin disease, 414
Annular pancreas, 297
Anorectal malformations (imperforate anus), 275–277
Anterior pituitary hormones, 382
Antibiotic-related (pseudomembranous) colitis, 2
Antibiotic therapy
  in bacterial meningitis, 103
  in conjunctivitis, 468
  in glomerular nephritis, 334
  in otitis media, 470–471
  prophylactic ocular in newborn, 76
  in syphilis, 485
  in vesicoureteral reflux, 336
Anticipation, genetic, 513
Anticipatory guidance, 117–119
Antidiuretic hormone (ADH, vasopressin), 340
  syndrome of inappropriate (SIADH), 388
Antigens, vs. allergens, 508
Anuria
  intrauterine, 84
  neonatal, 83–86
Aorta, coarctation, 194–195
Aortic stenosis, 195–196
APGAR mnemonic, 63
Apgar scores, 63, 64*t*
Aphthous stomatitis, 475
Aplastic anemia, 173–174
Apnea of infancy, 104–105
Appendicitis, 282–283
Arnold Chiari malformation, 397–398
Arrhythmias, 211–212
  atrial, 211
  ventricular, 212
Arterial catheters, 109
Arterial puncture, 13–14
Ascaris lumbricoides, 503
Aseptic meningitis, 465–467
Aspiration
  myringotomy with (tympanocentesis), 473
  tissue, 439
Association, genetic, 512
Asthma, 223–227
  classification of, 225
  exercise-induced, 147–148
  refractory period in, 147
  sports participation and, 147
  theophylline overdose signs, 227
Atopic dermatitis (eczema), 436–437
Atopy, 507–508
Atresia
  biliary, 339–340
  choanal, 73, 245
  esophageal, 300–303, *301*
  intestinal, 298–399, *299*
  laryngeal, 248
  pulmonary with intact ventricular septum, 199–200
  tricuspid, 204–205
  vaginal, 353
Atrial baffle repair, 190
Atrial septal defect, 190–191
Atrioventricular septal (canal) defect, 192–194
Atrophy, spinal muscular (SMA), 404
Attention deficit hyperactivity disorder (ADHD), 130–131
Atypical mycobacteria, 506
Auscultation, findings in pneumonia, 221

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Autoimmune neutropenia, 178
Autosomal condition, 512
Autosomal dominant trait, 513
Autosomal recessive trait, 513
Autosome, 512

Bacille Calmette-Guerin (BCG) vaccination, 501
Bacterial cystitis, 480
Bacterial endocarditis, 205–206
Bacterial gastroenteritis, 477–479
Bacterial meningitis, 462–465
  neonatal, 102–104
Ballard Score, New, 69
Barium swallow, in poisoning, 126
Becker muscular dystrophy, 404
Beckwith-Weidemann syndrome, 410
Bezoar, 306–307
Bicarbonate replacement, in renal tubular acidosis, 337
Bile duct, obstruction of, 312
Biliary atresia, 320–321
  correctable, 320
Biliary colic, 311
Bilirubin, 79–81
Biopsy, muscle, in neonatal hypotonia, 91
Birth control, 161
Blalock-Taussig shunt, 189
Blastema, nodular renal (nephroblastomatosis), 412
Bleeding (*see also* Hemorrhage)
  dysfunctional uterine, 159
  esophageal variceal, 324, 325
Blood and blood products, 20–22 (*see also* Hematologic disorders)
  cytomegalovirus negative, 20
Blood-borne infectious diseases, sports participation and, 146
Blood culture
  in cellulitis, 440
  epiglottitis, 241
  in meningitis, 464
  in pneumonia, 222
Blood gases, arterial (*see* ABGs)
Blood pressure (*see also* Hypertension)
  in major trauma, 51
  measurement in children, 342
Blood tests, in sepsis, 102
Blueberry muffin rash, 71
Bochdalek hernia, 216–217
Body fat measurement, 136
Body surface area (BSA), 15
Body temperatures, 111, 112
Bone age, 377
Bone marrow findings, in Fanconi anemia, 180
Bone marrow transplantation, in aplastic anemia, 174
Bowel stricture, 256
Bradycardia
  pathologic of newborn, 31
  toxic agents causing, 53
Branchial cleft remnants, 248
Breast-feeding, 23–24, 112
  thrush prevention, 474
  time to begin, 77
Breast milk, 23, 111
Breast tissue, in newborns, 71
Breslow classification, of melanoma, 433
Bronchogenic cyst, 235–236
Bronchoscopy, in pneumonia, 222
Brudzinski sign, 103, 462
BSA (body surface area), 15
Budd-Chiari syndrome, 323
Bullous impetigo, 439
Burkitt lymphoma, 417
Burn prevention, 118

Café au lait spots, 448
Caloric requirements, 23
  of athlete, 151
Cancer (*see also* Neoplastic disease)
  cervical, HPV and, 487
  penile, 77
  testicular, 345
Caput succedaneum, 72
Carcinoma
  embryonal, 351
  hepatocellular, 430
Cardiac arrest, 33–34
Cardiac catheterization, 197
  aortic stenosis, 196
  pulmonary atresia, 200
  pulmonary hypertension, 213
Cardiac complications, of congenital rubella, 491
Cardiogenic shock, 36, 38, 39
Cardiology, 188–214
  acquired heart disease, 205–214
  congenital heart defects, 190–205 (*see also* Congenital heart defects)
  general considerations, 188–190

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Cardiomegaly, in sickle cell disease, 171
Cardiomyopathy, 237–238
  dilated, 210
  hypertrophic, 208–209
  in sickle cell disease, 171
Cardiovascular fitness assessment, 135
Cardipulmonary arrest, 30
Car seats, 78
Catheterization
  arterial, 13, 14, 109
  nasal of newborn, 73
  suprapubic, 480
  umbilical cord, 66
Catheters, arterial, 13, 14, 109
CCAM (congenital cystic adenomatoid malformation), 237–238
CD4 counts, 496
Celiac sprue (gluten enteropathy), 263–266
Cells, Reed-Sternberg, 413
Cellulitis, 439–441
  erysipelas, 440
  orbital, 440
  periorbital, 440
Central vs. peripheral cyanosis, 38
Cephalohematoma, 72
Cerebral edema, 42, 43
Cerebral palsy, 398–399
Cerebral perfusion pressure (CPP), 42
Cerebrospinal fluid (CSF)
  diagnostic findings in, 12
  lumber puncture, 11, 12
  in meningitis, 463
  in tuberculosis, 499
Cervical cancer, HPV and, 160, 487
Cervical lymphadenitis, 506
Chancroid, 486
Charley horse (muscle hematoma), 138
Chédiak-Higashi syndrome, 177
Chest syndrome, acute, 196–197
Chicken pox (varicella), 493
Chlamydia infection, 486
Chlamydia trachomatis, 468
  in neonatal pneumonia, 100
Chloridorrhea, congenital, 281
Choanal atresia, 73, 245
Cholecystitis, 312
Choledochal cysts, 321–323
Cholelithiasis (gallstones), 311–313
Cholesterol stones, 311
Chordee, 74, 344
Choriocarcinoma, 351
Chromosome abnormalities
  in acute myelocytic leukemia, 420
  hypospadias in, 344
Chronic granulomatous disease (CGD), 177
Chronic lung disease (CLD), 95–96
Chronic renal failure, 330–332
Circumcision, 77
Clark melanoma staging, 433
Cleft
  branchial, remnants of, 248
  laryngeal, 247
Cloacal malformations, 277
Coagulation defects, 182–185
  disseminated intravascular coagulation (DIC), 185
  hemophilia A (Factor VIII deficiency), 182–184
  hemophilia B (Factor IX deficiency), 182–184
  von Willebrand disease, 184–185
Coarctation of aorta, 194–195
Cocaine, 152
Colic, biliary, 311
Colitis
  antibiotic-related (pseudomembranous), 263
  ulcerative, 257–260
Color vision, testing for, 121
Coma
  CT in, 47
  definition, 47
  differential diagnosis, 47
Comedones
  closed (whiteheads), 125
  open (blackheads), 125
Communicating hydrocele, 357
Communication, 1
Compartment syndrome, 10
Compensated shock, 35, 36
Complete blood count (CBC), in iron-deficiency anemia, 163
Compliance, lung, 89
Concussion, 41
  excessive repetitions, 144
  grades of, 142
  management of, 143–144
Condylomata acuminata (genital warts), 487
Congenital adrenal hyperplasia, 393
Congenital chloridorrhea, 281

