

FULL-LENGTH PRACTICE TEST 16

Practice Test 16: Pediatric Medicine

70 Questions — Recommended Time: 70 Minutes

1. A 3-week-old male infant is brought to the emergency department with projectile, non-bilious vomiting after every feeding for the past 5 days. The vomiting has been progressively worsening. He is hungry immediately after vomiting. Physical examination reveals a dehydrated infant with a palpable firm, olive-shaped mass in the right upper quadrant. Laboratory studies reveal a hypochloremic, hypokalemic metabolic alkalosis. Which of the following is the most likely diagnosis?

- A. Malrotation with midgut volvulus
- B. Gastroesophageal reflux disease
- C. Pyloric stenosis
- D. Duodenal atresia

2. A 2-day-old full-term male infant develops bilious (green) vomiting. Abdominal X-ray reveals a "double bubble" sign with air in the stomach and proximal duodenum and no distal bowel gas. The infant was born with Down syndrome (trisomy 21). Which of the following is the most likely diagnosis?

- A. Duodenal atresia
- B. Pyloric stenosis
- C. Necrotizing enterocolitis
- D. Hirschsprung disease

3. A 6-month-old infant is brought to the pediatrician for a well-child visit. The mother reports that the infant can sit with support, reaches for objects, transfers objects from one hand to the other, and babbles. The infant does not yet sit independently or crawl. Which of the following best describes this infant's developmental status?

- A. Significantly delayed — requires immediate evaluation
- B. Advanced for age
- C. Mildly delayed — warrants close monitoring
- D. Appropriate developmental milestones for a 6-month-old

4. A 4-year-old boy presents with a 2-week history of periorbital edema, abdominal swelling, and decreased urine output. Laboratory studies reveal serum albumin 1.5 g/dL, total cholesterol 410 mg/dL, and urinalysis showing 4+ protein with no hematuria. Serum creatinine is normal. Complement levels (C3, C4) are normal. Which of the following is the most likely diagnosis and expected response to treatment?

- A. Post-streptococcal glomerulonephritis — treated with antibiotics
- B. Minimal change disease — expected to respond to corticosteroids
- C. Focal segmental glomerulosclerosis — steroid resistant
- D. IgA nephropathy — treated with ACE inhibitors

5. A 2-month-old infant is brought to the emergency department with a rectal temperature of 100.6°F (38.1°C). The infant appears well but irritable. He was born at term without complications. Which of the following is the most appropriate management approach?

- A. Full sepsis workup (blood culture, urinalysis/culture, lumbar puncture, CBC) with empiric IV antibiotics pending results
- B. Reassurance and discharge home with follow-up in 24 hours
- C. Oral amoxicillin and outpatient follow-up in 48 hours
- D. Rectal acetaminophen and observation in the waiting room for 4 hours

6. A 5-year-old boy presents with a sudden onset of high fever (104°F), severe sore throat, drooling, muffled voice, and sitting in a tripod position with neck hyperextended. He appears toxic and has inspiratory stridor. He has not received routine childhood vaccinations. Which of the following is the most likely diagnosis?

- A. Croup (laryngotracheobronchitis)
- B. Peritonsillar abscess
- C. Acute epiglottitis
- D. Bacterial tracheitis

7. An 18-month-old child presents with a barking cough, inspiratory stridor, hoarseness, and low-grade fever following a 2-day upper respiratory infection. Symptoms are worse at night. An anteroposterior neck X-ray reveals subglottic narrowing ("steeple sign"). The child has mild retractions but is not in respiratory distress and maintains good oxygen saturation. Which of the following is the most appropriate treatment?

- A. Immediate intubation
- B. Single dose of oral or intramuscular dexamethasone
- C. IV antibiotics for suspected bacterial infection
- D. Nebulized racemic epinephrine alone without corticosteroids

8. A 3-month-old former premature infant (born at 28 weeks) presents with wheezing, tachypnea, nasal flaring, intercostal retractions, and decreased oral intake for 2 days. He initially had rhinorrhea and low-grade fever. Chest X-ray reveals bilateral hyperinflation with peribronchial thickening and patchy atelectasis. Rapid antigen testing of nasal secretions is positive for respiratory syncytial virus (RSV). Which of the following is the most likely diagnosis?

- A. Pneumonia
- B. Asthma exacerbation
- C. Foreign body aspiration
- D. Bronchiolitis

9. A 7-year-old boy is brought to the clinic because his mother noticed he has been drinking excessively, urinating frequently, and has lost 8 pounds over the past 3 weeks despite increased appetite. Random blood glucose is 385 mg/dL. Urinalysis reveals glucosuria and ketonuria. HbA1c is 11.2%. Which of the following is the most likely diagnosis?

- A. Type 1 diabetes mellitus
- B. Type 2 diabetes mellitus
- C. Diabetes insipidus
- D. Psychogenic polydipsia

10. A 10-year-old girl presents with fatigue, pallor, and easy bruising. Her mother reports frequent nosebleeds over the past month. Physical examination reveals petechiae, hepatosplenomegaly, and cervical lymphadenopathy. CBC reveals WBC 52,000/ μ L with 75% blasts, hemoglobin 6.5 g/dL, and platelet count 18,000/ μ L. Which of the following is the most likely diagnosis?

- A. Iron deficiency anemia
- B. Acute lymphoblastic leukemia (ALL)
- C. Immune thrombocytopenic purpura
- D. Aplastic anemia

11. A 4-day-old breastfed infant presents with jaundice. Total serum bilirubin is 18 mg/dL (predominantly unconjugated/indirect). The infant is feeding well, has adequate stool and urine output, and is otherwise healthy. Direct bilirubin is 0.5 mg/dL. Blood type is O-positive, and maternal blood type is O-positive. DAT (Coombs test) is negative. Which of the following is the most appropriate initial management?

- A. Exchange transfusion immediately
- B. Reassurance only — this is pathologic jaundice not requiring treatment
- C. Oral phenobarbital
- D. Phototherapy

12. A 14-year-old boy presents with a limp and dull ache in the left knee. He is tall for his age and very athletic, playing basketball daily. Physical examination reveals tenderness and a firm, non-tender mass at the distal femoral metaphysis. X-ray reveals a lytic and sclerotic lesion in the distal femur with periosteal elevation (Codman triangle) and a "sunburst" pattern of periosteal reaction. Which of the following is the most likely diagnosis?

- A. Ewing sarcoma
- B. Osteochondroma
- C. Osteosarcoma
- D. Growing pains

13. A 3-year-old unvaccinated child presents with a 3-day history of high fever, cough, coryza, and conjunctivitis. On examination, small white-blue spots on an erythematous base are noted on the buccal mucosa opposite the molars. A maculopapular rash is beginning to appear on the face and behind the ears, spreading downward. Which of the following is the most likely diagnosis?

- A. Measles (rubeola)
- B. Rubella (German measles)
- C. Roseola infantum
- D. Scarlet fever

14. A 6-year-old boy presents with the sudden onset of cola-colored urine, periorbital edema, and blood pressure of 130/85 mmHg. His mother reports he had impetigo on his legs 3 weeks ago. Urinalysis reveals dysmorphic red blood cells, red blood cell casts, and proteinuria. Serum C3 complement is low, and ASO titer is elevated. Which of the following is the most likely diagnosis?

- A. IgA nephropathy
- B. Post-streptococcal glomerulonephritis
- C. Minimal change disease
- D. Hemolytic uremic syndrome

15. A 2-year-old child presents with a palpable abdominal mass crossing the midline, discovered by the parent during bathing. The child has periorbital ecchymosis (raccoon eyes), and physical examination reveals a firm, irregular, non-tender abdominal mass. Urine catecholamines (homovanillic acid and vanillylmandelic acid) are markedly elevated. CT reveals a suprarenal mass with calcifications encasing the aorta and crossing the midline. Which of the following is the most likely diagnosis?

- A. Wilms tumor (nephroblastoma)
- B. Hepatoblastoma
- C. Rhabdomyosarcoma
- D. Neuroblastoma

16. A 3-year-old girl presents with a large, smooth, firm abdominal mass confined to the right flank. The mass does not cross the midline. Physical examination reveals hypertension (blood pressure 125/80 mmHg). She has no periorbital ecchymosis. Urinalysis reveals microscopic hematuria. CT reveals a well-circumscribed intrarenal mass with a distinct pseudocapsule compressing but not invading surrounding structures. Which of the following is the most likely diagnosis?

- A. Neuroblastoma
- B. Polycystic kidney disease
- C. Wilms tumor (nephroblastoma)
- D. Renal cell carcinoma

17. A 12-month-old infant presents with paroxysmal episodes of severe, colicky abdominal pain with drawing up of the legs, followed by periods of lethargy and pallor. The parent reports "currant jelly" stool (mixture of blood and mucus). Physical examination reveals a sausage-shaped mass in the right upper quadrant. Abdominal ultrasound reveals a "target sign" (concentric rings of bowel within bowel). Which of the following is the most likely diagnosis?

- A. Intussusception
- B. Meckel diverticulum
- C. Pyloric stenosis

D. Malrotation with volvulus

18. A 10-year-old boy presents with recurrent episodes of wheezing, coughing (especially at night and with exercise), chest tightness, and shortness of breath. Symptoms occur 4 days per week and wake him from sleep twice per month. Pulmonary function testing reveals FEV1 of 78% predicted with a 15% improvement after albuterol administration. He uses his rescue inhaler 3-4 times per week. Which of the following is the most appropriate classification of his asthma?

A. Intermittent asthma

B. Mild persistent asthma

C. Severe persistent asthma

D. Moderate persistent asthma

19. A 5-year-old girl presents with a widespread rash for 2 days. Physical examination reveals crops of lesions in different stages — erythematous macules, papules, vesicles, and crusted lesions — distributed predominantly on the trunk, face, and scalp. She has low-grade fever and mild pruritus. She has not received the varicella vaccine. Which of the following is the most likely diagnosis?

A. Hand-foot-and-mouth disease

B. Varicella (chickenpox)

C. Impetigo

D. Scabies

20. A 6-week-old infant presents with progressive tachypnea, a staccato cough, and bilateral conjunctivitis. He was born via vaginal delivery. The mother had inadequate prenatal care and was not screened for sexually transmitted infections. CBC reveals eosinophilia. Chest X-ray reveals bilateral diffuse interstitial infiltrates with hyperinflation. Which of the following is the most likely causative organism?

A. Respiratory syncytial virus

B. Streptococcus pneumoniae

- C. Bordetella pertussis
- D. Chlamydia trachomatis

21. A 4-year-old boy presents with high fever (104°F) for 6 days, bilateral non-exudative conjunctivitis, strawberry tongue, cracked erythematous lips, erythema and edema of the hands and feet, a polymorphous truncal rash, and right anterior cervical lymphadenopathy (2.5 cm node). Which of the following is the most serious potential complication of this condition?

- A. Coronary artery aneurysms
- B. Rheumatic heart disease
- C. Glomerulonephritis
- D. Encephalitis

22. A 7-year-old girl presents with a pruritic rash in the antecubital fossae and behind the knees bilaterally. Physical examination reveals erythematous, dry, lichenified, scaly patches in the flexural areas. She has a history of seasonal allergies and her father has asthma. Which of the following is the most likely diagnosis?

- A. Contact dermatitis
- B. Psoriasis
- C. Atopic dermatitis (eczema)
- D. Tinea corporis

23. A newborn male is noted to have bilious vomiting within the first 24 hours of life. Abdominal X-ray shows a paucity of distal bowel gas. Upper GI series reveals the duodenojejunal junction (ligament of Treitz) positioned to the right of the midline with a "corkscrew" appearance of the small bowel. Which of the following is the most likely diagnosis?

- A. Duodenal atresia
- B. Malrotation with midgut volvulus
- C. Pyloric stenosis

D. Necrotizing enterocolitis

24. A 15-year-old boy presents with acute onset of right testicular pain and swelling for 3 hours. Physical examination reveals a high-riding right testicle with a horizontal (transverse) lie, absent cremasteric reflex, and severe tenderness. Doppler ultrasound shows absent blood flow to the right testicle. Which of the following is the most appropriate management?

A. Oral antibiotics for epididymitis

B. NSAIDs and scrotal elevation

C. Urgent Doppler ultrasound before any intervention (already done)

D. Emergent surgical exploration with bilateral orchiopexy

25. An 8-month-old infant presents with a brief episode of stiffening of the body followed by a sudden flexion of the trunk and extremities ("jackknife" movements) occurring in clusters upon awakening. The episodes last 1-2 seconds each and occur in groups of 10-20. EEG reveals hypsarrhythmia (chaotic, high-amplitude, disorganized background). Which of the following is the most likely diagnosis?

