

# COMPREHENSIVE MOCK EXAM 9

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Supplemental to: Neonatology Board Review 2026-2027

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*American Board of Pediatrics (ABP) Neonatal-Perinatal Medicine Subspecialty Certification Examination — 200-question supplemental simulation, single-best-answer A–D format matching the parent book, with comprehensive coverage of current ABP NPM content domains*

## Board-Review Questions

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1. A newborn has persistent tachycardia, irritability, poor weight gain, and a goiter. The mother has Graves disease but underwent thyroid ablation several years ago and is now hypothyroid on replacement therapy. What is the most likely cause?

- A. Transplacental levothyroxine causing direct neonatal thyrotoxicosis
- B. Maternal iodine deficiency causing neonatal thyroid overactivity
- C. Transplacental maternal thyroid-stimulating receptor antibodies
- D. Placental conversion of thyroxine into thyroid-stimulating hormone

2. A 30-week infant is born to a mother with severe preeclampsia and fetal growth restriction. The infant has a platelet count of 82,000/ $\mu$ L but is otherwise stable. Which maternal condition most plausibly contributes to this neonatal finding?

- A. Uncomplicated maternal group B streptococcal colonization
- B. Placental insufficiency associated with maternal hypertensive disease
- C. Maternal iron deficiency without placental dysfunction
- D. Isolated maternal hypothyroidism treated throughout pregnancy

3. Minutes after birth, an infant has severe upper-airway obstruction during quiet breathing. Cyanosis resolves when an oral airway is placed, and no airflow is detected from either naris. Which diagnosis best explains the obstruction?

- A. Bilateral choanal atresia
- B. Isolated cleft palate
- C. Laryngomalacia
- D. Unilateral vocal-cord paralysis

4. A 25-week infant is delivered in a cold operating room. Which intervention most directly reduces evaporative heat loss during the first minutes after birth?
- A. Place the wet infant promptly in a polyethylene wrap or bag under a radiant warmer
  - B. Dry the infant completely and leave the skin exposed to room air
  - C. Delay thermal interventions until an axillary temperature is obtained
  - D. Apply cool saline packs while preparing respiratory support
5. During umbilical venous catheter placement for neonatal resuscitation, rapid emergency vascular access is needed for epinephrine. Which placement strategy is appropriate for immediate temporary use?
- A. Advance blindly until resistance is met deep within the liver
  - B. Place the catheter into an umbilical artery because venous access is contraindicated during resuscitation
  - C. Advance the catheter only far enough to obtain free-flowing venous blood, then secure it for emergency medication delivery
  - D. Delay all vascular medication until a peripherally inserted central catheter can be placed
6. A critically ill newborn requires an immediately life-saving procedure, but the parents cannot be reached despite reasonable attempts. Delay would place the infant at substantial risk of death. Which ethical approach is most appropriate?
- A. Ask an unrelated visitor to sign consent
  - B. Withhold all care until written parental consent is obtained regardless of urgency
  - C. Provide the emergency treatment that is necessary to protect the infant
  - D. Delay treatment until a court order is obtained even if the infant may die first

7. A previously stable 2-day-old develops green emesis and rapidly worsening perfusion. An urgent upper gastrointestinal study shows an abnormal duodenojejunal position with a corkscrew configuration of proximal small bowel. Which diagnosis requires immediate operative attention?

- A. Midgut volvulus due to intestinal malrotation
- B. Infant dyschezia
- C. Physiologic gastroesophageal reflux
- D. Uncomplicated meconium plug syndrome

8. A very preterm infant has spent several months in the NICU and is ready for discharge. Which hearing-screening method is preferred for infants with a prolonged NICU stay because it can detect auditory neuropathy?

- A. Visual acuity testing
- B. Automated auditory brainstem response
- C. Tympanometry alone
- D. Behavioral response to a hand clap alone

9. A 30-week infant on conventional ventilation has worsening oxygenation despite increasing mean airway pressure. Chest radiography shows diffuse atelectasis without a focal air leak. Which rescue mode can recruit lung using very small tidal oscillations around a set mean airway pressure?