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Congenital cystic adenomatoid malformation (CCAM), 237–238
Congenital diphragmatic hernia (CDH), 215–218
Congenital duodenal obstruction, 296–298
Congenital glucose-galactose malabsorption, 281
Congenital heart defects
  acute rheumatic fever, 206–207
  aortic stenosis, 195–196
  arrhythmias, 211–212
  atrioventricular septal (canal) defect, 192–194
  cardiomyopathy, 208–210
  coarctation of aorta, 194–195
  myocarditis, 207–208
  patent ductus arteriosis, 194
  pulmonary stenosis, 196–197
  ventricular septal defect, 191–192
  cyanosis and, 72
  cyanotic, 99, 200
    bacterial endocarditis, 205–206
    Eisenmenger syndrome, 214
    hypoplastic left heart syndrome, 203–204
    persistent truncus arteriosus, 200–201
    pulmonary atresia with intact ventricular septum, 199
    tetralogy of Fallot, 198–199
    total anomalous pulmonary venous return, 201–202
    transposition of great arteries (TGA), 197, 204
    tricuspid atresia, 204–205
Congenital heart lesions, 38
Congenital hepatic fibrosis, 325
Congenital hip dysplasia, 116
Congenital melanocytic nevi, 448
Congenital platelet disorders, 179–181
Congenital rubella, 491, 492
Conjunctivitis, 467–469
  allergic, 469
  epidemic, 468
Constipation, 287–289
  chronic, 289
  functional vs. Hirschsprung disease, 289
Continuous venovenous hemofiltration, 330
Contraception, 160–161
Contrast enema
  in Hirschsprung disease, 293
  in intestinal atresia, 300
  in intussusception, 274
Cord hydrocele, 357, *359*
Corticosteroids, 1
  in croup, 244
  in Henoch-Schönlein purpura, 280
  in meningitis, 465
  in otitis media, 469
Coxsackievirus, 475
CPAP (continuous positive airway pressure), 109
Cramps
  heat, 152
  menstrual, 159
Cranial nerves, perinatal evaluation, 75
Creatinine clearance, 330, 331
Crohn's disease, 260–262
Croup, 242–244
  vs. epiglottitis, 240
Cryoprecipitate, 22
Cryptorchidism, 344–345
CT (computed tomography)
  in coma, 47
  in pancreatitis, 283–285
Cushing disease/Cushing syndrome, 390–391
Chvostek sign, 372
Cyanosis
  central vs. peripheral, 38
  heart defects associated with, 72, 197
  neonatal, 87–88
  O_2 saturation in, 87
Cyanotic congenital heart disease (*see under* Congenital heart defects)
Cyclic neutropenia, 177
Cyst(s)
  bronchogenic, 235–236
  choledochal, 321–323
  esophageal duplication, 235
  lung, 220–221
Cystic fibrosis, 228–229
Cystitis, bacterial, 480
Cystourethrogram, voiding, 335
Cytomegalovirus negative blood, 20

D10, 28
Dactylitis, 482 (*see also* Sickle cell disease)

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Deformation, 512
Dehydration, 16–19
  in diabetic ketoacidosis, 368, 370
  hypernatramic, 17–18
  hyponatremic, 18–19
  isotonic, 16–17
Delayed puberty, 62, 378–380
Delivery room assessment, 69
Denys-Drash syndrome, 411
Dermatitis
  atopic (eczema), 436–437
  contact, 437–438
  seborrheic, 436
Desmopressin (dDAVP), 339
Development, 58–62 (*see also* Growth)
  Denver Developmental Screening Test, 61
  fine motor, 59–60
  gross motor, 59
  guidance for parents, 117
  language, 60–61
  puberty, 61–62
  sensory impairment and, 59
  social milestones, 60
Developmental delay, 127
  mental retardation and, 61
Developmental monitoring, 115
Diabetes insipidus, 338–339
  nephrogenic, 340–341
Diabetes mellitus, 364–370
  diabetic ketoacidosis (DKA), 368–370
  LGA newborn and maternal, 67
  sports participation and, 149
  sports to avoid, 149
  type 1, 364, 366
  type 2, 364, 365, 366
  types, 364
Diabetic ketoacidosis (DKA), 368–370
Dialysis
  continuous venovenous hemofiltration, 330
  peritoneal, 330
Diaper rash, 445
Diaphragmatic hernia, congenital, 215–218
Diarrhea
  causes of acute, 124
  causes of chronic, 124
  neonatal, 105–106
Diet (*see also* Nutrition)
  food allergies, 508–509
DiGeorge syndrome, 200, 373
Dilated cardiomyopathy, 210
Dimple, spinal, 74
Disorders of sexual development, 392–395
Disruption, genetic, 512
Disseminated intravascular coagulation (DIC), 185
Distributive shock, 36
Diverticulum, Meckel, 308
Documentation, 1
Dopamine, in shock, 40
Double-bubble sign, 297
Down syndrome (trisomy 21), 513–514
  Hirschsprung disease and, 293
  intubation in, 33
Drowning, 47–49
  definition of near, 47
Drug-related meningitis, 467
Dubowitz examination, 69
Duchenne muscular dystrophy, 404
Duct, thyroglossal, remnant, 248–249
Ductus arteriosus, patent, 194
Duodenal obstruction, congenital, 296–298
Dysfunctional uterine bleeding, 159
Dysmenorrhea, 159
Dysplasia, septo-optic, 381, 382
Dystrophy, muscular, 404

Echocardiography, 188, 206, 208
Eczema (atopic dermatitis), 436–437
Edema
  cerebral, 18, 42, 43
  in nephrotic syndrome, 332–333
Edrophonium chloride (Tensilon), 401
Eisenmenger syndrome, 214
Electroencephalography (EEG)
  in neonatal hypotonia, 91
  in seizures, 46, 397
Electrolytes (*see also* Fluid and electrolytes)
  GI fluid concentration, 17, 17*t*
  maintenance requirements, 15
Electromechanical dissociation, 33
Electromyography (EMG), in neonatal hypotonia, 91
ELISA test, 319
Embryonal carcinoma, 351
Emergencies
  acute cardiovascular collapse, 33–40
    cardiac arrest, 33–34
    shock, 35–40

WhiteKnightLove

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adrenal crisis, 390
environmental, 47–54
drowning, 47–49
hypothermia, 49–50
major trauma/pediatric aspects, 50–53
toxicology, 53–54
epiglottitis, 239–242
intracranial hemorrhage, 399–400
neurologic and mental status, 41–47
head trauma, 41–43
seizures, 43–46
unexplained coma, 47
respiratory distress, 31–33
resuscitation in children, 32
thyroid storm, 384
Emphysema, 219–220
lobar, 220
Encephalitis, herpes simplex, 83
Encephalocele, 402
Encephalopathy, hypoxic ischemic, 82, 83
Encopresis, 128–129, 290–291
Endocarditis, bacterial, 205–206
Endocrine disorders, 364–395 (*see also specific conditions*)
adrenocortical insufficiency, 389–390
ambiguous genitalia, 392–395
Cushing disease/Cushing syndrome, 390–391
delayed puberty, 378–380
diabetes insipidus, 338–341
diabetes mellitus, 364–370
growth hormone (GH) deficiency, 380–381
gynecomastia, 377–378
panhypituitarism, 382–383
parathyroid hormone (PTH), 370–373
hyperparathyroidism, 370–372
hypoparathyroidism, 372–373
pheochromocytoma, 391–392
precocious puberty, 375–377
SIADH, 388–389
thyroid, 383–388
hyperthyroidism, 383–386
hypothyroidism, 386–388
vitamin D metabolism, 373–375
Endodermal sinus tumors, 350
Endoscopic retrograde cholangiopancreatography (ERCP), 284, 312
Endotracheal tubes (ETT)
for infants, 65
size, 33
Endotrachel intubation (*see* Intubation)
End-stage renal disease (ESRD), in Henoch-Schönlein purpura, 280
Enema, contrast (*see* Contrast enema)
Enterobius vermicularis, 504
Enterocolitis, necrotizing (NEC), 254–257
Enuresis, 129
Environmental emergencies, 47–54 (*see also under* Emergencies)
Epidemic conjunctivitis, 468
Epididymitis, 346
Epidural hematoma, 41, 42
Epigastric hernia, 358
Epiglottitis, 239–242
differential diagnosis, 240
rifampin prophylaxis, 242
vs. croup, 240, 242
Epileptic seizures (*see* Seizures)
Epinephrine, in shock, 40
Epithelial ovarian tumors, 349, 352
ERCP (endoscopic retrograde cholangiopancreatography), 284, 312
Erysipelas, 440
Erythema infectiosum (fifth disease), 443, 492
Erythema toxicum, 71
Erythrocyte protoporphyrins (FEP), 164
Escherichia coli 0157:H7, 285
Esophageal atresia, 300–303, *301*
Esophageal duplication cyst, 235
Esophageal variceal bleeding, 325
Esophagoscopy, in poisoning, 126
Essential amino acids, 24
Essential fatty acids, 24
Ewing sarcoma, 426
Examination, physical, 113–132 (*see also* Physical examination)
Exercise-induced anaphylaxis, 148
Exercise-induced asthma, 147–148
Extracorporeal membrane oxygenation (ECMO), 217
Eye injuries, 145

Facial hemangioma (port wine stain), 454–455
Failure to thrive, 128

WhiteKnightLove

Freely you have received; freely give.