A. Infantile spasms (West syndrome)

B. Febrile seizures

C. Absence seizures

D. Benign myoclonus of infancy

26. A 6-year-old girl presents with her second episode of a generalized tonic-clonic seizure lasting 3 minutes. She has no fever or identifiable trigger. After the seizure, she is postictal with confusion for 15 minutes before returning to her normal baseline. Neurologic examination is normal. EEG reveals generalized spike-and-wave discharges. Brain MRI is normal. Which of the following is the most appropriate initial treatment?

- A. No treatment needed — single seizure only
- B. Phenobarbital
- C. A broad-spectrum antiepileptic drug (levetiracetam or valproic acid)
- D. Surgical evaluation

27. A 5-year-old girl presents with a sore throat, fever of 102°F, and a diffuse, fine, sandpaper-like erythematous rash that is most prominent in the skin folds (axillae, groin) with a circumoral pallor. Examination of the pharynx reveals tonsillar exudates and palatal petechiae. The tongue initially has a white coating that desquamates to reveal a red "strawberry tongue." Which of the following is the most likely diagnosis?

- A. Measles
- B. Scarlet fever (Group A Streptococcus)
- C. Kawasaki disease
- D. Staphylococcal scalded skin syndrome

28. A 2-year-old child is brought to the emergency department after witnessed ingestion of a button battery found on the floor approximately 2 hours ago. The child is currently asymptomatic with no coughing, drooling, or respiratory distress. Which of the following is the most appropriate next step?

- A. Emergent anteroposterior and lateral chest/abdominal X-ray to localize the battery, with immediate endoscopic removal if lodged in the esophagus
- B. Observation at home with instructions to monitor stool for passage
- C. Administration of activated charcoal
- D. Reassurance that button batteries pass harmlessly through the GI tract

29. A 4-year-old boy presents with a 3-week history of easy bruising, petechiae, and a nosebleed. He is otherwise well and active. He had a viral illness 4 weeks ago. CBC reveals an isolated platelet count of $15,000/\mu\text{L}$ with normal WBC, hemoglobin, and peripheral smear (no blasts). Which of the following is the most likely diagnosis?

- A. Acute lymphoblastic leukemia
- B. Aplastic anemia
- C. Henoch-Schönlein purpura
- D. Immune thrombocytopenic purpura (ITP)

30. A 9-year-old boy presents with palpable purpura on the lower extremities and buttocks, diffuse colicky abdominal pain, arthralgia of the knees and ankles, and microscopic hematuria. He had a recent upper respiratory infection. Physical examination reveals non-thrombocytopenic purpura (platelet count is normal). Which of the following is the most likely diagnosis?

- A. Immune thrombocytopenic purpura
- B. Kawasaki disease
- C. IgA vasculitis (Henoch-Schönlein purpura)
- D. Systemic lupus erythematosus

31. A full-term newborn is noted to have failure to pass meconium within the first 48 hours of life. The infant develops abdominal distension, bilious vomiting, and refusal to feed. Rectal examination produces an "explosive" gush of stool and gas. Barium enema reveals a transition zone between a narrowed distal segment and a dilated proximal segment. Which of the following is the most likely diagnosis?

- A. Meconium ileus
- B. Hirschsprung disease
- C. Necrotizing enterocolitis
- D. Imperforate anus

32. A 3-year-old child presents with a 2-day history of bloody diarrhea, abdominal cramps, and low-grade fever. Five days ago, the family attended a barbecue where hamburgers were served. Stool culture grows *E. coli* O157:H7. Three days later, the child develops pallor, petechiae, decreased urine output, and irritability. Laboratory studies reveal hemoglobin 7.5 g/dL, platelet count 45,000/ μ L, creatinine 3.2 mg/dL, and schistocytes on peripheral smear. Which of the following is the most likely complication?

- A. Hemolytic uremic syndrome (HUS)
- B. Thrombotic thrombocytopenic purpura
- C. Disseminated intravascular coagulation
- D. Immune thrombocytopenic purpura

33. A 15-year-old girl presents with primary amenorrhea. She is short-statured (height at the 3rd percentile), has a webbed neck, low posterior hairline, widely spaced nipples (shield chest), and cubitus valgus. Physical examination reveals absent breast development and streak gonads on pelvic ultrasound. Which of the following is the most likely diagnosis?

- A. Polycystic ovary syndrome
- B. Constitutional delay of puberty
- C. Kallmann syndrome
- D. Turner syndrome (45,X)

34. A 14-year-old obese boy (BMI 32) presents with a limp and left knee pain for the past 2 weeks. He denies trauma. Physical examination reveals limited internal rotation of the left hip and obligatory external rotation when the hip is flexed. X-ray of the hip reveals posterior displacement of the femoral epiphysis relative to the metaphysis on the frog-leg lateral view. Which of the following is the most likely diagnosis?

- A. Legg-Calvé-Perthes disease
- B. Slipped capital femoral epiphysis (SCFE)
- C. Septic arthritis of the hip
- D. Osgood-Schlatter disease

35. A 4-year-old boy presents with a limp and left hip pain for 3 weeks. He is afebrile and otherwise well. X-ray of the left hip reveals flattening and fragmentation of the femoral head with increased radiodensity ("crescent sign"). He is of short stature for his age. Which of the following is the most likely diagnosis?

- A. Transient synovitis
- B. Septic arthritis
- C. Legg-Calvé-Perthes disease (avascular necrosis of the femoral head)
- D. Slipped capital femoral epiphysis

36. A 6-year-old boy presents with a 2-day history of right hip pain, limp, and refusal to bear weight. He had a viral upper respiratory infection one week ago. He is afebrile with a temperature of 99.0°F. CBC and ESR are normal. Hip ultrasound reveals a small joint effusion. He has pain with internal rotation of the hip. Which of the following is the most likely diagnosis?

- A. Slipped capital femoral epiphysis
- B. Osteomyelitis
- C. Juvenile idiopathic arthritis
- D. Transient synovitis (toxic synovitis)

37. A 4-year-old girl presents with a painful, swollen right knee for the past 6 weeks. She also has daily high-spiking fevers (quotidian pattern — one or two temperature spikes above 102.2°F daily with return to normal), a salmon-colored evanescent macular rash on the trunk and extremities that appears with fevers, hepatosplenomegaly, and generalized lymphadenopathy. Laboratory studies reveal markedly elevated ESR, CRP, ferritin, and WBC. ANA and RF are negative. Which of the following is the most likely diagnosis?

- A. Systemic juvenile idiopathic arthritis (Still disease)
- B. Acute rheumatic fever
- C. Systemic lupus erythematosus
- D. Reactive arthritis

38. A 5-year-old boy presents with a 3-week history of fever, a new-onset heart murmur (grade III/VI pansystolic murmur at the apex), and migratory polyarthritis affecting the large joints. He had a sore throat 4 weeks ago that was not treated with antibiotics. ASO titer is elevated. ECG reveals a prolonged PR interval. Which of the following is the most likely diagnosis?

- A. Infective endocarditis
- B. Kawasaki disease
- C. Acute rheumatic fever
- D. Juvenile idiopathic arthritis

39. A 4-month-old infant presents with poor feeding, diaphoresis during feeding, tachypnea, and failure to thrive. Physical examination reveals tachycardia, hepatomegaly, and a holosystolic murmur heard best at the left lower sternal border. Chest X-ray reveals cardiomegaly with increased pulmonary vascular markings. Echocardiogram reveals a large ventricular septal defect with left-to-right shunting. Which of the following is the most likely cause of this infant's symptoms?

- A. Tetralogy of Fallot
- B. Congestive heart failure from left-to-right shunt (VSD)
- C. Coarctation of the aorta
- D. Atrial septal defect

40. A 1-day-old cyanotic newborn has oxygen saturations of 65% that do not improve with supplemental oxygen (failed hyperoxia test). Chest X-ray reveals an "egg-on-a-string" appearance (narrow mediastinum with an egg-shaped cardiac silhouette). Echocardiogram reveals the aorta arising from the right ventricle and the pulmonary artery arising from the left ventricle. Which of the following is the most likely diagnosis?

- A. Tetralogy of Fallot
- B. Truncus arteriosus
- C. Total anomalous pulmonary venous return
- D. Transposition of the great arteries

41. A 2-month-old infant is brought for a well-child visit. On examination, a continuous "machinery-like" murmur is heard best in the left infraclavicular area (below the left clavicle). Bounding peripheral pulses and a widened pulse pressure are noted. The infant was born prematurely at 30 weeks gestation. Which of the following is the most likely diagnosis?

- A. Patent ductus arteriosus (PDA)
- B. Ventricular septal defect
- C. Atrial septal defect
- D. Aortic stenosis

42. A 1-week-old infant presents with poor feeding, lethargy, tachypnea, and diminished femoral pulses compared to brachial pulses. Blood pressure in the right arm is 90/60 mmHg, but blood pressure in the lower extremities is 50/30 mmHg. Chest X-ray reveals cardiomegaly. Echocardiogram reveals a discrete narrowing of the aorta just distal to the left subclavian artery at the level of the ductus arteriosus. Which of the following is the most likely diagnosis?

- A. Aortic stenosis
- B. Patent ductus arteriosus
- C. Coarctation of the aorta
- D. Interrupted aortic arch

43. A 6-month-old infant with a known history of unrepaired tetralogy of Fallot presents with sudden onset of severe cyanosis, irritability, and inconsolable crying after a prolonged episode of crying. Oxygen saturation is 55%. The infant assumes a squatting position when placed on the examination table. On auscultation, the systolic ejection murmur associated with his condition has diminished. Which of the following is the most appropriate immediate intervention?

- A. Emergent surgical repair
- B. IV antibiotics
- C. Immediate intubation

D. Knee-to-chest positioning, supplemental oxygen, IV fluid bolus, and subcutaneous morphine or IV phenylephrine

44. A 12-year-old girl presents with fatigue, exertional dyspnea, and a fixed split S2 on cardiac auscultation. She has a grade II/VI systolic ejection murmur at the left upper sternal border. ECG reveals right axis deviation and incomplete right bundle branch block. Chest X-ray reveals cardiomegaly with enlarged pulmonary arteries. Echocardiogram reveals a secundum atrial septal defect with significant left-to-right shunting (Qp:Qs ratio of 2:1). Which of the following is the most likely diagnosis?

- A. Ventricular septal defect
- B. Atrial septal defect (ASD)
- C. Patent ductus arteriosus
- D. Mitral valve prolapse

45. A 2-year-old boy presents with a painless abdominal mass noticed by his mother during a bath. Physical examination reveals a smooth, firm, non-tender left flank mass that does not cross the midline. Blood pressure is elevated at 120/78 mmHg. CBC is notable for polycythemia. Urinalysis shows microscopic hematuria. Which of the following additional findings would be most characteristic of this tumor?

- A. Association with aniridia, genitourinary anomalies, and hemihypertrophy (WAGR syndrome or Beckwith-Wiedemann syndrome)
- B. Elevated urine catecholamines (HVA and VMA)
- C. Periorbital ecchymosis and opsoclonus-myoclonus
- D. Calcifications on CT scan that encase the great vessels

46. A 6-year-old girl with sickle cell disease presents with sudden onset of severe pallor, lethargy, tachycardia, and a markedly enlarged spleen. Her hemoglobin is 3.5 g/dL (baseline 7.5 g/dL) with a reticulocyte count of 12%. She is hemodynamically unstable. Which of the following is the most likely diagnosis?

- A. Vaso-occlusive crisis
- B. Aplastic crisis
- C. Acute chest syndrome
- D. Acute splenic sequestration crisis

47. A 6-year-old boy with sickle cell disease presents with acute onset of fever, headache, and left-sided weakness with facial droop. CT scan of the head reveals an ischemic stroke in the right middle cerebral artery territory. Which of the following is the most appropriate acute treatment?

- A. IV tissue plasminogen activator (tPA)
- B. Aspirin and clopidogrel
- C. Emergent exchange transfusion (reduce HbS to less than 30%)
- D. IV heparin anticoagulation

48. A 3-year-old boy presents with unilateral painless proptosis of the right eye, periorbital ecchymosis, and a palpable orbital mass. CT scan of the orbit reveals a soft tissue mass in the right orbit. Biopsy reveals small round blue cells. Urine vanillylmandelic acid (VMA) and homovanillic acid (HVA) are markedly elevated. Which of the following is the most likely diagnosis?