- A. Unassisted room-air breathing
- B. High-frequency oscillatory ventilation
- C. Synchronized cardioversion at the respiratory rate
- D. Intermittent blow-by oxygen without positive pressure

**10.** A newborn has flank swelling, gross hematuria, thrombocytopenia, and a unilateral enlarged kidney on ultrasonography. Which diagnosis is most likely?

- A.** Renal vein thrombosis
- B.** Simple physiologic hydronephrosis
- C.** Posterior urethral valves
- D.** Central diabetes insipidus

**11.** A preterm infant has hyponatremia during the second week while gaining weight appropriately. Urine sodium remains high. Which developmental feature contributes?

- A.** Complete absence of glomerular filtration
- B.** Mature distal tubules that conserve sodium better than term kidneys
- C.** Immature tubular sodium reabsorption causing renal sodium losses
- D.** Excessive aldosterone responsiveness causing sodium retention

**12.** A term infant has generalized hypotonia, a flat facial profile with epicanthal folds, a wide sandal gap, and an echocardiogram showing a complete atrioventricular canal. Which chromosomal disorder is most likely?

- A.** Trisomy 13
- B.** 22q11.2 deletion syndrome
- C.** Trisomy 21
- D.** Monosomy X

**13.** A 29-week fetus has severe growth restriction. Umbilical artery Doppler shows reversed end-diastolic flow, and the placenta is small with multiple infarcts. Which placental process best explains these findings?

- A.** Diffuse villous edema caused by fetal hydrops
- B.** Fetal vascular malperfusion from an umbilical venous thrombus
- C.** Acute ascending chorioamnionitis with neutrophilic inflammation
- D.** Maternal vascular malperfusion from uteroplacental insufficiency

**14.** A neonate requires airway support for bilateral posterior nasal obstruction. Examination also reveals an ocular coloboma, hypoplastic external genitalia, and abnormal semicircular canals. Which syndrome best accounts for this combination?

- A.** Achondroplasia
- B.** Turner syndrome
- C.** VACTERL association
- D.** CHARGE syndrome

**15.** A high-position umbilical arterial catheter is intentionally placed to reduce branch-vessel complications. Which radiographic tip location is generally targeted?

- A.** Descending thoracic aorta at approximately T6-T9
- B.** Portal vein within the hepatic circulation
- C.** Right atrium beyond the tricuspid valve
- D.** Main pulmonary artery proximal to its branches

**16.** A mother cannot provide her own milk for a 26-week infant. Which alternative is generally preferred over preterm formula when the priority is reducing necrotizing enterocolitis risk?

- A.** Diluted evaporated milk
- B.** Pasteurized donor human milk
- C.** Clear carbohydrate solution without protein
- D.** Unmodified goat milk

**17.** A well newborn has brief repetitive jerks of both arms only during sleep. The movements stop immediately when the infant is awakened, and the neurologic examination is normal. Which diagnosis is most likely?

- A.** Hyperekplexia with continuous rigidity
- B.** Infantile spasms
- C.** Benign neonatal sleep myoclonus
- D.** Status epilepticus

**18.** A term infant develops focal clonic seizures at 18 hours of age. Examination later shows asymmetric tone. Cranial ultrasound is unrevealing. Which study is most sensitive for detecting a suspected perinatal arterial ischemic stroke?

- A.** Plain radiographs of the skull in multiple views
- B.** Electromyography with nerve-conduction testing
- C.** Ultrasound examination of the lumbar spine
- D.** Brain MRI with diffusion-weighted sequences

**19.** A 1-day-old infant has a serum creatinine similar to the mother's value despite normal urine output. Why is creatinine difficult to interpret immediately after birth?

- A.** Early neonatal creatinine partly reflects maternal creatinine transferred across the placenta
- B.** Neonatal creatinine is determined only by bilirubin concentration
- C.** Creatinine is not filtered by the neonatal kidney at any gestational age
- D.** Maternal creatinine cannot cross the placenta

**20.** A newborn develops a persistent milky pleural effusion after thoracic surgery. Pleural fluid has a high triglyceride concentration and lymphocyte predominance after enteral feeds are established. What is the diagnosis?