Fallot, tetralogy of, 198–199
Familial atypical mole melanoma (FAMM) syndrome, 433
Family stresses, 119
Fanconi anemia, 180
Fanconi syndrome, 338
Fatigue
  heat, 152
  in school-age child, 122
Fat-soluble vitamins, 25
Fatty acid deficiency, 25
Fatty acids, essential, 25
Febrile seizures, 45
Felon, 459–460
Fever, 123
Fibrosis
  congenital hepatic, 325
  cystic, 228–229
Fifth disease (erythema infectiosum), 443, 492
Fine motor development, 59–60
Firearms, 118
  suicide and availability, 162
Fistula, tracheoesophageal, 300–303, *301*
Fluid restriction, in meningitis, 465
Fluids and electrolytes, 15–19 (*see also* IVs *and specific electrolytes*)
  in chronic renal failure, 330
  dehydration, 16–19
    hypernatremic, 17–18
    hyponatremic, 18–19
    isotonic, 16–17
  GI electrolyte concentration, 17, 17*t*
  maintenance fluid requirements, 15
  in renal failure, 329
Fluorescent treponemal antibody absorption (FTA-ABS) test, 485
Folate-deficiency anemia, 164–165
Folic acid, in pregnancy, 403
Fontan procedure, 190
Fontanel, 72
Food allergies, 508–509
Foreign body, airway, 227–228
Formula, types of, 111
Fowler-Stevens orchiopexy, 345
Fracture
  during delivery, 73
  skull, 41
Fragile X syndrome, 515, 516
Freckles, 447–448
Fresh frozen plasma, 21
Fungal infections, meningitis in, 467
Gallstones (cholelithiasis), 311–313
  cholesterol, 311
  pigmented, 311
Gardnerella infection, 488
Gastroenteritis, 477–479
  bacterial, 477–479
  viral, 477
Gastrointestinal (GI) disorders, 252–310
  anorectal malformations (imperforate anus), 275–277
  antibiotic-related (pseudomembranous) colitis, 263
  appendicitis, 282–283
  atresia
    esophageal, 300–303, *301*
    intestinal, 298–300, *299*
  bezoar, 306–307
  celiac sprue (gluten enteropathy), 263–266
  congenital duodenal obstruction, 296–298
  constipation, 287–289
  Crohn's disease, 260–262
  diarrhea, 124
    neonatal, 105–106
  encopresis, 128–129, 290–291
  functional abdominal pain in children, 291–293
  gastroenteritis, 477–479
  gastroesophageal reflux (GER), 266–270
  hemolytic-uremic syndrome (HUS), 285–287
  Henoch-Schönlein purpura, 277–280
  Hirschsprung disease, 293–294
    intestinal polyps, 305–306
  intestinal transport defects, 281
  intussusception, 272–274, *272*
  malrotation, 294–296
  Meckel diverticulum, 308
  necrotizing enterocolitis (NEC), 254–257
  pancreatitis, 283–285
  peptic ulcer disease (PUD), 270–272
  perianal/perirectal abscess, 309–310
  pica, 307
  pyloric stenosis, 304
  short-gut (short-bowel) syndrome, 252–254
  tracheoesophageal fistula, 300–303, *301*

WhiteKnightLove

'It is more blessed to give than to receive.

ulcerative colitis, 257–260
volvulus, 295, 296
vomiting, 124
Gastroschisis, 362–363
Gaucher disease, 174–175
Genetics, 520–525
definitions, 512–513
genetic syndromes, 513–517
(*see also* Congenital *entries and specific disorders*)
Genital herpes, 486–487
Genitalia (*see also* Genitourinary disorders *and specific organs*)
ambiguous, 392–395
Genital warts (condylomata acuminata), 487
Genitourinary disorders, 344–355 (*see also* Renal disorders)
cryptorchidism, 344–345
epididymitis, 346
hypospadias, 344
imperforate hymen, 355
neonatal anuria and oliguria, 83–85
ovarian tumors, 349–353
pelvic inflammatory disease (PID), 489
testicular torsion, 346–347
testicular tumors, 347–349
in tuberculosis, 499
urinary tract infection, 479–481
vaginal anomalies, 353–354
vaginal tumors, 354–355
German measles (rubella), 491–492
Germ cell tumors
ovarian, 351
testicular, 348
Germinoma, ovarian, 350
Gestational age, 67, *68*
Gingivostomatitis, 475–476
Glomerulonephritis, 334
Glucose-6-phosphate dehydrogenase deficiency (G6PDD), 171–172
Glucose-galactose malabsorption, congenital, 281
Glutamyl transpeptidase test, 312
Gluten enteropathy (celiac sprue), 263–266
Glycosylated hemoglobin (Hb1c) test, 365
Gonadal stromal tumors
testicular, 347, 349
Gonadoblastoma
ovarian, 349, 352
testicular, 348
Gonorrhea, 483
Granulocyte disorders, 177–179
Granulocyte transfusion, 21
Granuloma, umbilical, 107
Granulosa-theca cell tumors, 352
Graves disease, 383–386 (*see also* Hyperthyroidism)
neonatal, 385
Graves ophthalmopathy, 384
Great vessels, transposition of, 38, 72
Gross motor development, 59
Growth, 55–58, *56, 57* (*see also* Development)
failure
in chronic renal failure, 331
in Crohn's disease, 261
physical examination for, 115
pubertal, 155–157
rate, 55
retardation, symmetric, 67
short stature, 127
Tanner stages, 155
Growth hormone (GH) deficiency, 395–396
Guillain-Barré syndrome, 400–401
Gynecomastia, 377–378
pseudo, 378

Haemophilus influenzae type b, in epiglottitis, 239
Hair disorders, 460
alopecia, 466
hirsutism, 461
hypertrichosis, 461
tinea capitis, 445
trichotillomania, 460
Hand-foot-mouth disease, 444
Hangnail, 458
Hansel stain, 510
Harlequin color change, 73
Hashimoto's thyroiditis, 388
Hb1c (glycosylated hemoglobin) test, 365
Headache, 132–133
migraine, 132
tension, 132
Head and neck disorders, 239–251
branchial cleft remnants, 248
choanal atresia, 245

WhiteKnightLove

Freely you have received; freely give.

Head and neck disorders (*continued*)
craniosynostosis, 251
croup, 242–244
epiglottitis, 239–242
laryngeal cleft, 247
laryngeal web, 246
peritonsillar abscess, 250–251
Pierre Robin malformation, 244–245
subglottic stenosis, 247–248
thyroglossal duct remnant, 248–249
torticollis, 249–250
vocal cord paralysis, 246
Head injuries, 41–43, 142–145
second impact syndrome, 144
stingers, 145
Hearing loss, in meningitis, 465
Hearing test, 120
Heart defects, congenital (*see* Congenital heart defects)
Heart murmurs, 72
age to look for, 115
Heat, acclimatization to, 150
Heat cramps, 152
Heat exhaustion, 152
Heat fatigue, 152
Heat injury, 152–154
Heat stroke, 152
Heat syncope, 152
Helicobacter pylori, 272
Hemangioma, 452–453
facial (port wine stain), 454
Hematocolpos, 354
Hematologic disorders, 163–187 (*see also* Anemia)
anemias
aplastic, 173–174
hemolytic, 171–173
coagulation defects, 182–185 (*see also* Coagulation defects)
Gaucher disease, 174–175
granulocyte disorders, 177–179
hemoglobinopathies, 167–171
sickle cell disease, 169–171
thalassemia, 167–168
heparin-induced thrombocytopenia, 186
neonatal alloimmune thrombocytopenia, 186–187
platelet disorders, 179–182 (*see also* Platelet disorders)
polycythemia, 175–176
Hematoma
epidural, 41, 42
muscle (Charley horse), 138
subdural, 41
subungual, 459
Hematuria, microscopic, 480
Hemoglobinopathies, 167–171 (*see also under* Hematologic disorders)
Hemolytic anemias, 171–173
glucose-6-phosphate dehydrogenase deficiency, 171–172
spherocytosis, 172–173
Hemolytic-uremic syndrome (HUS), 181, 285–287, 328
Hemophilia A (Factor VIII deficiency), 182–184
Hemophilia B (Factor IX deficiency), 182–184
Hemorrhage (*see also* Bleeding)
intracranial, 399–400
periventricular/intraventricular of newborn, 92–93
posthemorrhagic hydrocephalus and, 93
Henoch-Schönlein purpura, 277–280
Hepatic fibrosis, congenital, 325
Hepatobiliary disorders, 311–325 (*see also* Biliary disorders; Liver disorders)
Alagille syndrome, 325–327
biliary atresia, 320–321
choledochal cysts, 321–323
cholelithiasis (gallstones), 311–313
congenital hepatic fibrosis, 325
hepatitis, 313–319
portal hypertension, 323–324
Hepaticojejunostomy, 321
Hepatitis
A, 314–316, *315*
B, 316–318, *317*
C, 318–319
chronic active vs. chronic persistent, 313
D, 319
E, 319
immunization, 316, 318
Hepatitis B surface antigen (HBsAg), 316, *317*
Hepatoblastoma, 428–430
Hepatocellular carcinoma, 430–431
Hepatoma, 430–431
Hepatosplenomegaly, 325

WhiteKnightLove

'It is more blessed to give than to receive.