- A. Retinoblastoma
- B. Neuroblastoma with orbital metastasis
- C. Rhabdomyosarcoma of the orbit
- D. Lymphoma

49. A 2-year-old child presents with a white pupillary reflex (leukocoria) in the left eye, noticed by the parents in photographs. Ophthalmologic examination reveals an intraocular mass. Family history reveals the father had a similar condition requiring eye removal as an infant. Which of the following is the most likely diagnosis?

- A. Retinoblastoma
- B. Congenital cataract
- C. *Toxocara canis* infection
- D. Coats disease

50. A 14-year-old boy presents with knee pain and a painful swelling of the proximal tibia. X-ray reveals a lytic lesion in the diaphysis of the tibia with a multilayered periosteal reaction described as "onion-skin" appearance. Biopsy reveals small round blue cells arranged in rosettes. Which of the following is the most likely diagnosis?

- A. Osteosarcoma
- B. Osteochondroma
- C. Chondrosarcoma
- D. Ewing sarcoma

51. A 2-month-old infant is brought for routine well-child visit. The mother asks about the recommended immunization schedule. Which of the following vaccines is routinely administered at the 2-month well-child visit?

- A. Measles, mumps, rubella (MMR)
- B. Varicella
- C. DTaP (diphtheria, tetanus, acellular pertussis), IPV (inactivated polio), Hib (*Haemophilus influenzae* type b), PCV13 (pneumococcal conjugate), and rotavirus
- D. Hepatitis A

52. A 3-year-old boy is brought to the emergency department by his mother who noticed bruises on his back, buttocks, and upper arms in various stages of healing. The child appears withdrawn and avoids eye contact. The mother states the bruises are from "falling down the stairs." Physical examination reveals patterned bruises consistent with a belt buckle mark, and a cigarette burn on the forearm. Which of the following is the most appropriate next step?

- A. Accept the caregiver's explanation and document findings
- B. Report suspected child abuse to child protective services, obtain full skeletal survey, and ensure the child's safety
- C. Confront the mother about the inconsistent history
- D. Discharge the child with a follow-up appointment in 1 week

53. A 4-year-old girl presents with a limp and refusal to bear weight on the right leg. She is febrile (103.5°F), appears ill, and refuses to move the right hip. The right hip is held in flexion, abduction, and external rotation. Laboratory studies reveal WBC 18,000/ μ L, ESR 65 mm/hr, and CRP 8.5 mg/dL. Hip ultrasound reveals a large joint effusion. Which of the following is the most appropriate next step?

- A. Urgent joint aspiration (arthrocentesis) of the right hip for Gram stain, cell count, and culture
- B. Oral ibuprofen and close outpatient follow-up
- C. MRI of the hip before any intervention
- D. IV antibiotics without joint aspiration

54. A 15-month-old child has a generalized tonic-clonic seizure lasting 3 minutes in the context of a rapidly rising fever to 104°F from an acute otitis media. The child has no prior history of seizures and returns to baseline neurologic function within 30 minutes. There is no family history of epilepsy. Which of the following is the most likely diagnosis?

- A. Epilepsy requiring long-term anticonvulsant therapy
- B. Meningitis
- C. Simple febrile seizure
- D. Complex febrile seizure

55. A 3-year-old child is brought to the emergency department after sudden onset of choking, coughing, and unilateral wheezing while playing with small toys. The child appears non-toxic but has decreased breath sounds on the right side. Chest X-ray reveals hyperinflation of the right lung with mediastinal shift to the left on expiratory film. Which of the following is the most likely diagnosis?

- A. Asthma exacerbation
- B. Pneumonia
- C. Croup
- D. Foreign body aspiration

56. A 6-month-old infant is brought to the clinic with a flat, erythematous, well-demarcated lesion on the left side of the forehead that has been present since birth. The lesion does not change in size with crying. The mother reports it has not grown. Which of the following is the most likely diagnosis?

- A. Infantile hemangioma
- B. Port-wine stain (nevus flammeus)
- C. Capillary hemangioma (strawberry hemangioma)
- D. Mongolian spot

57. A 3-day-old full-term infant is found to be jaundiced with a total bilirubin of 15 mg/dL. The mother's blood type is O-positive and the infant's blood type is A-positive. Direct Coombs test (DAT) is positive. Peripheral smear reveals spherocytes and reticulocytosis. Which of the following is the most likely cause of this infant's jaundice?

- A. ABO hemolytic disease of the newborn
- B. Physiologic jaundice
- C. Breast milk jaundice
- D. Gilbert syndrome

58. A 7-year-old boy presents with recurrent headaches, vomiting (especially in the morning), unsteady gait, and progressive clumsiness over the past month. Neurologic examination reveals truncal ataxia, bilateral papilledema, and a positive Romberg sign. MRI of the brain reveals a posterior fossa mass arising from the cerebellar vermis with obstructive hydrocephalus. Which of the following is the most likely diagnosis?

- A. Craniopharyngioma
- B. Brainstem glioma
- C. Medulloblastoma (or pilocytic astrocytoma of the cerebellum)
- D. Supratentorial ependymoma

59. A 5-year-old boy presents with progressive proximal muscle weakness — he has difficulty climbing stairs, rising from the floor using the Gowers maneuver (walking hands up his thighs), and waddles when walking. Physical examination reveals calf pseudohypertrophy. CK is markedly elevated at 15,000 U/L. His maternal uncle was wheelchair-bound by age 12. Which of the following is the most likely diagnosis?

- A. Becker muscular dystrophy
- B. Myasthenia gravis
- C. Polymyositis
- D. Duchenne muscular dystrophy

60. A 10-day-old infant born at home to a mother with no prenatal care presents with poor feeding, irritability, and a bulging anterior fontanelle. Temperature is 100.8°F. Lumbar puncture reveals CSF with elevated WBCs (predominantly neutrophils), elevated protein, low glucose, and Gram stain showing gram-positive rods. Which of the following is the most likely causative organism?

- A. *Escherichia coli*
- B. *Listeria monocytogenes*
- C. *Neisseria meningitidis*
- D. *Streptococcus pneumoniae*

61. A 4-year-old previously healthy boy presents with sudden onset of limping, irritability, and refusal to move his right arm. The parents report no history of trauma. On examination, there is tenderness, warmth, and swelling over the distal right femur. He has a temperature of 102°F. WBC is 15,000/ μ L, ESR is 55 mm/hr, CRP is elevated, and blood culture is pending. MRI of the femur reveals a metaphyseal bone abscess with periosteal elevation and surrounding soft tissue edema. Which of the following is the most likely diagnosis?

- A. Acute hematogenous osteomyelitis
- B. Ewing sarcoma
- C. Osteosarcoma
- D. Transient synovitis

62. A 2-year-old child presents with a 5-day history of fever, bilateral cervical lymphadenopathy, conjunctivitis, oral mucositis, and a diffuse erythematous rash. The child meets criteria for Kawasaki disease. Echocardiogram is normal. Which of the following is the most appropriate treatment to reduce the risk of coronary artery aneurysms?

- A. Oral penicillin V for 10 days
- B. High-dose oral corticosteroids alone
- C. Intravenous immunoglobulin (IVIG) plus high-dose aspirin
- D. IV antibiotics for 14 days

63. A 16-year-old boy presents with progressive fatigue, exercise intolerance, and a grade III/VI crescendo-decrescendo systolic ejection murmur heard best at the right upper sternal border with radiation to the carotid arteries. The murmur is preceded by a systolic ejection click. He has a history of a bicuspid aortic valve diagnosed in childhood. Which of the following is the most likely diagnosis?

- A. Mitral regurgitation
- B. Aortic stenosis (from bicuspid aortic valve)
- C. Hypertrophic cardiomyopathy
- D. Pulmonary stenosis

64. A 10-year-old girl is brought to the emergency department after accidental ingestion of approximately 20 tablets of her mother's acetaminophen (500 mg each) approximately 4 hours ago. She currently feels nauseated but has no other symptoms. Serum acetaminophen level is plotted on the Rumack-Matthew nomogram and falls above the treatment line. Which of the following is the most appropriate treatment?

- A. Activated charcoal alone as definitive treatment
- B. Observation without treatment since she is asymptomatic
- C. Immediate liver transplant evaluation
- D. N-acetylcysteine (NAC)

65. A 3-year-old boy presents with painless rectal bleeding with bright red blood mixed with stool. The bleeding is intermittent and self-limited. He is otherwise healthy with normal growth and development. Physical examination and CBC are normal. A Meckel scan (technetium-99m pertechnetate scan) reveals abnormal uptake in the right lower quadrant. Which of the following is the most likely diagnosis?

- A. Meckel diverticulum
- B. Juvenile polyp
- C. Intussusception
- D. Anal fissure

66. A 7-day-old breast-fed infant presents with lethargy, poor feeding, vomiting, jaundice, and hepatomegaly. Newborn screening results are pending. Urine shows reducing substances (positive Clinitest) but negative glucose oxidase test. Escherichia coli sepsis is identified on blood culture. Which of the following is the most likely diagnosis?

- A. Maple syrup urine disease
- B. Galactosemia
- C. Hereditary fructose intolerance
- D. Phenylketonuria

67. A 15-year-old girl presents with a 6-month history of progressive weight loss (20 pounds), amenorrhea, excessive exercising, and self-induced restriction of food intake. She expresses intense fear of gaining weight despite a BMI of 16.5 (severely underweight). She has dry skin, lanugo hair on her face and arms, and bradycardia (heart rate 48 bpm). Which of the following is the most likely diagnosis?

- A. Bulimia nervosa
- B. Major depressive disorder
- C. Anorexia nervosa
- D. Hyperthyroidism

68. A 4-year-old boy presents with a new onset of periorbital and scrotal edema. Urinalysis reveals 4+ proteinuria with no hematuria. His 24-hour urine protein is 55 mg/kg/day (nephrotic range). Serum albumin is 1.8 g/dL. Complement levels are normal. He is started on prednisone 2 mg/kg/day. After 4 weeks of daily corticosteroid therapy, his proteinuria completely resolves. Which of the following best characterizes the expected clinical course?

- A. Progression to end-stage renal disease within 5 years
- B. Complete cure with no risk of relapse
- C. Steroid resistance requiring cyclophosphamide
- D. Steroid-responsive nephrotic syndrome with approximately 50-75% chance of relapse, but excellent long-term renal prognosis

69. A 14-month-old child presents to the emergency department after the parents witnessed the child put a small coin in their mouth followed by gagging and drooling. The child now has persistent drooling, dysphagia, and refuses to eat. There is no stridor or respiratory distress. Chest X-ray reveals a round radiopaque foreign body at the level of the cricopharyngeus muscle (upper esophagus). Which of the following is the most appropriate management?

- A. Emergent endoscopic removal of the esophageal foreign body
- B. Observation for 24 hours to allow spontaneous passage into the stomach
- C. Oral laxative to facilitate passage
- D. Barium swallow as the next diagnostic step

70. A full-term newborn delivered via cesarean section develops progressive respiratory distress within the first few hours of life. The infant has tachypnea (respiratory rate 80), mild grunting, and mild retractions. Oxygen saturation is 92% on room air and improves to 98% with supplemental oxygen. Chest X-ray reveals prominent perihilar streaking, fluid in the interlobar fissures, and hyperinflation. The symptoms gradually improve over 24-48 hours with supportive care. Which of the following is the most likely diagnosis?

- A. Respiratory distress syndrome (hyaline membrane disease)
- B. Transient tachypnea of the newborn
- C. Meconium aspiration syndrome
- D. Pneumothorax

PRACTICE TEST 16: ANSWER KEY

WITH EXPLANATIONS

Pediatric Medicine

1. C. Pyloric stenosis. Hypertrophic pyloric stenosis is caused by progressive hypertrophy and hyperplasia of the pyloric smooth muscle, producing gastric outlet obstruction. It typically presents at 2-6 weeks of age (classically in firstborn males) with progressive, projectile, non-bilious vomiting — the non-bilious nature is critical because the obstruction is proximal to the ampulla of Vater. The infant remains hungry after vomiting ("hungry vomiter"). The palpable olive-shaped mass in the right upper quadrant (hypertrophied pylorus) is pathognomonic but found in only approximately 70-80% of cases. Hypochloremic, hypokalemic metabolic alkalosis results from loss of hydrochloric acid in vomitus. Ultrasound is the diagnostic study of choice, revealing pyloric muscle thickness greater than 3 mm and pyloric channel length greater than 15 mm. Treatment is surgical pyloromyotomy (Ramstedt procedure) after fluid and electrolyte correction.