- A.** Empyema from untreated bacterial pneumonia
- B.** Transudative effusion from isolated hypoalbuminemia
- C.** Hemothorax from ongoing arterial bleeding
- D.** Chylothorax from lymphatic leakage

**21.** A preterm infant has recurrent regurgitation after feeds but is growing, has no apnea temporally linked to events, and has no hematemesis. Which statement is most accurate?

- A.** Gastroesophageal reflux is common and often reflects developmental physiology
- B.** Acid suppression is required for all visible regurgitation
- C.** Reflux in preterm infants proves cow-milk protein allergy
- D.** Every reflux episode indicates a surgical emergency

**22.** A 30-week infant is clinically stable but not yet ready for large enteral volumes. Small-volume trophic feeds are started. What is the main rationale?

- A.** Intentionally induce intestinal hypoperfusion to prevent overgrowth
- B.** Replace all parenteral nutrition immediately regardless of tolerance
- C.** Provide enteral stimulation while larger nutritional needs are met gradually
- D.** Prevent bilirubin conjugation by limiting hepatic blood flow

**23.** A patient at 29 weeks is expected to deliver imminently. Magnesium sulfate is administered before birth for a fetal indication. What neonatal outcome is this strategy primarily intended to improve?

- A.** Prevention of neonatal hypoglycemia through increased glycogen storage
- B.** Elimination of early-onset group B streptococcal infection
- C.** Closure of the fetal ductus arteriosus before delivery
- D.** Reduction in the risk of cerebral palsy among survivors of very preterm birth

**24.** A cyanotic newborn has a harsh systolic murmur. Echocardiography shows ventricular septal defect, overriding aorta, right ventricular outflow tract obstruction, and right ventricular hypertrophy. Which diagnosis is present?

- A.** Tetralogy of Fallot
- B.** Truncus arteriosus
- C.** Total anomalous pulmonary venous return
- D.** Hypoplastic left heart syndrome

**25.** A former 25-week infant has a gradual fall in hemoglobin over several weeks, a low reticulocyte count, and no evidence of bleeding or hemolysis. Which diagnosis is most likely?

- A.** Acute splenic rupture
- B.** Massive fetomaternal hemorrhage
- C.** Immune hemolytic disease
- D.** Anemia of prematurity

**26.** An infant born to a mother with diabetes becomes jittery at 12 hours. Glucose is normal, but total and ionized calcium are low. Which condition is most likely?

- A.** Familial hypocalciuric hypercalcemia causing neonatal symptoms
- B.** Early neonatal hypocalcemia associated with maternal diabetes
- C.** Primary neonatal hyperparathyroidism with skeletal demineralization
- D.** Vitamin D intoxication producing neonatal hypercalcemia

**27.** A newborn with a urea-cycle defect becomes lethargic and tachypneic. Ammonia is markedly elevated, glucose is normal, and the anion gap is not increased. Which acid-base pattern may be seen early?

- A.** Respiratory alkalosis from hyperammonemia-induced hyperventilation
- B.** High-anion-gap ketoacidosis as the defining early feature
- C.** Severe metabolic alkalosis from chloride retention
- D.** Isolated respiratory acidosis from central hypoventilation

**28.** A term infant develops respiratory distress, hypotension, and temperature instability at 8 hours of life. The mother was colonized with group B Streptococcus and received no intrapartum antibiotics. Which pathogen is most likely?

- A.** Candida parapsilosis after several weeks of parenteral nutrition
- B.** Respiratory syncytial virus acquired from daycare attendance
- C.** Group B Streptococcus acquired around the time of delivery
- D.** Coagulase-negative staphylococcus acquired from a long-term central line

**29.** Thick meconium is present at delivery. After warming and stimulation, the newborn remains limp and apneic and the heart rate is 80/min. Which action should take precedence over attempts to clear meconium from the trachea?

- A.** Perform routine endotracheal suction before any ventilation is attempted
- B.** Wait for spontaneous respirations while giving blow-by oxygen
- C.** Begin positive-pressure ventilation without delaying for routine tracheal suction
- D.** Provide chest compressions immediately because meconium is present

**30.** A 28-week infant on mechanical ventilation has adequate oxygenation but a PaCO<sub>2</sub> of 68 mm Hg with respiratory acidosis. If all else is unchanged, which adjustment most directly increases minute ventilation?