Hernia, 356–380, *378* (*see also* Abdominal wall defects)
  Bochdalek, 215–218
  congenital diaphragmatic (CDH), 215–218
  epigastric, 358
  indirect inguinal, 356
  inguinal, 356–358
  Littré, 308, 358
  Morgagni, 215, 216, 217, 218
  Richter's, 358
  sliding, 358
  umbilical, 359–360
Herpes simplex encephalitis, 71
Herpes simplex infection
  genital, 486–487
  gingivostomatitis in, 475–476
Herpes simplex rash, 71
Hgb H disease, 167
High-protein supplements, 151
Hip dysplasia, 74, 116
Hirschsprung disease, 87, 129, 293–294
  vs. functional constipation, 289
Hirsutism, 461
HIV (*see* Human immunodeficiency virus)
HIV DNA polymerase chain reaction test, 495
Hodgkin disease, 413–415
  Ann Arbor classification, 414
  secondary neoplasms, 415
  staging laparotomy in, 414
Homicide, adolescents and, 161
Hormone(s)
  anterior pituitary, 380, 382
  antidiuretic (ADH, vasopressin), 340
    syndrome of inappropriate (SIADH), 388
Human chorionic gonadotropin (hCG), 348, 351
Human immunodeficiency virus (HIV), 494–496
Human papillomavirus (HPV), 487
Hydrocele, 357
  communicating, 357
  cord, 357, *359*
Hydrocephalus, 403–404
  communicating, 403
  noncommunicating, 403
  posthemorrhagic, 93
  X-linked, 404
Hydrocolpos, 354
Hydrometrocolpos, 354
Hydrops fetalis, 167, 492
Hymen, imperforate, 355
Hymenoptera anaphylaxis, 148
Hyperalimentation, 27, 28, 110 (*see also* Total parenteral nutrition)
Hyperbilirubinemia, 79–81
  clinical evaluation, 80
  differential diagnosis, 80
  treatment, 81
Hypercalciuria, 342
Hypercapnia, permissive, 217
Hypercoagulable states, 187
  in nephrotic syndrome, 333
Hypergonadotropic hypogonadism, 379
Hyperkalemia, 329
Hyperparathyroidism, 370–372
Hyperphosphatemia, in hypoparathyroidism, 372
Hyperplasia
  adrenal, 390
  congenital adrenal, 38, 393
Hypertelorism, ocular, 72
Hypertension
  in Henoch-Schönlein purpura, 280
  in pheochromocytoma, 391
  portal, 323–324
  pulmonary, 213–214
  persistent of newborn (PPHN), 98–99
  renal, 342–343
  in renal failure, 329
  toxic agents causing, 53
Hyperthermia, toxic agents causing, 53
Hyperthyroidism, 383–386
  thyroid storm, 384
Hypertrichosis, 461
Hypocalcemia
  in hypoparathyroidism, 372
  transient neonatal, 373
Hypoglycemia, in diabetes mellitus, 368
Hypoglycemic shock, 37
Hypogonadism
  hypergonadotropic, 379
  hypogonadotropic, 379
Hypokalemia, in renal tubular acidosis, 337
Hyponatremia, 389
Hyponatremic dehydration, 18–19
Hypoparathyroidism, 372–373
Hypoplasia, congenital adrenal, 38
Hypoplastic left heart syndrome, 203–204

WhiteKnightLove

Freely you have received; freely give.

Hypospadias, 74, 344
Hypotension, toxic agents causing, 53
Hypothermia, 349–50
  toxic agents causing, 53
Hypothyroidism, 386–388
  acquired, 388
  congenital, 386–387
Hypotonia, neonatal, 90–91
Hypovolemic shock, 35
Hypoxemia, 98
Hypoxic ischemic encephalopathy, 82, 83

Idiopathic thrombocytopenic purpura (ITP), 181–182
IDM (infant of diabetic mother), 69
Immunizations, 117 (*see also* Vaccination; Vaccines)
  adolescent, 157
  hepatitis A, 316
  hepatitis B, 318
  at school physical, 120
Immunodeficiency disorders, lymphoma and, 416
Immunoglobulin, therapeutic uses, 21
Immunotherapy, in allergy, 511
Imperforate anus, 275–277
Imperforate hymen, 355
Impetigo, 439
Incontinence, fecal (encopresis), 290–291
Indirect inguinal hernia, 356
Infection (*see also* Infectious diseases; Sepsis)
  bacterial endocarditis, 205–206
  fever and, 123
  neonatal sepsis, 101
  in nephrotic syndrome, 333
  neutropenia in, 178
  pyelonephritis, 481–482
  urinary tract infection (UTI), 479–481
Infection control, universal precautions, 146, 318
Infectious diseases, 461–506 (*see also specific conditions*)
  atypical mycobacteria, 506
  blood-borne, 146
  common viral syndromes, 489–494 (*see also* Viral syndromes *and specific syndromes*)
  conjunctivitis, 467–469
  dactylitis, 482
  fifth disease (erythema infectiosum), 443, 492
  gastroenteritis, 477–479
  gingivostomatitis, 475–476
  hepatitis, 313–319
  HIV, 494–496
  lymphadenitis, 476
  meningitis, 462–467
  mononucleosis, 146
  otitis media, 469–473
  parasitic infections, 503–505 (*see also* Parasitic infections)
  pertussis (whooping cough), 501–502
  pharyngitis, 474–475
  roseola, 492–493
  sexually transmitted diseases (STDs), 482–489 (*see also* Sexually transmitted diseases)
  sports participation and, 145–149
  thrush, 473–475
  tuberculosis, 497–501
  varicella (chicken pox), 493–494
Infiltration, of IV fluid, 10
Inflammatory bowel disease (IBD)
  Crohn's disease, 260–262
  ulcerative colitis, 257–260
Ingrown toenail, 458–459
Inguinal hernia, indirect, 356
Injury
  overuse, 138
  prevention, 118
  traumatic, 138
Insect stings and bites, 510–511
Insulin, 367, 369
Intensive care
  environment, 111–112
  monitors, 109–110
  in newborns, 109–112
  nutrition during, 110–111
  questions parents ask, 112
  respirators, 109
Interferons, in hepatitis B, 318
International Classification of Epileptic Seizures, 44*t*
International Neuroblastoma Staging System, 408
Intestinal atresia, 298–300, 299
Intestinal perforation, 256, 257, 363
Intestinal polyps, 305–306
Intestinal transport defects, 281 (*see also specific conditions*)

WhiteKnightLove

'It is more blessed to give than to receive.

Intracranial hemorrhage, 399–400
intraventricular, 400
subdural, 399
Intracranial pressure increase (ICP), 42
in meningitis, 464
Intrahepatic portal hypertension, 323
Intralipids, 28
Intraosseous lines, 51
Intravenous administration (*see* IVs)
Intraventricular hemorrhage of newborn, 92–93
Intubation
endotracheal tube (ETT) size, 33
of newborn, 64–65
Intussusception, 272–274, *272*
in Henoch-Schönlein purpura, 278
Iron-deficiency anemia, 163–164
Isoniazid
seizures caused by, 45
side effects, 500
in tuberculosis, 500
Isotonic crystalloid, in major trauma, 51
Itch-scratch cycle, 436
IVs
bicarbonate replacement, 337
blood and blood products, 20–22
complications of, 10
in diabetic ketoacidosis, 368
fluid and electrolytes, 15–19
in gastroenteritis, 477, 479
intraosseous lines, 51
in major trauma, 50–53
in renal failure, 329
total parenteral nutrition (TPN), 27–29, 110–111
venipuncture for, 9–11

JAGGED gene, 326
Jaundice (*see also* Hyperbilirubinemia)
in newborn, 79–81
Jones criteria, for rheumatic fever, 206
Juvenile intestinal polyps, 305–306

Kasabach-Merritt syndrome, 180
Kasai portoenterostomy, 321
Keratinization, 125
Kernig sign, 103, 462
Kidney disorders, 328–332 (*see also* Genitourinary disorders; Renal disorders)
Klinefelter syndrome, 515
Kostmann syndrome, 178
Kugelberg-Welander disease, 405
Kwashiorkor, 26