2. A. Duodenal atresia. Duodenal atresia is a congenital complete obstruction of the duodenal lumen resulting from failure of recanalization during fetal development. The "double bubble" sign on abdominal X-ray — two air-fluid levels representing air in the stomach and proximal duodenum with no distal bowel gas — is pathognomonic. Bilious vomiting within the first 24-48 hours of life indicates obstruction distal to the ampulla of Vater. Duodenal atresia has a strong association with Down syndrome (trisomy 21), occurring in approximately 25-30% of cases. Other associated anomalies include congenital heart defects, annular pancreas, and malrotation. Polyhydramnios is common prenatally due to impaired fetal swallowing. Treatment is surgical duodenoduodenostomy. Prognosis is excellent with timely surgical intervention.

3. D. Appropriate developmental milestones for a 6-month-old. At 6 months, expected developmental milestones include sitting with support (independent sitting typically develops at 6-7 months), reaching for and grasping objects, transferring objects between hands, and babbling (consonant-vowel combinations like "ba-ba," "da-da"). Crawling typically begins at 8-10 months, so its absence at 6 months is completely normal. Gross motor milestones follow a cephalocaudal progression — head control (3-4 months), rolling (4-5 months), sitting with support (5-6 months), sitting independently (6-7 months), crawling (8-10 months), pulling to stand (9-10 months), and walking (12-15 months). Fine motor development progresses from a palmar grasp (4-5 months) to a raking grasp (6-7 months) to a pincer grasp (9-10 months). Red flags warranting evaluation include failure to reach for objects by 5 months, inability to sit with support by 6 months, or absence of babbling by 9 months.

4. B. Minimal change disease — expected to respond to corticosteroids. Minimal change disease (MCD) is the most common cause of nephrotic syndrome in children aged 2-6 years, accounting for approximately 80% of pediatric nephrotic syndrome. The full nephrotic syndrome includes heavy proteinuria (greater than 3.5 g/day or 40 mg/m²/hour in children), hypoalbuminemia (less than 2.5 g/dL), hyperlipidemia, and edema. Normal complement levels distinguish MCD from post-streptococcal glomerulonephritis and membranoproliferative GN. Light microscopy appears normal — the only abnormality is diffuse podocyte foot process effacement on electron microscopy. MCD is highly steroid-responsive, with approximately 90% of children achieving complete remission with prednisone (2 mg/kg/day for 4-6 weeks followed by taper). Renal biopsy is not performed initially in children with classic presentation; it is reserved for steroid-resistant cases or atypical features.

5. A. Full sepsis workup with empiric IV antibiotics. Fever in an infant younger than 60 days (particularly under 28 days) is a serious concern because neonates have an immature immune system with limited ability to localize infection, and serious bacterial infections (SBI) including bacteremia, UTI, and meningitis occur in approximately 7-13% of febrile young infants. Even well-appearing infants under 28 days require a full sepsis evaluation — blood culture, urinalysis with urine culture (catheterized specimen), lumbar puncture for CSF analysis and culture, and CBC. Empiric IV antibiotics (ampicillin plus gentamicin or ampicillin plus cefotaxime) are administered pending culture results. For infants 29-60 days, risk stratification tools (Rochester criteria, Step-by-Step approach, Febrile Infant Lab Score) may guide management, but most protocols still recommend full evaluation and empiric antibiotics. This conservative approach is essential because clinical appearance alone cannot reliably exclude serious bacterial infection in this age group.

6. C. Acute epiglottitis. Acute epiglottitis is a rapidly progressive, life-threatening infection of the epiglottis and supraglottic structures, most classically caused by *Haemophilus influenzae* type b (Hib) in unvaccinated children. The classic presentation includes the "4 Ds" — Dysphagia, Drooling, Distress, and Dysphonia (muffled "hot potato" voice) — plus high fever, toxic appearance, and inspiratory stridor. The child characteristically sits in the tripod position (leaning forward with neck hyperextended and chin protruding) to maximize airway diameter. Direct visualization of the pharynx with a tongue depressor is absolutely contraindicated as it may precipitate complete airway obstruction. Lateral neck X-ray reveals the "thumbprint sign" (swollen epiglottis). Management requires immediate airway protection (controlled intubation in the operating room by the most experienced provider) followed by IV antibiotics (ceftriaxone or cefotaxime). The Hib vaccine has dramatically reduced the incidence of epiglottitis.

7. B. Single dose of oral or intramuscular dexamethasone. Croup (laryngotracheobronchitis) is the most common cause of acute upper airway obstruction in children aged 6 months to 3 years, predominantly caused by parainfluenza virus (types 1 and 2). Mild croup (barking cough, hoarseness, stridor at rest with mild retractions but no significant respiratory distress) is treated with a single dose of dexamethasone (0.15-0.6 mg/kg orally or intramuscularly). Dexamethasone reduces airway edema and inflammation, providing benefit within 2-4 hours lasting up to 72 hours. Nebulized racemic epinephrine is added for moderate-to-severe croup (significant stridor at rest, marked retractions, agitation), providing rapid but

temporary relief (lasting approximately 2 hours). The steeple sign on AP neck X-ray represents subglottic narrowing. Most cases are self-limited, resolving within 3-7 days.

8. D. Bronchiolitis. Bronchiolitis is the most common lower respiratory tract infection in infants under 2 years of age, with RSV being the most common cause (accounting for approximately 50-80% of cases). It typically occurs in winter epidemics. The clinical course begins with upper respiratory symptoms (rhinorrhea, low-grade fever) progressing to lower respiratory involvement (wheezing, tachypnea, retractions, hypoxia) over 2-3 days. Former premature infants are at highest risk for severe disease. Chest X-ray reveals hyperinflation, peribronchial thickening, and patchy atelectasis. Management is primarily supportive — supplemental oxygen for hypoxia (saturation below 90%), nasal suctioning, hydration support, and close monitoring. Bronchodilators, corticosteroids, and antibiotics are not recommended for routine bronchiolitis. Palivizumab (monoclonal antibody against RSV) is given prophylactically to high-risk infants during RSV season.

9. A. Type 1 diabetes mellitus. Type 1 diabetes is the most common form of diabetes in children and adolescents, caused by autoimmune destruction of pancreatic beta cells (mediated by T-lymphocytes) leading to absolute insulin deficiency. Classic presentation includes the triad of polyuria (osmotic diuresis from glucosuria), polydipsia (compensatory for fluid loss), and weight loss (despite polyphagia, because without insulin, glucose cannot enter cells, and the body catabolizes fat and muscle). Random blood glucose greater than 200 mg/dL with classic symptoms confirms the diagnosis. HbA1c of 11.2% reflects chronic hyperglycemia over the preceding 2-3 months. Autoantibodies (anti-GAD65, anti-insulin, anti-IA-2, anti-ZnT8) confirm autoimmune etiology. Diabetic ketoacidosis (DKA) is the initial presentation in approximately 25-40% of children with new-onset type 1 diabetes. Treatment is lifelong insulin therapy (basal-bolus regimen or insulin pump).

10. B. Acute lymphoblastic leukemia (ALL). ALL is the most common pediatric malignancy, accounting for approximately 25% of all childhood cancers, with peak incidence between ages 2-5 years. ALL results from malignant clonal proliferation of lymphoid precursors (lymphoblasts) in the bone marrow. Bone marrow replacement by blasts causes pancytopenia — anemia (pallor, fatigue), thrombocytopenia (petechiae, bruising, bleeding), and functional leukopenia (despite elevated WBC). Hepatosplenomegaly and lymphadenopathy result from extramedullary blast infiltration. Bone pain from marrow expansion is common. Peripheral blood showing greater than 20% blasts with confirmation on bone marrow biopsy (greater than 25% blasts) establishes the diagnosis. Immunophenotyping identifies B-cell precursor ALL (most common, approximately 85%). Prognosis in childhood ALL is excellent, with cure rates exceeding 85-90% with modern multi-agent chemotherapy.

11. D. Phototherapy. Neonatal jaundice is extremely common, affecting approximately 60% of term and 80% of preterm neonates. This 4-day-old breast-fed infant has unconjugated (indirect) hyperbilirubinemia at 18 mg/dL, which exceeds phototherapy thresholds based on the Bhutani nomogram (hour-specific bilirubin plotted against age in hours determines risk zone and treatment threshold). The predominantly indirect bilirubin, negative DAT, and same blood group (O+/O+) make hemolytic disease unlikely. This presentation is consistent with breastfeeding jaundice (inadequate intake leading to decreased bilirubin

elimination) versus breast milk jaundice (substances in breast milk inhibiting conjugation). Phototherapy converts unconjugated bilirubin into water-soluble photoisomers that can be excreted without hepatic conjugation. Exchange transfusion is reserved for bilirubin approaching exchange levels (typically greater than 25 mg/dL in term infants) or for rapidly rising bilirubin unresponsive to phototherapy. Breastfeeding should be continued with encouragement of frequent feedings.

12. C. Osteosarcoma. Osteosarcoma is the most common primary malignant bone tumor in children and adolescents, with peak incidence during the adolescent growth spurt (10-20 years). It most commonly affects the metaphyseal region of long bones, particularly the distal femur (most common site), proximal tibia, and proximal humerus — areas of rapid bone growth. Classic radiographic findings include a mixed lytic and sclerotic lesion, Codman triangle (periosteal elevation at the tumor margin from subperiosteal new bone formation), and a "sunburst" or "sunray" pattern (radiating spicules of reactive bone extending perpendicular to the cortex). Serum alkaline phosphatase and LDH may be elevated. Treatment is neoadjuvant chemotherapy followed by limb-salvage surgery or amputation, then adjuvant chemotherapy. Five-year survival for localized disease is approximately 65-70%.

13. A. Measles (rubeola). Measles is a highly contagious viral illness caused by the measles virus (paramyxovirus family), transmitted via respiratory droplets. The prodromal phase features the "3 Cs" — Cough, Coryza, and Conjunctivitis — plus high fever. Koplik spots (pathognomonic enanthem — small white-blue spots on an erythematous base on the buccal mucosa opposite the molars) appear 1-2 days before the rash and are the earliest clinical sign specific to measles. The exanthem is a maculopapular rash that begins on the face and behind the ears and spreads cephalocaudally (downward to the trunk and extremities). Complications include pneumonia (most common cause of death), otitis media, croup, encephalitis, and subacute sclerosing panencephalitis (SSPE — a rare, fatal late complication occurring years after infection). Measles is preventable with the MMR vaccine (first dose at 12-15 months, second dose at 4-6 years).

14. B. Post-streptococcal glomerulonephritis. PSGN is the classic post-infectious glomerulonephritis in children, occurring 1-3 weeks after streptococcal pharyngitis or 3-6 weeks after streptococcal skin infection (impetigo, as in this case). It results from glomerular deposition of immune complexes (containing streptococcal antigens and host antibodies), activating complement and triggering inflammatory damage. The nephritic presentation includes hematuria (cola-colored urine from dysmorphic RBCs), RBC casts (pathognomonic for glomerulonephritis), hypertension, edema (particularly periorbital in children), and mild-moderate proteinuria. Low C3 with normal C4 reflects alternative complement pathway activation. Elevated ASO titer confirms recent streptococcal infection. Electron microscopy reveals characteristic subepithelial "humps." Treatment is supportive — salt and fluid restriction, antihypertensives, loop diuretics for edema. Prognosis is excellent in children, with greater than 95% complete recovery.

15. D. Neuroblastoma. Neuroblastoma is the most common extracranial solid tumor in children and the most common malignancy of infancy, arising from neural crest cells of the sympathetic nervous system. Most tumors originate in the adrenal medulla (approximately 40%) or paravertebral sympathetic ganglia.

Key distinguishing features from Wilms tumor include crossing the midline (neuroblastoma encases and displaces vessels rather than displacing the kidney), calcifications on imaging (present in approximately 80-90%), periorbital ecchymosis ("raccoon eyes" from orbital metastases), and elevated urine catecholamines (HVA and VMA, elevated in greater than 90% of cases). Opsoclonus-myoclonus syndrome ("dancing eyes, dancing feet") is a paraneoplastic manifestation. Neuroblastoma demonstrates unique biology — spontaneous regression can occur in infants (stage 4S), while older children often present with advanced disease. MYCN amplification is the most important adverse prognostic factor.