- A.** Increase inspired oxygen concentration alone
- B.** Lower alveolar ventilation by increasing dead-space volume
- C.** Decrease minute ventilation while keeping oxygen unchanged
- D.** Increase effective tidal ventilation or ventilator rate

**31.** A newborn has jaundice and anemia. The mother is blood group O and the infant is group A. Direct antiglobulin testing is positive. Which process is most likely?

- A.** G6PD deficiency with a negative antibody test
- B.** Biliary atresia causing conjugated hyperbilirubinemia
- C.** ABO immune-mediated hemolysis
- D.** Physiologic jaundice without hemolysis

**32.** A term newborn's heart rate is 50/min despite at least 30 seconds of ventilation that clearly produces chest movement. What is the next intervention?

- A.** Give naloxone before any circulatory support
- B.** Switch to blow-by oxygen and reassess after spontaneous breathing begins
- C.** Start coordinated chest compressions while continuing effective ventilation
- D.** Continue ventilation alone for several additional minutes

**33.** A 900-g infant receives 10% dextrose intravenously at 4 mL/kg/h. What glucose infusion rate is being delivered?

- A.** Approximately 6.7 mg/kg/min
- B.** Approximately 16.7 mg/kg/min
- C.** Approximately 0.7 mg/kg/min
- D.** Approximately 40 mg/kg/min

**34.** A 25-week infant receives almost no enteral nutrition for several weeks because of severe illness. Which factor contributes to parenteral-nutrition-associated liver disease?

- A.** Phototherapy causing obstruction of the extrahepatic bile ducts
- B.** Human milk exposure causing direct hepatotoxicity
- C.** Caffeine therapy causing universal neonatal cholestasis
- D.** Lack of enteral stimulation together with prolonged parenteral nutrient exposure

**35.** A 26-week infant has a large urine output and rising serum sodium on day 2. Which management principle is most appropriate?

- A.** Ignore daily weight because extracellular fluid never changes after birth
- B.** Use one fixed fluid volume for every preterm infant throughout the first week
- C.** Adjust fluids using serial weight, sodium, urine output, and environmental losses
- D.** Correct sodium rapidly without considering ongoing water losses

**36.** A newborn has hypotonia, poor feeding, genital hypoplasia, and later develops a strong tendency toward hyperphagia. Which imprinting disorder is most likely?

- A.** Prader-Willi syndrome
- B.** Beckwith-Wiedemann syndrome
- C.** Angelman syndrome
- D.** Noonan syndrome

**37.** A newborn receives positive-pressure ventilation, but the heart rate remains 70/min and the chest is not moving. What is the best immediate response?

- A.** Begin volume expansion without reassessing ventilation technique
- B.** Perform ventilation corrective steps to establish visible chest movement
- C.** Stop ventilation and provide free-flow oxygen alone
- D.** Proceed directly to synchronized cardioversion for presumed arrhythmia

**38.** Prenatal polyhydramnios was noted. After birth, an infant pools saliva and chokes with feeding; radiography shows a feeding tube coiled in the upper mediastinum with air in the stomach. Which congenital anomaly is most likely?

- A.** Hypertrophic pyloric stenosis causing gastric outlet obstruction
- B.** Esophageal atresia with or without tracheoesophageal fistula
- C.** Isolated gastroesophageal reflux without anatomic interruption
- D.** Hirschsprung disease with distal functional obstruction

**39.** Point-of-care lung ultrasound is performed for suspected pneumothorax. Which finding most specifically supports pneumothorax when identified at the transition between normal and abnormal pleura?

- A.** Visible lung sliding throughout the scanned field
- B.** A normal seashore sign on M-mode
- C.** Diffuse bilateral B-lines with normal sliding
- D.** A discrete lung point at the transition zone

**40.** A newborn with cystic fibrosis develops abdominal distention and failure to pass meconium. Radiography shows a soap-bubble appearance in the right lower quadrant. Which diagnosis is most likely?