Labia minora, gestational age and, 74
Laboratory tests (*see* Tests *and specific tests*)
Ladd's procedure, 296
Language development, 60–61
Lanugo, 71
Laryngeal atresia, 248
Laryngeal cleft, 247
Laryngeal web, 246
Lead toxicity, screening for, 120
Lentigines, 448
Leukemia, 418–421
acute lymphoblastic (ALL), 419–420
acute myelocytic (AML), 420–421
Leukemic infiltrates, testicular, 347
Leukocyte adhesion deficiency (LAD), 177
LGA (large for gestational age), 67–69
Li-Fraumeni syndrome, 422, 425
Limp, 131–132
Lithotripsy, 342
Littré hernia, 308, 358
Liver disorders, 311–327 (*see also* Hepatobiliary disorders)
Alagille syndrome, 325–327
congenital hepatic fibrosis, 325
hepatitis, 313–319
hepatoblastoma, 428–430
hepatoma, 430–431
Liver trauma, 52
Lobar emphysema, 220
Lumbar puncture, 11–12
in meningitis, 463, 464, 466
traumatic, 12
Lung compliance, 89
Lung cysts, 220–221
Lung disease, chronic (CLD), 94, 95
Lung disorders (*see* Respiratory/thoracic disorders *and specific conditions*)
Lymphadenitis, 476
cervical, 506
Lymphangioma, 453–454
Lymphoma
Hodgkin disease, 413–415
non-Hodgkin, 416–418

WhiteKnightLove

Freely you have received; freely give.

Macrocephaly, 403–404
Magnetic retrograde cholangiopancreatography (MRCP), 322
Malformation
  anorectal, 275–277
  arnold-chiari, 397–398
  congenital cystic adenomatoid, 237–238
  defined, 512
  syndrome defined, 512
Malrotation, gut, 294–296
Mantoux tuberculin skin test, 498
Marasmus, 26
Marijuana, 152
Marmorata, 74
Math readiness testing, 121
McBurney's point, 282
McCune-Albright syndrome, 376
MCT oil, 25
Measles (roseola), 443, 489–490
  German (rubella), 491–492
Meckel diverticulum, 308
Meconium, failure to pass, 86–87
Meconium aspiration syndrome, 96–98
Meconium staining, 69
Medulloblastoma, 427
Megacolon, toxic (*see* Hirschsprung's disease)
Melanoma, 433–434
  Breslow classification, 433
  Clark staging, 433
MEN (multiple endocrine neoplasia), 371
Menarche, 62, 155 (*see also* Menstruation)
Mendelian trait or condition, 513
Meningitis
  aseptic, 465–467
  bacterial, 462–465
  neonatal bacterial, 102–104
  in tuberculosis, 499
Meningocele, 402
Menstruation, 158–159
  dysfunctional uterine bleeding, 159
  dysmenorrhea, 159
  oligomenorrhea, 158
Mental retardation
  developmental delay and, 61, 127
  hypothyroidism and, 386
  vitamin supplementation in, 26
Mesoblastic nephroma, 412
Metabolic acidosis, 37
Metabolic disease screening, 116
Metabolic disorders (*see* Endocrine disorders *and specific disorders*)
Metaiodobenzylguinine (MIBG) test, 407
Methionine, 165
Metronidazole, in ocular larva migrans, 488
Micrognathia, 245
Microscopic hematuria, 480
Migraine, 132
Milia, 71
Milk, breast, 23, 111
Miosis, toxic agents causing, 53
Mitotic karyorrhexis index, 409
Mitral valve prolapse, 232
Mongolian spots, 71, 447
Mononucleosis, 146
Morgagni hernia, 215–218
Mortality, adolescent, 161–162
Mosaicism, 513
MRCP (magnetic retrograde cholangiopancreatography), 322
MUDPILES acronym, 54
Multifactorial trait defined, 513
Multiple endocrine neoplasia (MEN), 371, 392
Mumps, 490
Muscle biopsy, in neonatal hypotonia, 91
Muscle hematoma (Charley horse), 138
Muscular atrophy, spinal (SMA), 404
Muscular dystrophy, 404
  Becker, 404
  Duchenne, 404
Musculoskeletal disorders
  in Henoch-Schönlein purpura, 278
  limp, 131–132
Musculoskeletal injuries, 138
  grading, 141
  management, 138–142
    acute, 138–139
    advanced level rehabilitation, 140–142
    early rehabilitation, 139
    PRICEM mnemonic, 139
    return to action, 140
Myasthenia gravis, 401
Mycobacteria, atypical, 506

WhiteKnightLove

'It is more blessed to give than to receive.

Mycobacterium sp., 467
Mydriasis, toxic agents causing, 53
Myelomeningocele, 403
Myer-Cotton system, 247
Myocarditis, 207–208
Myringotomy with aspiration (tympanocentesis), 473

Nail disorders, 458–460
  hangnail, 458
  ingrown toenail, 458–459
  paronychia, 459
  subungual hematoma, 459
Nasal catheterization, 73
Necrosis, soft tissue, 10
Necrotizing enterocolitis (NEC), 254–257
Neisseria gonorrhoeae, 468
Neonatal alloimmune thrombocytopenia, 186–187
Neonatal bacterial meningitis, 102–104
Neonatal cyanosis, 87–88
Neonatal Graves disease, 385–386
Neonatal hypocalcemia, transient, 373
Neonatal hypotonia, 90–91
Neonatal pneumonia, 100–101
Neonatal sepsis, 101–102
Neonate (*see* Newborn)
Neoplasia, multiple endocrine (MEN), 371, 392
Neoplastic disorders (*see also* Cancer; Tumors)
  astrosarcoma, 427–428
  Ewing sarcoma, 426
  hepatoblastoma, 428–430
  hepatoma, 430–431
  Hodgkin disease, 413–415
  leukemia, 418–421
  medulloblastoma, 427
  melanoma, 433–434
  neuroblastoma, 406–410
  non-Hodgkin lymphoma, 416–418
  osteogenic sarcoma, 424–425
  ovarian tumors, 349–353
  pheochromocytoma, 391–392
  retinoblastoma, 421–422
  rhabdomyosarcoma, 422–424
  teratoma, 431–432
  tumor lysis syndrome, 418
  vaginal tumors, 354–355
  Wilms tumor, 410–412
Nephritis, in Henoch-Schönlein purpura, 278
Nephroblastomatosis (nodular renal blastema), 412
Nephrogenic diabetes insipidus, 340–341
Nephroma, mesoblastic, 412
Nephrotic syndrome, 332–333
Neural-crest conditions, 407
Neural tube defects, 402–403
  anencephaly, 402
  encephalocele, 402
  meningocele, 402
  myelomeningocele, 403
  spinal bifida occulta, 403
Neuroblastoma, 406–410
  International Neuroblastoma Staging System, 408–409
  mitotic karyorrhexis index, 409
  Shimada classification, 409, 409*t*
Neurofibromatosis type 1 (von Recklinghausen disease), 392, 516
Neurologic disorders, 396–405
  Arnold Chiari malformation, 397–398
  cerebral palsy, 398–399
  Guillain-Barré syndrome, 400–401
  hydrocephalus, 403–404
  intracranial hemorrhage, 399–400
  Kugelberg-Welander disease, 405
  macrocephaly, 403–404
  muscular dystrophies, 404
  myasthenia gravis, 401
  neural tube defects, 402–403
  Reye syndrome, 405
  seizures, 43–46, 396–397 (*see also* Seizures)
  spinal muscular atrophy (SMA), 404
  Werdnig-Hoffman disease, 404
Neurologic emergencies, 41–47 (*see also under* Emergencies)
Neurologic evaluation, of newborn, 67–78
Neutropenia, 177
  autoimmune, 178
  cyclic, 177
Nevi, congenital melanocytic, 448
New Ballard Score, 69
Newborn
  car seat required for, 78
  common clinical conditions, 79–91