16. C. Wilms tumor (nephroblastoma). Wilms tumor is the most common renal malignancy of childhood, with peak incidence between ages 3-4 years. It presents as a smooth, firm, painless abdominal mass confined to one flank that characteristically does not cross the midline (distinguishing it from neuroblastoma). Hypertension occurs in approximately 25% of cases from renin secretion by the tumor. Microscopic hematuria is found in approximately 25% of patients. CT reveals a well-circumscribed intrarenal mass with a pseudocapsule. Associated syndromes include WAGR syndrome (Wilms tumor, Aniridia, Genitourinary anomalies, intellectual disability — WT1 deletion on chromosome 11p13) and Beckwith-Wiedemann syndrome (macroglossia, omphalocele, hemihypertrophy, organomegaly — WT2 on chromosome 11p15.5). Treatment is nephrectomy followed by chemotherapy (vincristine, dactinomycin, with or without doxorubicin based on staging and histology). Prognosis is excellent with overall survival exceeding 90%.

17. A. Intussusception. Intussusception is the most common cause of intestinal obstruction in children aged 6 months to 3 years, occurring when a proximal segment of bowel (intussusceptum) telescopes into an adjacent distal segment (intussusciens). The ileocolic junction is the most common site (approximately 90%). Classic presentation includes paroxysmal colicky abdominal pain (child draws up legs and screams) alternating with periods of lethargy, "currant jelly" stool (blood and mucus from mucosal ischemia), and a palpable sausage-shaped mass in the right upper quadrant. Ultrasound reveals the pathognomonic "target sign" or "doughnut sign" (concentric rings from bowel-within-bowel). Most cases in children are idiopathic (hypertrophied Peyer patches from preceding viral illness serve as lead points). Air or hydrostatic (saline) enema is both diagnostic and therapeutic (approximately 80-95% successful reduction). Surgical intervention is reserved for failed enema reduction, perforation, or peritonitis.

18. D. Moderate persistent asthma. Asthma classification in children is based on symptom frequency, nighttime awakenings, rescue inhaler use, interference with activity, and lung function. Moderate persistent asthma is defined by daily symptoms (this child has symptoms 4 days/week), nighttime awakenings more than once per week but not nightly (this child wakes twice monthly, consistent with mild persistent or borderline moderate), rescue inhaler use daily, some activity limitation, and FEV1 60-80% predicted. This child's FEV1 of 78% with significant bronchodilator reversibility (15% improvement) places him in the moderate persistent category. Treatment is a medium-dose inhaled corticosteroid (ICS) or low-dose ICS plus a long-acting beta-2 agonist (LABA) as step 3-4 therapy per NAEPP guidelines. All asthma patients should have a rescue short-acting beta-agonist (albuterol) and an asthma action plan.

19. B. Varicella (chickenpox). Varicella is caused by varicella-zoster virus (VZV), a highly contagious herpesvirus transmitted via respiratory droplets and direct contact with vesicular fluid. The hallmark clinical feature is a generalized vesicular rash with lesions in multiple stages of development simultaneously (macules → papules → vesicles → pustules → crusts) — described as "crops" of lesions in varying stages. The rash begins on the trunk and face and spreads centrifugally. Individual vesicles are described as "dewdrop on a rose petal" (clear vesicle on an erythematous base). Complications include secondary bacterial skin infection (most common), pneumonia (especially in adolescents and adults), cerebellar ataxia, encephalitis, and Reye syndrome (if aspirin is administered). The varicella vaccine (live attenuated) has dramatically reduced incidence. Treatment is supportive for uncomplicated cases; oral acyclovir is recommended for adolescents, adults, and immunocompromised patients.

20. D. Chlamydia trachomatis. Chlamydial pneumonia in infants (afebrile pneumonia syndrome) is acquired through vertical transmission during vaginal delivery from an infected mother. It typically presents between 4-12 weeks of age with a distinctive clinical picture — gradual onset tachypnea, staccato cough (distinctive repetitive, machine-gun-like cough), absence of fever (afebrile or low-grade), and bilateral conjunctivitis (which often precedes the pneumonia by 1-2 weeks). Peripheral eosinophilia is a characteristic laboratory finding. Chest X-ray reveals bilateral diffuse interstitial infiltrates with hyperinflation. Diagnosis is confirmed by Chlamydia PCR of nasopharyngeal specimens. Treatment is oral erythromycin or azithromycin for 14 days (erythromycin) or 5 days (azithromycin). The mother and her sexual partner should also be treated. Prenatal screening and treatment of maternal chlamydial infection prevents neonatal disease.

21. A. Coronary artery aneurysms. This presentation describes Kawasaki disease (mucocutaneous lymph node syndrome), diagnosed by fever lasting 5 or more days plus 4 of 5 clinical criteria — bilateral non-exudative conjunctivitis, oral changes (strawberry tongue, cracked lips), extremity changes (erythema, edema, later desquamation), polymorphous rash, and cervical lymphadenopathy (typically unilateral, node greater than 1.5 cm). Coronary artery aneurysms are the most feared complication, developing in 15-25% of untreated children and potentially causing myocardial infarction, sudden death, and chronic coronary artery disease. Treatment with IVIG (2 g/kg single infusion) plus high-dose aspirin (80-100 mg/kg/day during acute phase, then low-dose 3-5 mg/kg/day for 6-8 weeks) within the first 10 days of illness reduces coronary aneurysm risk to approximately 3-5%. Echocardiographic monitoring of coronary arteries is essential during acute illness and follow-up.

22. C. Atopic dermatitis (eczema). Atopic dermatitis is the most common chronic inflammatory skin condition in children, affecting approximately 10-20% of children. It is part of the "atopic triad" — atopic dermatitis, allergic rhinitis, and asthma — with a strong genetic component (family history of atopy is present in approximately 70% of cases). Age-dependent distribution patterns are characteristic — infants (face, extensor surfaces), children (flexural areas — antecubital and popliteal fossae, wrists, ankles), and adults (hands, eyelids, flexural areas). Key features include intense pruritus ("the itch that rashes"), xerosis (dry skin), erythematous papules and plaques with lichenification (thickened skin from chronic scratching), and a relapsing-remitting course. Treatment follows a stepwise approach — daily emollients

(cornerstone of therapy), topical corticosteroids for flares, calcineurin inhibitors (tacrolimus, pimecrolimus) for sensitive areas, and systemic therapy (dupilumab) for severe refractory disease.

23. B. Malrotation with midgut volvulus. Malrotation is a congenital anomaly of intestinal rotation and fixation during embryologic development, occurring when the midgut fails to complete its normal 270-degree counterclockwise rotation around the superior mesenteric artery. This creates abnormal mesenteric attachments (Ladd bands) and a narrow mesenteric base predisposing to midgut volvulus (twisting of the small bowel around the SMA). Bilious vomiting in a neonate is midgut volvulus until proven otherwise — this is a surgical emergency because volvulus can cause complete mesenteric vascular occlusion, leading to midgut necrosis within hours. Upper GI series is the gold standard, revealing the duodenojejunal junction (ligament of Treitz) displaced to the right of the midline with a "corkscrew" or "bird's beak" appearance. Treatment is emergent Ladd procedure (detorsion, division of Ladd bands, widening of the mesenteric base, appendectomy, and placement of the small bowel in the right abdomen and colon in the left).

24. D. Emergent surgical exploration with bilateral orchiopexy. Testicular torsion is a urologic emergency with the highest incidence during adolescence (particularly ages 12-18). Absent blood flow on Doppler ultrasound confirms torsion. Emergent surgical exploration must be performed within 6 hours of symptom onset for optimal testicular salvage (approximately 90-100% salvage rate within 6 hours, declining to less than 10% after 24 hours). Bilateral orchiopexy (fixation of both testes with non-absorbable sutures to the dartos fascia) is performed during the same operation because the bell-clapper deformity predisposing to torsion is bilateral in approximately 80% of cases. The contralateral testicle must be fixed even if it appears normal. If the affected testicle is non-viable upon exploration, orchiectomy is performed with contralateral orchiopexy. The key clinical features — sudden onset, high-riding testicle, horizontal lie, and absent cremasteric reflex — should prompt immediate surgical consultation.

25. A. Infantile spasms (West syndrome). Infantile spasms are a severe epileptic encephalopathy typically presenting between 4-8 months of age. The classic triad (West syndrome) consists of infantile spasms (sudden, brief, symmetric flexion or extension of the trunk and extremities occurring in clusters, often upon awakening), hypsarrhythmia on EEG (chaotic, high-voltage, disorganized background with multifocal spike-and-wave discharges — one of the most characteristic EEG patterns in pediatric epilepsy), and developmental regression. Causes include tuberous sclerosis (most common identifiable cause), perinatal hypoxic-ischemic injury, brain malformations, Down syndrome, and cryptogenic (no identifiable cause). Treatment is vigabatrin (first-line for tuberous sclerosis-associated spasms) or ACTH/high-dose prednisolone (first-line for cryptogenic and other causes). Prognosis varies — cryptogenic cases may have better outcomes, while symptomatic cases often progress to Lennox-Gastaut syndrome.

26. C. A broad-spectrum antiepileptic drug (levetiracetam or valproic acid). After a second unprovoked seizure, the diagnosis of epilepsy is established (epilepsy is defined as two or more unprovoked seizures occurring more than 24 hours apart, or one unprovoked seizure with a high probability of recurrence). Initiation of antiepileptic drug (AED) therapy is appropriate. For generalized

tonic-clonic seizures with generalized spike-and-wave discharges on EEG, a broad-spectrum AED is preferred. Levetiracetam is widely used as first-line due to its broad-spectrum efficacy, favorable side effect profile, and minimal drug interactions. Valproic acid is also effective for generalized epilepsy but has significant side effects (hepatotoxicity, teratogenicity, weight gain, thrombocytopenia). Narrow-spectrum AEDs (carbamazepine, phenytoin) can worsen generalized epilepsy syndromes. The goal is monotherapy at the lowest effective dose. Brain MRI and EEG are essential baseline studies.

27. B. Scarlet fever (Group A Streptococcus). Scarlet fever is a clinical syndrome caused by Group A Streptococcus (*Streptococcus pyogenes*) producing erythrogenic exotoxin (streptococcal pyrogenic exotoxin). It presents with streptococcal pharyngitis (fever, sore throat, tonsillar exudates, palatal petechiae) plus a characteristic diffuse, fine, sandpaper-textured erythematous rash that blanches with pressure. Pastia lines (linear petechiae in skin folds — axillae, antecubital fossae, groin) and circumoral pallor (pale area around the mouth contrasting with the flushed face) are distinctive features. The tongue progresses from a white-coated "white strawberry tongue" to a red "red strawberry tongue" as the coating desquamates. Desquamation of fingertips and toes occurs 1-2 weeks after the acute illness. Treatment is penicillin or amoxicillin for 10 days to eradicate GAS and prevent acute rheumatic fever. Rapid streptococcal antigen test or throat culture confirms the diagnosis.

28. A. Emergent X-ray and immediate endoscopic removal if in esophagus. Button battery ingestion is a pediatric emergency — batteries lodged in the esophagus can cause severe tissue injury within 2 hours through electrical current generation (producing hydroxide at the negative pole), pressure necrosis, and leakage of alkaline contents. Complications include esophageal perforation, tracheoesophageal fistula, aorto-esophageal fistula (potentially fatal massive hemorrhage), mediastinitis, and stricture formation. Immediate anteroposterior and lateral chest/abdominal X-rays are essential to localize the battery — on AP view, a button battery shows a characteristic "double-ring" or "halo" sign (step-off at the junction of the two battery components), distinguishing it from a coin. If lodged in the esophagus, emergent endoscopic removal is mandatory regardless of symptoms or duration. If the battery has passed into the stomach, management depends on the child's age, battery size, and symptoms — most batteries in the stomach pass spontaneously.

29. D. Immune thrombocytopenic purpura (ITP). ITP in children typically presents as acute onset of bruising, petechiae, and mucosal bleeding 1-4 weeks after a viral illness, in an otherwise well-appearing child. The hallmark is isolated severe thrombocytopenia with a completely normal remainder of the CBC and peripheral smear — normal WBC (no blasts, excluding leukemia), normal hemoglobin (no red cell line involvement, excluding aplastic anemia), and no schistocytes (excluding TTP/HUS). This distinguishes ITP from more serious diagnoses. Childhood ITP is usually self-limited, with approximately 80% resolving spontaneously within 6 months regardless of treatment. Treatment is based on bleeding severity rather than platelet count — observation alone for mild symptoms (bruising, petechiae), IVIG (rapid platelet increase within 24-48 hours) or corticosteroids for significant mucosal bleeding, and anti-Rh(D) immunoglobulin for Rh-positive children.