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Newborn (*continued*)
anuria and oliguria, 83–86
failure to pass meconium, 86–87
jaundice, 79–81
neonatal cyanosis, 87–88
neonatal hypotonia, 90–91
neonatal seizures, 81–83
respiratory distress, 89
diseases of, 92–108
apnea of infancy and SIDS, 104–105
meconium aspiration syndrome, 96–98
neonatal bacterial meningitis, 102–104
neonatal diarrhea, 105–106
neonatal pneumonia, 100–101
neonatal sepsis, 101–102
periventricular/intraventricular hemorrhage, 92–93
persistent pulmonary hypertension of newborn (PPHN), 98–99
respiratory distress syndrome (RDS), 94–96
transient tachypnea of newborn (TTN), 96
umbilical abnormalities, 106–107
umbilical hernia, 107–108
IDM (infant of diabetic mother), 69
intensive care, 109–112
LGA (large for gestational age), 67–69
pathologic bradycardia of, 31
perinatal care and evaluation, 63–78
Apgar scores, 63, 64*t*
complete physical examination, 69
delivery room assessment, 69
evaluation, 67–78, *68*
laboratory studies, 78
neurologic examination, 75
resuscitation, 64–66
umbilical cord catheterization, 66
respiratory rate of normal, 70
vitamin K supplementation, 25, 76
well-child visits for, 113–119
Nitric oxide, in pulmonary hypertension, 217
Nodular renal blastema (nephroblastomatosis), 412
Non-Hodgkin lymphoma, 416–418
associated immunodeficiency conditions, 416
Burkitt, 417
large-cell, 417
lymphoblastic, 419–420
NOTCH2 gene, 326
Notes and presentations, 1–3
admission notes, 2
documentation and communication, 1
oral presentations, 2–3
progress notes, 2
SOAP notes, 2
Nutrition, 23–29
breast-feeding, 23–24
in celiac sprue, 263
in diabetes mellitus, 367
folic acid in pregnancy, 403
food allergies, 508–509
guidance for parents, 118–119
during intensive care, 110–111
IV and parenteral hyperalimentation, 27, 110
pica, 307
sports participation and, 150–152
total parenteral (TPN), 27–29
types of formula, 111
Nutritional anemias, 163–166 (*see also under* Anemia)

O2 saturation, in cyanosis, 87, 88
Obesity, 128
heat acclimatization and, 150
Obstruction
of common bile duct, 312
congenital duodenal, 296–298
urinary, 85
Ocular hypertelorism, 72
Ocular larva migrans, 504
Oligomenorrhea, 158
Oliguria
classification of, 328
neonatal, 83–86
Omphalocele, 360–362
Ophthalmologic complications, of congenital rubella, 491
Ophthalmopathy, Graves, 384
Oral presentations, 2–3
Orbital cellulitis, 440
Orchiopexy, 345
Osteodystrophy, renal, 332
Osteogenic sarcoma, 424–425
Otitis media, 469–473
acute (AOM), 469–471

WhiteKnightLove

'It is more blessed to give than to receive.

chronic (COM), 472–473
conjunctivitis and, 468
with effusion (OME), 471–472
recurrent, 471
Ovarian tumors, 349–353
choriocarcinoma, 351
embryonal carcinoma, 351
endodermal sinus, 350
epithelial, 349, 352
germinoma, 350
gonadoblastoma, 352
granulosa-theca cell, 352
Sertoli-Leydig cell, 352
sex cord stromal, 349
teratoma, 350
Overuse injury, 138, 141
Oximetry, in pneumonia, 221
Oxygen
conventional rate, 109
high-frequency, 109
intensive-care administration, 110
transcutaneous, 110

Pacemakers, 211
Pain, abdominal, 124
Palate abnormalities, in Pierre Robin malformation, 245
Pancreas, annular, 297
Pancreatitis, 283–285
Panencephalitis, subacute sclerosing (SSPE), 490
Panhypopituitarism, 382–383
Paraganglioma, 391
Paralysis, vocal cord, 246
Parasitic infections, 503–505
hookworm, 505
lice (pediculosis), 446–447
meningitis in, 467
pinworms, 504–505
roundworm, 503
scabies, 446
visceral larva migrans (toxocariasis), 503–504
whipworm, 505
Parathyroid hormone (PTH) disorders, 370–373
hyperparathyroidism, 370–372
hypoparathyroidism, 372–373
Parenteral nutrition, 253
peripheral (PPN), 27
total (TPN), 27–29
Paronychia, 459
Patent ductus arteriosis, 194
Pathologic bradycardia of newborn, 31
Pectus carinatum, 234
Pectus excavatum, 232–234
Pediatric Advanced Life Support (PALS) manual, 34
Pediatric procedures, 9–14
arterial puncture, 13
lumbar puncture, 11–12
suprapubic puncture, 13
venipuncture, 9–11
Pelvic inflammatory disease (PID), 489
Penicillin (*see also* Antibiotic therapy)
in syphilis, 485
Penile cancer, 487
Penile ulcers, 487
Penis
appropriate size of infant, 394
newborn evaluation, 74
Peptic ulcer disease (PUD), 270–272
Percussion, findings in pneumonia, 221
Perforation, intestinal, 257
Perfusion pressure, cerebral (CPP), 42, 43
Perianal/perirectal abscess, 309–310
Perinatal care and evaluation, 63–78 (*see also under* Newborn)
Perineal reconstruction, 395
Periorbital cellulitis, 440, 441
Peripheral parenteral nutrition (PPN), 27
Peripheral vs. central cyanosis, 38
Peritoneal dialysis, 330
Periventricular hemorrhage of newborn, 92–93
Permissive hypercapnia, 217
Persistent pulmonary hypertension of newborn (PPHN), 98–99
Persistent truncus arteriosus, 200–201
Persistent urachal remnant, 107–108
Pertussis (whooping cough), 501–502
Petechiae, in bacterial meningitis, 462
Pharyngitis, 474–475
Pheochromocytoma, 391–392
Phobia, school, 129–130
Physical disabilities, school and, 119
Physical examination, 113–132
common clinical conditions, 123
abdominal pain, 124
acne, 125–126
attention deficit hyperactivity disorder (ADHD), 130–131

WhiteKnightLove

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Physical examination (*continued*)
developmental delay, 127
diarrhea, 124
encopresis, 128–129
enuresis, 129
failure to thrive, 128
fever, 123
headache, 132
limp, 131–132
obesity, 128
poisoning, 126
respiratory distress, 123
school phobia, 129–130
short stature, 127
vomiting, 124
wheezing, 123
preparticipation sports assessment, 134–137
school physical, 119–122
certifications, 120
immunizations given at, 120
math readiness, 121
screening laboratory tests, 120
size issues, 121–122
speech evaluation, 121
vs. well-child visit, 119
well-child visit, 113–119
anticipatory guidance, 117–119
developmental monitoring, 115
growth variable tracking, 115
immunizations, 117
measurements to take, 114–115
for newborn, 114
scheduling of, 114
screening laboratory tests, 116–117
vs. school physical, 119
Pica, 307
Pierre Robin malformation, 244–245
choanal atresia in, 245
Pigmented gallstones, 311
Pinworms, 504–505
Plasma, fresh frozen, 21
Platelet disorders, congenital, 179–181
Fanconi anemia, 180
hemolytic-uremic syndrome, 181
idiopathic thrombocytopenic purpura (ITP), 181–182
Kasabach-Merritt syndrome, 180
TAR syndrome, 179
Wiskott-Aldrich syndrome, 179
Platelet dysfunction, 21
Platelet transfusions, 21
Pneumonia, 221–223
in meconium aspiration syndrome, 97
neonatal, 100–101
Pneumonitis, in meconium aspiration syndrome, 97
Pneumothorax, 230–231
Poisoning, 126
prevention, 126
theophylline overdose signs, 227
Polycythemia, 175–176
Polydactyly, 74
Polymorphonuclear neutrophil leukocytes (PMNs), 463
Polyps, intestinal, 305–306
Portal hypertension, 323–324
intrahepatic, 323
suprahepatic, 324
Portal vein thrombosis, 324
Portosytemic shunt, 324
Port wine stain (facial hemangioma), 454
Posthemorrhagic hydrocephalus, 93
Posttraumatic respiratory distress, 31
Potassium supplements, 153
Pott disease (spondylitis), 499
Power drinks contraindicated, 153
Precocious pseudopuberty, 376
Precocious puberty, 62, 156, 349, 375–377
central, 375
peripheral, 376
Pregnancy, adolescent, 160–161
Prehn's sign, 347
Premature adrenarche, 376, 377
Premature menarche, 380
Premature thelarche, 376, 377
Prematurity (*see also* Intensive care)
questions parents ask, 112
Pressure
cerebral perfusion (CPP), 42
increased intracranial (ICP), 42
PRICEM mnemonic, 139
Primary (primitive) reflexes, 76
Procedures, pediatric, 9–14 (*see also* Pediatric procedures *and specific procedures*)
Progress notes, 2
Prostaglandin E1
complications of, 38
in shock, 39
Protein, in TPN, 28
Protein requirements, 23

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Proteus syndrome, 456
Pseudogynecomastia, 378
Pseudomembranous (antibiotic-related) colitis, 263
Pseudomonas sp., in neonatal pneumonia, 100
Psoas sign, 282
Psychological conditions, school phobia, 130
Puberty, 61–62, 155–157 (*see also* Adolescent medicine)
  delayed, 62, 156, 378–380
  precocious, 62, 156, 349, 375–377
Pulmonary atresia with intact ventricular septum, 199–200
Pulmonary disorders (*see* Respiratory/thoracic disorders *and specific conditions*)
Pulmonary fitness evaluation, 136
Pulmonary hypertension, 213–214
  persistent of newborn (PPHN), 98–99
Pulmonary sequestration, 236–237
Pulmonary stenosis, 196–197
Pulmonary venous return, total anomalous, 201–202
Pulse oximetry, 110
Puncture
  arterial, 13–14
  lumbar, 11–12, 463
  suprapubic, 13, 480
Purpura
  in bacterial meningitis, 462
  Henoch-Schõnlein, 277–280
  idiopathic thrombocytopenic (ITP), 181–182
Pyelonephritis, 481–482
Pyloric stenosis, 304
Pyuria, 480

Rapid plasma reagin (RPR) test, 484
Rashes, 443, 444
  blueberry muffin, 71
  herpes simplex, 71
RBCs (*see also* Blood and blood products)
  packed, 20
Reactive airway disease (*see* Asthma)
Red reflex, 73
Reed-Sternberg cells, 413
Reflex
  primary (primitive), 76
  red, 73
Reflux, vesicoureteral (VUR), 335–336, 481
Refractory period, in asthma, 147
Rehabilitation
  advanced level, 140–142
  early, 139
  return to action phase, 140
Renal dialysis, 329
Renal disorders, 347–363
  abscess, 435
  diabetes insipidus, 338–341
  end-stage renal disease (ESRD), 331
  Fanconi syndrome, 338
  glomerular disease, 331
  glomerulonephritis, 334
  in Henoch-Schönlein purpura, 278, 280
  hypertension, 364–343
  impetigo and, 439
  minimal-change disease, 332, 333
  nephrotic syndrome, 332–333
  pyelonephritis, 481–482
  renal failure
    acute, 328–330
    chronic, 330–332
  renal stones (nephrolithiasis), 341–342
  renal tubular acidosis (RTA), 336–337
  tumors, 411
  vesicoureteral reflux (VUR), 335–336
  Wilms tumor, 410–411
Renal failure (*see under* Renal disorders)
Renal malformations, 84
Renal or perirenal abscess, 482
Renal osteodystrophy, 332
Renal stones (nephrolithiasis), 341–342
Renal tubular acidosis (RTA), 336–337
Respirators, for newborns, 109
Respiratory burst, 177
Respiratory distress, 31–33, 123–132
  in epiglottitis, 240
  intrathoracic vs. extrathoracic obstruction, 31
  of newborn, 94–96, 105–107
  posttraumatic, 31
Respiratory syncytial virus, 242
Respiratory/thoracic disorders, 215–238
  airway foreign body, 227–228
  asthma, 223–227
  bronchogenic cyst, 235–236
  chylothorax, 231–232
  congenital cystic adenomatoid malformation (CCAM), 237–238

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Respiratory/thoracic disorders (*continued*)
congenital diaphragmatic hernia (CDH), 215–218
cystic fibrosis, 228–229
emphysema, 219–220
lobar, 220
esophageal duplication cyst, 235
lung cysts, 220–221
pectus deformity, 232–234
pneumonia or pneumonitis, 221–223
pneumothorax, 230–231
pulmonary sequestration, 236–237
tracheomalacia, 229–230
tuberculosis, 497–501
Resuscitation, 30–31
in epiglottitis, 239
in major trauma, 50, 52
in near drowning, 48
newborn, 64–66
in shock, 39–40
Retardation, symmetric growth, 67
Retinoblastoma, 421–422
Retinoic acid, 125
adverse drug effects, 125
Rhabdomyosarcoma, 422–424
testicular, 349
vaginal, 354, 355
Rheumatic fever, acute, 206–207
Rhinitis, allergic, 509–510
Rhinoconjunctivitis, 509
Richter's hernia, 358
Rickets, 374
vitamin D-resistant, 374
X-linked hypophosphatemic, 281
Rickettsia, meningitis and, 467
Rifampin
in epiglottitis, 242
side effects, 500
in tuberculosis, 500
Roseola, 443, 492–493
Rotavirus, 106
Rothmund-Thomson syndrome, 425
Rovsing's sign, 282
Rubella (German measles), 443, 491–492
congenital, 491
Rubeola, 489–490

Sacrococcygeal teratoma, 432
Salmonella gastroenteritis, 478
Salt tablets, 153
Sarcoma
Ewing, 426
osteogenic, 424–425
Scalded skin syndrome, staphylococcal, 442
Scarlet fever, 442
Schilling test, 166
School phobia, 129–130
School physical, 119–122 (*see also under* Physical examination)
Sclerosis, tuberous, 516–517
Screening, 116–117
as school physical, 120
Seborrheic dermatitis, 436
Seizures, 43–46, 396–397
common pathologic causes, 45*t*
febrile, 45
in head trauma, 43
International Classification of Epileptic, 44*t*
motor vs. nonmotor, 43
neonatal, 83
overdose-related, 45
partial complex, 396
physiologic results, 44
sports participation and, 149
tonic-clonic, 396
treatment, 45
Seminoma, 349
Sensory impairment, development and, 59
Sepsis, neonatal, 101–102
Septal defect
atrial, 190–191
ventricular, 191–192
Septo-optic dysplasia, 381
Sequence, genetic, 512
Sequestration, pulmonary, 236–237
Sertoli-Leydig cell tumors, ovarian, 349, 352
Serum amylase test, 312
Serum bilirubin test, 312
Sex cord stromal tumors, ovarian, 349
Sexual development, disorders of, 392–395
Sexually transmitted diseases (STDs), 159–160
chancroid, 486
chlamydia, 485–486
Gardnerella infection, 488
genital herpes, 486–487
gonorrhea, 483

WhiteKnightLove

'It is more blessed to give than to receive.

HIV, 494–496
human papillomavirus, 487
pelvic inflammatory disease (PID), 489
syphilis, 484–485
trichomoniasis, 488
Shimada classification, of neuroblastoma, 409, 409*t*
Shock, 35
cardiogenic, 36
compensated, 35, 36
complications of treatment, 40
in congenital heart anomalies, 38
definition, 35
diagnosis, 39
differential diagosis, 35, 37
distributive, 36
hypovolemic, 35
treatment, 39–40
types of, 35, 36
uncompensated, 35, 37
Short-gut (short-bowel) syndrome, 252–254, 256
Short stature, 127
Shunt, 189
Blalock-Tausig, 189
portosytemic, 325
Shwachman-Diamond syndrome, 173, 265
Sickle α- thalassemia, 169, 170
Sickle β-thalassemia, 169, 170
Sickle cell disease, 169–171
dactylitis and, 482
screening for, 116
Sickle disease, 169
Sickle SC disease, 169
Sickle thalassemia, 169
Sickle trait, 169
SIDS, 104–105
Sign
Brudzinski, 103, 462
Chvostek, 372
double-bubble, 297
Kernig, 103, 462
Prehn's, 347
psoas, 282
Rovsing's, 282
sunburst, 425
Trousseau, 372
Sinus tachycardia, 211
Skin disorders, 449–463
acne, 125–126
atopic dermatitis (eczema), 436–437
cellulitis, 439–441
congenital melanocytic nevi, 448
felon, 459–460
hemangioma, 452–453
hypo- and hyperpigmentation, 447–450
impetigo, 439
lymphangioma, 453–454
melanoma, 433–434
mongolian spots, 71, 447
Proteus syndrome, 456
rashes, 443
Sturge-Weber syndrome, 454–455
tinea versicolor, 446
Skull fracture, 41–43
Sliding hernia, 358
Small-for-gestational age (SGA) infants, meconium aspiration syndrome in, 97
Smoking, 152
guidance for parents, 117, 119
SOAP notes, 2
Social developmental milestones, 60
Soft tissue necrosis, 10
Speech evaluation, 121
Spherocytosis, 172–173
Spina bifida occulta, 403
Spinal dimple, 74
Spinal muscular atrophy (SMA), 404
Spondylitis (Pott disease), 499
Spontaneous intestinal perforation (SIP), 257
Sports drinks, 153, 154
Sports medicine, 149–177
age for participation, 134
eye injuries, 145
general issues in, 133–134
head injuries, 142–145
medical issues related to, 145–154
heat injury, 152–154
infectious disease, 145–147
nutrition, 150–152
steroids, drugs, and substances, 152
musculoskeletal injury, 138
preparticipation evaluation, 134–137
cardiovascular fitness, 135
general health, fitness, and nutrition, 136
laboratory tests, 136
potential recommendations, 137