30. C. IgA vasculitis (Henoch-Schönlein purpura). IgA vasculitis (formerly HSP) is the most common systemic vasculitis in children, typically affecting children aged 3-10 years. It is an IgA-mediated small vessel vasculitis producing the classic tetrad — palpable purpura (non-thrombocytopenic, typically on the lower extremities and buttocks), arthralgia/arthritis (large joints, non-destructive), abdominal pain (colicky, from mesenteric vasculitis with risk of intussusception), and renal involvement (microscopic hematuria, proteinuria — IgA nephropathy). Normal platelet count is essential for distinguishing HSP purpura from ITP. Most cases follow an upper respiratory infection. The condition is usually self-limited (resolving within 4-6 weeks), with treatment being supportive (NSAIDs for arthralgia, analgesics for abdominal pain). Long-term prognosis depends on the degree of renal involvement — close monitoring of urinalysis and blood pressure for at least 6 months after diagnosis is recommended.

31. B. Hirschsprung disease. Hirschsprung disease (congenital aganglionic megacolon) results from failure of neural crest cell migration to the distal colon during fetal development, producing an aganglionic segment (lacking ganglion cells in the Meissner and Auerbach plexuses) that cannot relax, causing functional obstruction. The rectosigmoid region is most commonly affected (approximately 80%). The hallmark presentation is failure to pass meconium within the first 48 hours of life (normal is within 24-48 hours) followed by abdominal distension, bilious vomiting, and feeding intolerance. The "squirt sign" — explosive expulsion of stool and gas upon rectal examination — is characteristic. Barium enema reveals a transition zone between the narrow aganglionic segment and the dilated proximal normal colon. Diagnosis is confirmed by rectal suction biopsy showing absence of ganglion cells and hypertrophied nerve trunks with increased acetylcholinesterase staining. Definitive treatment is surgical pull-through procedure.

32. A. Hemolytic uremic syndrome (HUS). HUS is the most common cause of acute kidney injury in young children. Typical (diarrhea-associated) HUS is triggered by Shiga toxin-producing *E. coli* (STEC), particularly serotype O157:H7 from undercooked ground beef, unpasteurized milk, or contaminated water. The classic triad includes microangiopathic hemolytic anemia (schistocytes from mechanical RBC shearing through damaged microvasculature), thrombocytopenia (platelet consumption in microthrombi), and acute kidney injury (from glomerular and arteriolar endothelial damage). HUS develops in approximately 5-15% of children with STEC infection, typically 5-10 days after diarrhea onset. Treatment is supportive — IV fluids, transfusions, monitoring electrolytes, and dialysis for severe renal failure. Antibiotics are contraindicated as they may increase Shiga toxin release. Most children (approximately 70-85%) recover renal function completely.

33. D. Turner syndrome (45,X). Turner syndrome is the most common sex chromosome abnormality in females, caused by complete or partial absence of one X chromosome (45,X in approximately 50% of cases; mosaicism or structural X abnormalities in the remainder). Classic phenotypic features include short stature (most consistent finding, present in virtually all patients), webbed neck (pterygium colli), low posterior hairline, shield chest with widely spaced nipples, cubitus valgus, and streak gonads (fibrous ovarian tissue without follicles, causing primary amenorrhea and absent secondary sexual characteristics). Associated anomalies include bicuspid aortic valve (most common cardiac anomaly, approximately 15-30%), coarctation of the aorta, horseshoe kidney, and lymphedema in infancy. Diagnosis is confirmed by

karyotype analysis. Treatment includes growth hormone therapy (for short stature) and estrogen replacement therapy (to induce puberty and maintain secondary sexual characteristics and bone health).

34. B. Slipped capital femoral epiphysis (SCFE). SCFE is the most common hip disorder in adolescents, characterized by posterior and inferior displacement (slippage) of the proximal femoral epiphysis relative to the metaphysis through the hypertrophic zone of the growth plate. Classic presentation is an obese adolescent (BMI above the 95th percentile) with insidious onset of hip or knee pain (referred pain to the knee is common and may delay diagnosis) and limp. The pathognomonic examination finding is obligatory external rotation of the hip with flexion (the leg externally rotates as the hip is passively flexed). Frog-leg lateral hip X-ray is the most sensitive view, revealing posterior displacement of the epiphysis (Klein line — a line drawn along the superior femoral neck fails to intersect the epiphysis). Treatment is urgent surgical fixation with in situ screw fixation to prevent further slippage and avascular necrosis. The contralateral hip must be evaluated as bilateral involvement occurs in approximately 20-40%.

35. C. Legg-Calvé-Perthes disease. Legg-Calvé-Perthes disease is idiopathic avascular necrosis of the femoral head in children, typically affecting boys aged 4-10 years (peak age 5-7). The disease progresses through four stages — initial/necrosis (ischemia, femoral head appears dense on X-ray), fragmentation (resorption of necrotic bone, femoral head appears fragmented with mixed lucency and sclerosis), reossification (new bone formation), and remodeling (final shape of the femoral head). Presentation includes insidious onset of limp with hip or groin pain (sometimes referred to the knee), limited range of motion (especially abduction and internal rotation), and muscle atrophy. The "crescent sign" (subchondral fracture line visible as a radiolucent crescent) on X-ray indicates subchondral collapse. Short stature may be associated. Treatment goals include maintaining femoral head containment within the acetabulum — observation, abduction bracing, or surgical containment (femoral or pelvic osteotomy) based on age and severity.

36. D. Transient synovitis (toxic synovitis). Transient synovitis is the most common cause of acute hip pain and limp in children aged 3-8 years. It is a self-limited inflammatory condition of the hip joint, often following a viral upper respiratory infection by 1-2 weeks. Key features include low-grade or no fever, preserved range of motion (though internal rotation may be painful), normal or mildly elevated inflammatory markers (ESR, CRP), and small hip effusion on ultrasound. The critical distinction is from septic arthritis — the Kocher criteria help differentiate the two: non-weight bearing, fever greater than 101.3°F, ESR greater than 40 mm/hr, and WBC greater than 12,000/μL. Meeting zero or one Kocher criteria strongly suggests transient synovitis (less than 3% probability of septic arthritis). Treatment is supportive — rest, NSAIDs, and activity modification, with expected resolution within 1-2 weeks.

37. A. Systemic juvenile idiopathic arthritis (Still disease). Systemic JIA is a distinct subtype of juvenile idiopathic arthritis characterized by prominent systemic inflammatory features that distinguish it from other JIA subtypes. The hallmark features include quotidian (daily) high-spiking fevers (typically one or two spikes above 102.2°F with return to normal or below normal between spikes), evanescent salmon-colored macular rash (appears with fevers and disappears as fever resolves), and arthritis (may be absent initially, developing weeks to months after systemic symptoms). Hepatosplenomegaly, lymphadenopathy,

and serositis (pericarditis, pleuritis) are common. Laboratory findings include markedly elevated inflammatory markers (ESR, CRP, ferritin), leukocytosis, thrombocytosis, and anemia of chronic disease. ANA and RF are typically negative. Macrophage activation syndrome (MAS) is a life-threatening complication. Treatment includes NSAIDs, systemic corticosteroids, and biologic agents (IL-1 inhibitors — anakinra, canakinumab; IL-6 inhibitor — tocilizumab).

38. C. Acute rheumatic fever. Acute rheumatic fever (ARF) is a systemic inflammatory disease occurring 2-4 weeks after untreated Group A Streptococcal pharyngitis, caused by molecular mimicry between streptococcal M protein and host cardiac, joint, and CNS tissue. Diagnosis uses the revised Jones criteria — evidence of preceding GAS infection (elevated ASO titer, positive throat culture) plus two major criteria OR one major plus two minor criteria. Major criteria include carditis (new murmur, pancarditis), migratory polyarthritis (most common manifestation, large joints), Sydenham chorea, erythema marginatum (evanescent pink rings on trunk), and subcutaneous nodules. Minor criteria include arthralgia, fever, elevated ESR/CRP, and prolonged PR interval on ECG. This patient has two major criteria (carditis — new murmur, and migratory polyarthritis) plus evidence of GAS infection (elevated ASO). Treatment includes aspirin or NSAIDs for arthritis, corticosteroids for severe carditis, and penicillin to eradicate GAS. Secondary prophylaxis with monthly IM benzathine penicillin G prevents recurrence.

39. B. Congestive heart failure from left-to-right shunt (VSD). Ventricular septal defect is the most common congenital heart defect. Large VSDs produce significant left-to-right shunting (blood flows from the high-pressure left ventricle to the lower-pressure right ventricle), resulting in pulmonary overcirculation and volume overload of the left atrium and ventricle. As pulmonary vascular resistance naturally falls in the first weeks of life, the left-to-right shunt increases, leading to congestive heart failure typically manifesting at 4-8 weeks of age. Symptoms include poor feeding, diaphoresis during feeding (increased metabolic demand), tachypnea, failure to thrive, and hepatomegaly. The classic murmur is a holosystolic (pansystolic) murmur at the left lower sternal border. Chest X-ray shows cardiomegaly and increased pulmonary vascular markings. Medical management includes diuretics (furosemide), afterload reduction (captopril), and caloric supplementation. Surgical closure is indicated for large VSDs with heart failure unresponsive to medical therapy.

40. D. Transposition of the great arteries. TGA is the most common cyanotic congenital heart defect presenting in the newborn period, resulting from the aorta arising from the right ventricle and the pulmonary artery arising from the left ventricle, creating two parallel circulations. This produces severe cyanosis at birth that does not improve with supplemental oxygen (failed hyperoxia test — PaO₂ remains below 100 mmHg despite 100% FiO₂). Survival depends on mixing between the two circulations through a PDA, ASD, or VSD. Chest X-ray reveals the "egg-on-a-string" (or "egg-shaped heart") appearance — the narrow mediastinum results from the anteroposterior relationship of the great arteries. Immediate management includes prostaglandin E₁ (PGE₁) to maintain PDA patency. Balloon atrial septostomy (Rashkind procedure) provides temporary mixing through the atrial septum. Definitive surgical repair is the arterial switch operation (Jatene procedure) performed within the first 2 weeks of life.

41. A. Patent ductus arteriosus (PDA). The ductus arteriosus is a fetal vascular structure connecting the pulmonary artery to the aorta, normally closing within 24-72 hours after birth in response to increased oxygen tension and decreased prostaglandin levels. PDA is the persistence of this connection, more common in premature infants. The classic continuous "machinery-like" murmur (systolic and diastolic components) heard best in the left infraclavicular area is pathognomonic. Bounding pulses and widened pulse pressure result from diastolic runoff (aortic blood flowing through the PDA into the pulmonary artery during diastole). In premature infants, pharmacologic closure with indomethacin or ibuprofen (prostaglandin synthesis inhibitors) is first-line treatment. In full-term infants and older children, transcatheter device closure or surgical ligation is performed. PDA must be closed to prevent left heart volume overload, pulmonary hypertension, and eventual Eisenmenger syndrome.

42. C. Coarctation of the aorta. Coarctation of the aorta is a discrete narrowing of the aortic lumen, typically at the aortic isthmus just distal to the origin of the left subclavian artery near the ductus arteriosus insertion. The hallmark clinical finding is upper-to-lower extremity blood pressure discrepancy — hypertension in the upper extremities with diminished or absent femoral pulses and hypotension in the lower extremities. Critical coarctation in neonates presents with circulatory shock as the ductus arteriosus closes (blood flow to the lower body was dependent on the PDA). Immediate management is PGE1 infusion to reopen the PDA and restore lower body perfusion. Associated findings include bicuspid aortic valve (most common associated anomaly, approximately 50-85%), Turner syndrome, and intracranial aneurysms. In older children, rib notching on chest X-ray (from enlarged intercostal collateral arteries) and a "3 sign" (pre-stenotic and post-stenotic aortic dilation) may be seen. Surgical repair or balloon angioplasty with stent placement is definitive treatment.