WhiteKnightLove

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Sports medicine (*continued*)
pulmonary fitness, 136
Tanner stages, 133, 135
Spots
café au lait, 448
mongolian, 71, 447
Sprain, 138
Sputum culture, in pneumonia, 222
SSPE (subacute sclerosing panencephalitis), 490
Stain, Hansel, 510
Staphylococcal scalded skin syndrome, 442
Stenosis
aortic, 195–196
pulmonary, 196–197
pyloric, 304
subglottic, 247–248
Stingers, 145
Stomatitis, aphthous, 475
Stones (calculi) (*see* Gallstones; Kidney stones)
Strabismus, testing for, 121
Strain, 138
Streptococcal pharyngitis, 474–475
Streptococcus sp., in neonatal pneumonia, 100, 101
Stresses, family, 119
Stricture, bowel, 256
Stroke, heat, 152
Sturge-Weber syndrome, 454–455
Stuttering, 121
Subacute sclerosing panencephalitis (SSPE), 490
Subdural hematoma, 41
Subglottic stenosis, 247–248
Substance abuse, 152
Subungual hematoma, 459
Sudden infant death syndrome (SIDS), 105
Suicide, adolescents and, 161–162
Suicide attempts, as "acting out," 162
Sunburst sign, 425
Supplements
high-protein, 151
potassium, 153
vitamin, 26, 151
Suprahepatic portal hypertension, 323
Suprapubic puncture, 13, 480
Symmetric growth retardation, 67
Syncope, heat, 152
Syndactyly, 74
Syphilis, 484–485

Tachycardia
sinus, 211
toxic agents causing, 53
Tachypnea
toxic agents causing, 53
transient of newborn, 96
Tanner stages, 155
Tape test, 504
TAR syndrome, 179
Teeth, eruption of first, 58
Tension headache, 132
Teratoma, 431–432
ovarian, 350
sacrococcygeal, 432
treatment, 349
Terms, common abbreviations, 3–8
Test(s)
alkaline phosphatase, 312
α-fetoprotein, 348
carcinoembryonic antigen (CEA), 351
CD4 counts, 496
creatinine clearance, 330–331
Denver Developmental Screening Test, 61, 115
ELISA, 319
fluorescent treponemal antibody absorption (FTA-ABS), 485
glutamyl transpeptidase, 312
glycosylated hemoglobin (Hb1c), 365
Hansel stain, 510
hearing, 120
hepatitis B surface antigen (HBsAg), 316, *317*
HIV DNA polymerase chain reaction, 495
Mantoux tuberculin, 498
metaiodobenzylguanine (MIBG), 407
rapid plasma reagin (RPR), 484
Schilling, 166
screening, 116–117, 127
serum amylase, 312
serum bilirubin, 312
tape for pinworms, 504
thyroid function, 249, 383
Venereal Disease Research Laboratory (VDRL), 484
vision test, 121
water-deprivation, 339

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'It is more blessed to give than to receive.

Testicles, undescended (cryptorchidism), 344–345
Testicular cancer, 345
Testicular torsion, 346–347
Testicular tumors, 347–349
  germ cell, 347
  gonadal stromal (non-germ cell), 347, 349
  rhabdomyosarcoma, 347, 349
  seminoma, 349
  yolk sac, 348
Tetralogy of Fallot, 198–199
Thalassemia, 167–168
Thal intermedia, 168
Thal major, 168
Thal trait, 167, 168
Thelarche, premature, 376
Theophylline overdose, 227
Thrombocytopenia, 28, 205–206
  heparin-induced, 186
  in meningitis, 464
  neonatal alloimmune, 186–187
Thrombosis, portal vein, 324
Thrush, 473–475
Thyroglossal duct remnant, 248–249
Thyroid disorders, 397–402
  hyperthyroidism, 383–386
  hypothyroidism, 386–388
Thyroid function tests, 249, 383
Thyroiditis, Hashimoto's, 388
Thyroid storm, 384
Tinea capitis, 445
Tinea versicolor, 446
Tissue aspiration, in cellulitis, 439
Tobacco products (*see also* Smoking)
  guidance for parents, 119
Tobacco use, 152
Toenail, ingrown, 458–459
Torsion, testicular, 346–347
Torticollis, 249–250
Total anomalous pulmonary venous return, 201–202
Total parenteral nutrition (TPN), 27–29
Total proctocolectomy, in ulcerative colitis, 259
Toxic megacolon, 258
Toxic shock syndrome, 442–443
Toxocariasis, 503–504
Tracheoesophageal fistula, 300–303, *301*
Transcutaneous oxygen, 110
Transient neonatal hypocalcemia, 373
Transient tachypnea of newborn (TTN), 96
Transplantation, bone marrow, in aplastic anemia, 174
Transposition of great vessels, 38, 72
Trauma, 50–53
  head, 41–43
Traumatic injury, 138
Traumatic lumbar puncture, 12
Treponema pallidum, 484–485
Trichomonas vaginalis, 483, 488
Trichomoniasis, 488
Trichotillomania, 460
Trichuris trichuria, 505
Tricuspid atresia, 204–205
Tripod position, 240
Trisomy 13, 513, 514
Trisomy 18, 514
Trisomy 21 (*see* Down syndrome)
Trousseau sign, 372
Tuberculosis, 497–501
Tuberous sclerosis, 516, 517
Tubes
  endotracheal (ETT), for infants, 65
  tympanostomy, 473
Tumor lysis syndrome, 418
Tumors (*see also* Neoplastic disorders)
  lymphangioma, 453–454
  ovarian, 349–353
  paragangliomas, 391
  pheochromocytoma, 391–392
  testicular, 347–349
  vaginal, 354–355
Turner syndrome, 380, 515
21-Hydroxylase deficiency, 393
22q11 deletion syndrome, 373
Tympanocentesis, 473
Tympanostomy tubes, 473
Tzanck prep, 475

Ulcerative colitis, 257–260
Ulcers
  peptic, 270–272
Umbilical cord
  abnormalities, 106–107
    omphalitis, 106–107
    persistent urachal remnant, 107–108
    umbilical granuloma, 107
  catheterization, 66
  vessels normal to, 73
Umbilical hernia, 107–108, 359–360

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Uncompensated shock, 35, 37
Undescended testicles (cryptorchidism), 345
Universal precautions, 146
Urachal remnant, persistent, 107–108
Urinalysis, screening, 117
Urinary tract infection (UTI), 480–481
Urogenital disorders, ambiguous genitalia, 392–395
Uterine bleeding, dysfunctional, 159

Vaccination
  bacille Calmette-Guerin (BCG), 501
  pertussis, 502
Vaccine (*see also* Immunization)
  hepatitis B, 318
VACTERL association, 301–302
Vaginal anomalies, 353–354
Vaginal atresia, 353
Vaginal tumors, 354–355
Varicella, 493–494
  chicken pox, 493
  vaccine, 493
  varicella-zoster immune globulin (VZIG), 493
Vasoocclusive crisis, in sickle cell disease, 171
Vegetarian diets, 26
Venereal Disease Research Laboratory (VDRL) test, 484
Venipuncture, 9–11
Ventricular arrhythmias, 212
Ventricular septal defect, 191–192
Vernal conjunctivitis, 469
Vernix caseosa, 71
Vesicles
  penile herpetic, 487
  skin in newborn, 83
Vesicoureteral reflux (VUR), 335–336, 481
Viral gastroenteritis, 477
Viral meningitis, 467
Viral syndromes, 496–503
  fifth disease, 492
  measles (rubeola), 489–490
  mumps, 490
  roseola, 492–493
  rubella (german measles), 491–492
  varicella, 493–494
Virus, respiratory syncytial, 242
Visceral larva migrans (toxocariasis), 503–504
Visual acuity, 121
Visual tests, at school physical, 121
Vitamin B_{12}, 26
Vitamin B_{12} deficiency
  pernicious anemia, 165–166
Vitamin D-resistant rickets, 338
Vitamin K, in newborn, 25, 76
Vitamins
  fat-soluble, 25
  in TPN, 28
  supplementation, 26
    sports participation and, 151
Vitiligo, 449
Vocal cord paralysis, 246
Voiding cystourethrogram (VCUG), 335, 481
Volvulus, 296
Vomiting, causes of, 124
von Hippel-Lindau disease, 392
von Recklinghausen disease (neurofibromatosis type 1), 392, 516
von Willebrand disease, 184–185

Waardenburg syndrome, 456
  Hirschsprung disease and, 293–294
WAGR syndrome, 411
Warts, genital (condylomata acuminata), 487
Water-deprivation test, 339
WBC, in sepsis, 102
Web, laryngeal, 246
Weight lifting vs. weight training, 133
Weight loss, rapid unexplained, 151
Well-child visit, 113–119 (*see also under* Physical examination)
Werdnig-Hoffman disease, 404
Werner syndrome, 422
Wheezing, 123
Whipworm, 505
Whooping cough (pertussis), 501–502
WHO rehydration guidelines, 478
Wiskott-Aldrich syndrome, 179

X-linked hydrocephalus, 404
X-linked hypophosphatemic rickets, 281

Yolk sac tumors, testicular, 348

Zidovudine, 495
Zoster, 494

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'It is more blessed to give than to receive.

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Freely you have received; freely give.