43. D. Knee-to-chest positioning, oxygen, fluids, and morphine or phenylephrine. Tetralogy of Fallot (TOF) consists of four components — right ventricular outflow tract obstruction (RVOTO, most important determinant of severity), overriding aorta, ventricular septal defect, and right ventricular hypertrophy. Hypercyanotic ("tet") spells occur when dynamic infundibular (subpulmonic) spasm acutely worsens RVOTO, increasing right-to-left shunting through the VSD and producing severe cyanosis. The murmur diminishes during a spell because less blood flows across the obstructed RVOT. Immediate management includes knee-to-chest positioning (increases systemic vascular resistance, reducing right-to-left shunting), supplemental oxygen, IV fluid bolus, subcutaneous morphine (reduces agitation and hyperpnea that worsen the spell), and IV phenylephrine (alpha-agonist that increases SVR). Calming the child and avoiding agitation are essential. Surgical repair is definitive — complete intracardiac repair is typically performed at 3-6 months of age.

44. B. Atrial septal defect (ASD). ASD is the most common congenital heart defect presenting in adulthood, though it can present in childhood. The secundum ASD (defect in the fossa ovalis region, approximately 70% of ASDs) is the most common type. The pathognomonic finding is a fixed split S2 — the second heart sound is widely split and does not vary with respiration (normally, S2 splitting widens during inspiration). This fixed splitting results from the left-to-right shunt maintaining constant right ventricular volume regardless of respiratory phase. The systolic ejection murmur at the left upper sternal border results from increased flow across the pulmonary valve (not from flow across the ASD itself). ECG

shows right axis deviation and incomplete right bundle branch block from right ventricular volume overload. Significant ASDs (Qp:Qs greater than 1.5:1) are closed to prevent right heart failure, paradoxical embolism, and pulmonary hypertension. Transcatheter device closure is the preferred method for secundum ASDs.

45. A. Association with aniridia, genitourinary anomalies, and hemihypertrophy. This presentation describes Wilms tumor (nephroblastoma), the most common renal malignancy of childhood. Key associations include WAGR syndrome (Wilms tumor, Aniridia — absence of the iris, Genitourinary anomalies — cryptorchidism, hypospadias, and intellectual disability — caused by deletion of chromosome 11p13 including WT1 gene) and Beckwith-Wiedemann syndrome (macrosomia, macroglossia, omphalocele, hemihypertrophy, organomegaly, hypoglycemia — associated with WT2 on chromosome 11p15.5). Denys-Drash syndrome (WT1 mutation, diffuse mesangial sclerosis, male pseudohermaphroditism) also carries increased Wilms tumor risk. Elevated urine catecholamines (option B), periorbital ecchymosis and opsoclonus-myoclonus (option C), and calcifications encasing great vessels (option D) are all features of neuroblastoma.

46. D. Acute splenic sequestration crisis. Acute splenic sequestration is a life-threatening emergency in sickle cell disease, occurring when sickled red blood cells become trapped in the splenic sinusoids, causing rapid pooling of blood within the spleen. This produces acute splenomegaly, a precipitous drop in hemoglobin (often 2 g/dL or more below baseline), hypovolemic shock (from intravascular volume depletion), and reticulocytosis (bone marrow response to acute anemia). It typically affects children aged 6 months to 5 years with HbSS (before autosplenectomy occurs). Immediate treatment is aggressive IV fluid resuscitation and emergent blood transfusion to restore intravascular volume. After stabilization and resolution, splenectomy is recommended to prevent recurrence (recurrence rate approximately 50%). This contrasts with aplastic crisis (parvovirus B19 infection causing temporary marrow suppression with reticulocytopenia) and vaso-occlusive crisis (pain without acute hemoglobin drop).

47. C. Emergent exchange transfusion. Stroke is one of the most devastating complications of sickle cell disease, occurring in approximately 11% of children with HbSS by age 20. Ischemic stroke results from vaso-occlusion of cerebral arteries by sickled red blood cells and progressive vasculopathy (intimal hyperplasia). Emergent exchange transfusion (partial exchange to reduce HbS to less than 30% while avoiding hyperviscosity) is the standard acute treatment, simultaneously reducing HbS concentration and restoring normal oxygen-carrying capacity. Simple transfusion is used if exchange is not immediately available. tPA is generally not recommended for sickle cell stroke due to increased hemorrhagic transformation risk. After the acute event, chronic transfusion therapy (to maintain HbS below 30%) is continued indefinitely to prevent recurrent stroke. Transcranial Doppler ultrasonography screening (annually for children aged 2-16 with HbSS) identifies children at high risk for stroke before it occurs.

48. B. Neuroblastoma with orbital metastasis. Neuroblastoma is the third most common pediatric malignancy and the most common extracranial solid tumor in children. Metastatic spread to the orbit is a classic presentation, producing unilateral or bilateral proptosis and periorbital ecchymosis ("raccoon eyes"). Elevated urine catecholamine metabolites (VMA and HVA) are present in greater than 90% of

neuroblastomas because the tumor arises from sympathetic neural crest cells that produce catecholamines. Histologically, neuroblastoma is classified as a small round blue cell tumor (along with Ewing sarcoma, rhabdomyosarcoma, lymphoma, and Wilms tumor). Homer-Wright pseudorosettes (neuroblasts surrounding a central neuropil core) are a characteristic histologic pattern. MIBG (metaiodobenzylguanidine) scan is used for staging and detection of metastatic disease. Prognosis depends on age (better under 18 months), stage, MYCN amplification status, and tumor histology.

49. A. Retinoblastoma. Retinoblastoma is the most common intraocular malignancy of childhood, with a mean age of diagnosis at 18 months. Leukocoria (white pupillary reflex) is the most common presenting sign, often first noticed in photographs when the affected eye shows a white reflection instead of the normal red reflex. Strabismus is the second most common presentation. Retinoblastoma occurs in hereditary (approximately 40%, bilateral, germline RB1 mutation on chromosome 13q14) and sporadic (approximately 60%, unilateral, somatic mutation) forms. The father's history of a similar condition requiring enucleation strongly suggests the hereditary form following autosomal dominant inheritance with high penetrance. According to Knudson's "two-hit hypothesis," hereditary retinoblastoma requires only one additional somatic mutation (the germline mutation is the first hit), while sporadic cases require two somatic mutations. Hereditary retinoblastoma carriers have increased lifetime risk of secondary malignancies (especially osteosarcoma).

50. D. Ewing sarcoma. Ewing sarcoma is the second most common primary malignant bone tumor in children and adolescents (after osteosarcoma), with peak incidence at 10-15 years. It characteristically affects the diaphysis (midshaft) of long bones and flat bones (pelvis, ribs, scapula). The hallmark radiographic finding is a multilayered ("onion-skin") periosteal reaction from successive layers of reactive bone formation in response to the tumor. Histologically, Ewing sarcoma consists of uniform small round blue cells arranged in rosettes (Homer-Wright rosettes). The characteristic cytogenetic abnormality is t(11;22) translocation producing the EWS-FLI1 fusion gene (present in approximately 85% of cases). CD99 (MIC2) is a sensitive immunohistochemical marker. Ewing sarcoma is distinguished from osteosarcoma by its diaphyseal location, onion-skin periosteal reaction (versus sunburst), and small round blue cell histology. Treatment is neoadjuvant chemotherapy, local control (surgery and/or radiation), and adjuvant chemotherapy.

51. C. DTaP, IPV, Hib, PCV13, and rotavirus. The 2-month well-child visit is one of the most vaccine-dense visits in the childhood immunization schedule. Vaccines administered at 2 months include DTaP (diphtheria, tetanus, acellular pertussis — first dose of a 5-dose series), IPV (inactivated poliovirus — first dose), Hib (Haemophilus influenzae type b conjugate — first dose), PCV13 (13-valent pneumococcal conjugate — first dose), rotavirus (oral live attenuated — first dose, must be initiated before 15 weeks of age), and hepatitis B (second dose if the birth dose was given). MMR and varicella vaccines are not given until 12-15 months because maternal antibodies interfere with immune response to live vaccines. Hepatitis A vaccine begins at 12 months. Understanding the immunization schedule is essential for the PANCE.

52. B. Report suspected child abuse to child protective services. This presentation has multiple red flags for non-accidental trauma (child abuse) — bruises in various stages of healing (indicating repeated

injury over time), bruises in atypical locations for accidental injury (back, buttocks, upper arms — protected areas not commonly injured in falls), patterned bruises (belt buckle marks indicating inflicted injury with an identifiable object), cigarette burn (pathognomonic for intentional injury), and an inconsistent history (3-year-old children do not typically fall down stairs in a pattern producing bilateral buttock and back bruising). Healthcare providers are mandated reporters — all 50 states require reporting of suspected child abuse to child protective services (CPS). A full skeletal survey (AP and lateral views of the entire skeleton) is indicated in all children under 2 years with suspected abuse and selectively in children 2-5 years, to identify occult fractures. Classic fractures of abuse include metaphyseal corner ("bucket handle") fractures, posterior rib fractures, and fractures in various stages of healing.

53. A. Urgent joint aspiration of the right hip. This presentation raises high concern for septic arthritis of the hip — the most important orthopedic emergency in pediatrics. All four Kocher criteria are met — non-weight bearing, fever greater than 101.3°F, ESR greater than 40 mm/hr, and WBC greater than 12,000/ μ L — giving a predicted probability of septic arthritis greater than 99%. CRP greater than 2.0 mg/dL has been added as a fifth predictor. Urgent joint aspiration (arthrocentesis) is both diagnostic and therapeutic — synovial fluid analysis revealing WBC greater than 50,000/ μ L with greater than 90% neutrophils, positive Gram stain, or positive culture confirms the diagnosis. *Staphylococcus aureus* is the most common cause in all age groups beyond the neonatal period. Treatment is emergent surgical irrigation and debridement (particularly for the hip joint) combined with IV antibiotics. Delays in treatment can lead to permanent joint destruction, avascular necrosis of the femoral head, growth disturbance, and sepsis.

54. C. Simple febrile seizure. Simple febrile seizures are the most common seizure type in children, occurring in approximately 2-5% of children aged 6 months to 5 years. They are defined as generalized tonic-clonic seizures lasting less than 15 minutes, occurring once within a 24-hour period, in a child with a febrile illness (temperature greater than 100.4°F) and no history of prior afebrile seizures or CNS abnormality. This child meets all criteria — generalized seizure, brief duration (3 minutes), single episode, in the context of fever from acute otitis media, with rapid return to baseline. Complex febrile seizures are defined by focal features, duration greater than 15 minutes, or recurrence within 24 hours. Simple febrile seizures require no routine laboratory workup, EEG, or neuroimaging. Long-term anticonvulsant therapy is not indicated. The recurrence risk is approximately 30% with the next febrile illness. Importantly, simple febrile seizures do not increase the risk of developing epilepsy.

55. D. Foreign body aspiration. Foreign body aspiration is a common and potentially life-threatening emergency in children aged 6 months to 3 years, occurring when small objects (most commonly peanuts, grapes, hot dogs, coins, small toy parts) are inhaled into the tracheobronchial tree. The right mainstem bronchus is more commonly affected due to its wider diameter and more vertical orientation. Classic presentation includes sudden onset of choking, coughing, and unilateral wheezing in a previously healthy child. The foreign body acts as a ball-valve mechanism — allowing air in during inspiration but trapping it during expiration, producing unilateral air trapping (hyperinflation). Expiratory chest X-ray reveals hyperinflation of the affected lung with mediastinal shift to the contralateral side (away from the affected

side during expiration). Inspiratory films may appear normal. Diagnosis and treatment is rigid bronchoscopy with foreign body removal.

56. B. Port-wine stain (nevus flammeus). A port-wine stain is a congenital capillary vascular malformation present at birth that does not change in size, does not proliferate, and persists lifelong. It appears as a flat, well-demarcated, erythematous to violaceous patch that does not blanch completely with pressure and does not change with crying (distinguishing it from infantile hemangiomas, which are not present at birth, proliferate during the first year, and eventually involute). Port-wine stains in the distribution of the ophthalmic branch (V1) of the trigeminal nerve raise concern for Sturge-Weber syndrome (leptomeningeal angiomatosis causing seizures, intellectual disability, and glaucoma). Treatment is pulsed dye laser therapy for cosmetic improvement. Infantile hemangiomas (strawberry hemangiomas) are the most common tumors of infancy — they are typically not present at birth, appear within the first few weeks, rapidly proliferate, and spontaneously involute over years.

57. A. ABO hemolytic disease of the newborn. ABO hemolytic disease occurs when maternal anti-A or anti-B antibodies (IgG, which crosses the placenta) attack fetal red blood cells expressing A or B antigens. It most commonly affects type A or B infants born to type O mothers (who naturally produce IgG anti-A and anti-B). The positive direct Coombs test (DAT) confirms antibody-coated fetal red blood cells. Spherocytes on peripheral smear result from partial phagocytosis of antibody-coated RBCs by splenic macrophages, and reticulocytosis reflects the bone marrow's compensatory response to hemolysis. ABO incompatibility is generally milder than Rh disease because A and B antigens are widely expressed on non-erythroid tissues (reducing antibody targeting of RBCs) and because fetal A and B antigen expression is not fully developed. Treatment is phototherapy for hyperbilirubinemia, with exchange transfusion reserved for severe cases. Physiologic jaundice is excluded by the positive DAT and hemolytic findings.

58. C. Medulloblastoma (or pilocytic astrocytoma of the cerebellum). Posterior fossa tumors are the most common brain tumors in children, with medulloblastoma and pilocytic astrocytoma being the two most frequent types. Medulloblastoma is a highly malignant embryonal tumor of the cerebellum arising from the cerebellar vermis (midline), typically in children aged 4-8 years. It presents with signs of cerebellar dysfunction (truncal ataxia, dysmetria, gait unsteadiness) and obstructive hydrocephalus (headache worse in the morning, vomiting, papilledema, sixth nerve palsy) from compression of the fourth ventricle. Pilocytic astrocytoma (the most common pediatric brain tumor overall) also arises in the posterior fossa but is typically a cerebellar hemisphere lesion, cystic with a mural nodule, and carries an excellent prognosis. Medulloblastoma has a propensity for CSF dissemination (drop metastases). Treatment includes surgical resection, craniospinal radiation (for children over 3 years), and chemotherapy.

59. D. Duchenne muscular dystrophy. DMD is the most common and most severe muscular dystrophy, caused by X-linked recessive mutations in the dystrophin gene (Xp21), resulting in absent or severely reduced dystrophin protein — a critical structural protein linking the intracellular cytoskeleton to the extracellular matrix in muscle fibers. Progressive proximal muscle weakness begins by age 2-5 years. The Gowers maneuver (using hands to "walk up" the legs when rising from the floor) demonstrates proximal

hip girdle weakness. Calf pseudohypertrophy (replacement of muscle with fibrofattite tissue) is characteristic. CK is markedly elevated (10,000-50,000 U/L) from ongoing muscle destruction. X-linked inheritance explains the affected maternal uncle. Disease progression is relentless — wheelchair dependence by age 12, respiratory failure and cardiomyopathy in the late teens to twenties. Corticosteroids (prednisone or deflazacort) are the mainstay of treatment, slowing disease progression. Gene therapy and exon-skipping therapies are emerging treatments.

60. B. Listeria monocytogenes. Neonatal meningitis is caused by different organisms depending on timing. Early-onset (less than 7 days) is caused by Group B Streptococcus (most common), *E. coli* (second most common), and *Listeria monocytogenes*. Late-onset (7-28 days) includes these organisms plus coagulase-negative staphylococci. The Gram stain revealing gram-positive rods is the critical clue — *Listeria monocytogenes* is the only common neonatal meningitis pathogen that is a gram-positive rod. *E. coli* is a gram-negative rod, GBS is a gram-positive coccus, and *S. pneumoniae* is a gram-positive diplococcus. Lack of prenatal care increases the risk of *Listeria* transmission through contaminated food (unpasteurized dairy, deli meats). Importantly, *Listeria* is intrinsically resistant to cephalosporins — this is why ampicillin is included in empiric neonatal meningitis regimens (ampicillin plus gentamicin or ampicillin plus cefotaxime). Bulging fontanelle, irritability, and poor feeding are the most common signs of meningitis in neonates.

61. A. Acute hematogenous osteomyelitis. Acute hematogenous osteomyelitis is the most common form of bone infection in children, with *Staphylococcus aureus* being the causative organism in approximately 70-80% of all age groups beyond the neonatal period. The metaphysis of long bones (distal femur, proximal tibia, proximal humerus) is the most commonly affected location because the sluggish blood flow through the sinusoidal venous loops in the metaphyseal region promotes bacterial seeding. Clinical features include localized bone pain, warmth, swelling, tenderness, refusal to use the affected extremity, fever, and elevated inflammatory markers (WBC, ESR, CRP). Blood cultures are positive in approximately 30-50% of cases. MRI is the imaging modality of choice, revealing marrow edema, periosteal reaction, and abscess formation with high sensitivity and specificity. Treatment is IV antistaphylococcal antibiotics (nafcillin, cefazolin, or vancomycin for MRSA) for 4-6 weeks and surgical drainage of any abscess.

62. C. Intravenous immunoglobulin (IVIG) plus high-dose aspirin. Treatment of Kawasaki disease aims to reduce inflammation and prevent coronary artery aneurysms. The standard regimen is IVIG (2 g/kg as a single infusion over 10-12 hours) plus high-dose aspirin (80-100 mg/kg/day in 4 divided doses during the acute febrile phase). IVIG administered within the first 10 days of illness reduces the incidence of coronary aneurysms from approximately 15-25% to 3-5%. Once the fever resolves (typically 36-48 hours after IVIG), aspirin is reduced to low-dose (3-5 mg/kg/day) for its antiplatelet effect, continued for 6-8 weeks or until echocardiographic follow-up confirms no coronary abnormalities. Aspirin is the only NSAID used in Kawasaki disease. Refractory cases (persistent fever despite IVIG) may receive a second IVIG dose or IV methylprednisolone. Serial echocardiograms are performed to monitor coronary artery dimensions.

63. B. Aortic stenosis (from bicuspid aortic valve). Bicuspid aortic valve is the most common congenital cardiac anomaly, present in approximately 1-2% of the population. While many patients remain asymptomatic throughout childhood, the abnormal valve architecture (two leaflets instead of three) creates turbulent flow and progressive calcification, leading to aortic stenosis (most commonly) or aortic regurgitation in adolescence or young adulthood. The classic murmur of aortic stenosis is a crescendo-decrescendo (diamond-shaped) systolic ejection murmur heard best at the right upper sternal border (aortic area) with radiation to the carotid arteries. A systolic ejection click preceding the murmur indicates a valvular etiology (rather than subvalvular or supra-aortic). Symptoms of significant stenosis include fatigue, exercise intolerance, exertional syncope, and angina. Echocardiography confirms the diagnosis and quantifies severity. Intervention (balloon valvuloplasty or surgical valve replacement) is indicated for severe stenosis.

64. D. N-acetylcysteine (NAC). Acetaminophen overdose is the most common cause of acute liver failure in the United States. The toxic dose is approximately 150 mg/kg in children. This child ingested approximately 10,000 mg (approximately 200+ mg/kg, assuming average weight for a 10-year-old). Acetaminophen is metabolized by the liver — at therapeutic doses, the toxic metabolite NAPQI is conjugated by glutathione. In overdose, glutathione stores are depleted, allowing NAPQI to cause direct hepatocellular necrosis. The Rumack-Matthew nomogram (plotting serum acetaminophen level against time since ingestion at 4 hours or later) guides treatment — levels above the treatment line indicate probable hepatotoxicity and warrant NAC administration. NAC is the specific antidote, replenishing glutathione stores and providing a substrate for alternative NAPQI detoxification. NAC is most effective when administered within 8 hours of ingestion but provides benefit up to 72 hours after ingestion.

65. A. Meckel diverticulum. Meckel diverticulum is the most common congenital anomaly of the gastrointestinal tract, resulting from incomplete obliteration of the omphalomesenteric (vitelline) duct. It follows the "rule of 2s" — 2% of the population, 2 feet from the ileocecal valve, 2 inches long, 2 types of ectopic tissue (gastric and pancreatic), 2 years of age at most common presentation, and 2:1 male-to-female ratio. Painless rectal bleeding (bright red or maroon) is the most common presentation in children, caused by ectopic gastric mucosa producing acid that ulcerates adjacent ileal mucosa. A Meckel scan (technetium-99m pertechnetate scintigraphy) detects ectopic gastric tissue with approximately 85% sensitivity and 95% specificity. Treatment is surgical excision (diverticulectomy or segmental small bowel resection).

66. B. Galactosemia. Classic galactosemia is an autosomal recessive disorder caused by deficiency of galactose-1-phosphate uridylyltransferase (GALT), resulting in inability to metabolize galactose (a component of lactose in breast milk and standard formulas). Toxic metabolites (galactose-1-phosphate and galactitol) accumulate, causing hepatotoxicity (jaundice, hepatomegaly, coagulopathy), cataracts, intellectual disability, and increased susceptibility to *E. coli* sepsis (a characteristic and potentially fatal early complication). The key diagnostic clue is urine positive for reducing substances (Clinitest, which detects any reducing sugar including galactose) but negative for glucose oxidase test (which is specific for glucose only) — indicating the presence of a non-glucose reducing sugar (galactose) in the urine.

Treatment is immediate elimination of all galactose and lactose from the diet (soy-based formula). Newborn screening detects galactosemia through measurement of GALT activity.

67. C. Anorexia nervosa. Anorexia nervosa is a serious psychiatric disorder characterized by restriction of energy intake leading to significantly low body weight, intense fear of gaining weight or persistent behavior interfering with weight gain, and disturbance in body image perception. BMI less than 17.5 or below the 5th percentile for age in adolescents indicates severe underweight. Physical signs include lanugo hair (fine body hair, a compensatory thermoregulatory response), dry skin, hair loss, hypothermia, bradycardia (from vagal tone increase and metabolic adaptation), hypotension, and amenorrhea (from hypothalamic suppression of GnRH). Medical complications include electrolyte abnormalities (hypokalemia, hypophosphatemia), osteoporosis, cardiac arrhythmias (prolonged QTc), refeeding syndrome, and death (anorexia has the highest mortality rate of any psychiatric disorder). Treatment requires a multidisciplinary approach — nutritional rehabilitation, psychotherapy (family-based therapy is first-line for adolescents), and medical monitoring.

68. D. Steroid-responsive nephrotic syndrome with approximately 50-75% chance of relapse but excellent long-term prognosis. Minimal change disease in children is characterized by its excellent response to corticosteroids — approximately 90% of children achieve complete remission (resolution of proteinuria) within 4-8 weeks of prednisone therapy. However, relapse is common, occurring in approximately 50-75% of steroid-responsive patients. Despite frequent relapses, the long-term renal prognosis is excellent — MCD rarely progresses to chronic kidney disease or ESRD. Frequent relapsers (2 or more relapses within 6 months or 4 or more within 12 months) or steroid-dependent patients may require steroid-sparing agents (cyclophosphamide, mycophenolate mofetil, calcineurin inhibitors, or rituximab). Steroid resistance (failure to achieve remission after 8 weeks of prednisone) should prompt renal biopsy to evaluate for focal segmental glomerulosclerosis, which has a less favorable prognosis.

69. A. Emergent endoscopic removal of the esophageal foreign body. Esophageal foreign bodies in children require prompt intervention, particularly when lodged in the upper esophagus at the cricopharyngeus muscle (the most common site of esophageal foreign body impaction, accounting for approximately 60-70% of cases). Coins are the most commonly ingested foreign body in children. Symptoms of esophageal impaction include drooling, dysphagia, refusal to eat, and neck/chest pain. Esophageal foreign bodies should be removed within 24 hours to prevent complications (ulceration, perforation, aspiration). Objects at the cricopharyngeus level, sharp objects, and batteries lodged in the esophagus require emergent removal. Once a coin passes into the stomach, observation is generally appropriate as most objects pass spontaneously through the GI tract. Endoscopic removal using rigid or flexible endoscopy is the preferred method.

70. B. Transient tachypnea of the newborn. TTN (also called "wet lung") is the most common cause of respiratory distress in term and late preterm newborns, resulting from delayed clearance of fetal lung fluid. During vaginal delivery, thoracic compression and catecholamine surge facilitate lung fluid absorption — cesarean delivery (as in this case) bypasses this mechanism, making TTN more common. Presentation includes tachypnea (respiratory rate often 60-80/min), mild grunting, mild retractions, and mild

hypoxemia that responds well to supplemental oxygen. Chest X-ray reveals prominent perihilar streaking (engorged perihilar lymphatics from retained fluid), fluid in the interlobar fissures, and mild hyperinflation. TTN is self-limited, with symptoms resolving within 24-72 hours as the fluid is absorbed. This distinguishes TTN from respiratory distress syndrome (hyaline membrane disease — affects preterm infants, ground-glass appearance with air bronchograms) and meconium aspiration syndrome (post-term, meconium-stained amniotic fluid, coarse irregular opacities).