- A.** Necrotizing enterocolitis
- B.** Biliary atresia
- C.** Pyloric stenosis
- D.** Meconium ileus

**41.** A delivery-room team is performing coordinated compressions and ventilations for persistent severe neonatal bradycardia after effective ventilation. To deliver about 90 compressions and 30 inflations each minute, which compression-to-ventilation ratio should be used?

- A.** Thirty compressions to two ventilations
- B.** Five compressions to one ventilation
- C.** Three compressions to one ventilation
- D.** Fifteen compressions to two ventilations

**42.** A term newborn has profound cyanosis from d-transposition of the great arteries despite prostaglandin infusion. Echocardiography shows a very restrictive atrial communication. Which intervention most directly improves systemic oxygenation?

- A.** Diuretic therapy to reduce pulmonary blood flow
- B.** Balloon atrial septostomy to improve interatrial mixing
- C.** Pharmacologic closure of the ductus arteriosus
- D.** Immediate ligation of the foramen ovale

**43.** A newborn has conotruncal heart disease, hypocalcemia, and absent thymic tissue on imaging. Which genetic diagnosis is most likely?

- A.** Achondroplasia from an FGFR3-related skeletal dysplasia
- B.** Prader-Willi syndrome from chromosome 15 imprinting
- C.** Trisomy 18, also called Edwards syndrome
- D.** Chromosome 22q11.2 deletion syndrome

**44.** A newborn has in utero exposure to HIV. Maternal HIV RNA was below 50 copies/mL from 20 weeks of gestation through delivery with sustained treatment adherence. Which neonatal antiretroviral approach is appropriate for this low-risk situation?

- A.** Zidovudine prophylaxis alone for a short course beginning soon after birth
- B.** Delay prophylaxis until the first infant antibody test becomes positive
- C.** No antiretroviral medication because maternal therapy eliminates all residual risk
- D.** A three-drug presumptive treatment regimen for every exposed infant regardless of risk

**45.** A healthy term infant develops mild unconjugated jaundice at 48 hours, feeds well, and has a normal examination. Bilirubin rises slowly and then begins to fall. Which process is most likely?

- A.** Physiologic neonatal hyperbilirubinemia
- B.** Severe hemolytic disease with rapid bilirubin rise
- C.** Neonatal hepatitis causing conjugated hyperbilirubinemia
- D.** Biliary atresia

**46.** A newborn appears well initially but develops shock at 36 hours. The right arm blood pressure exceeds the leg pressure, and femoral pulses are weak. Which diagnosis is most likely?

- A.** Uncomplicated patent foramen ovale
- B.** Small muscular ventricular septal defect
- C.** Isolated secundum atrial septal defect
- D.** Critical coarctation of the aorta

**47.** A 5-day-old infant has fever, lethargy, seizures, and a bulging fontanelle. Lumbar puncture shows neutrophilic pleocytosis, low CSF glucose, and high protein. Which process is most likely?

- A.** Simple physiologic jaundice
- B.** Transient tachypnea of the newborn
- C.** Apnea of prematurity
- D.** Bacterial meningitis

**48.** A female newborn has diffuse hand and foot lymphedema, a webbed neck, widely spaced nipples, and coarctation of the aorta. Which diagnosis is most likely?

- A.** Noonan syndrome in a male infant
- B.** Trisomy 21
- C.** Turner syndrome
- D.** Klinefelter syndrome

**49.** A newborn has severe anemia, hydrops, and high-output cardiac failure. Maternal infection with parvovirus B19 occurred several weeks earlier. What fetal cell line is primarily affected?

- A.** Pancreatic beta cells
- B.** Mature neutrophils only
- C.** Erythroid precursor cells
- D.** Megakaryocytes only

**50.** A 25-week infant has recurrent apnea with bradycardia after infection, anemia, glucose disturbance, and temperature instability have been excluded. Which medication is commonly used to reduce these events?

- A.** Caffeine citrate to stimulate respiratory drive
- B.** Prostaglandin E1 to maintain respiratory rhythm
- C.** Indomethacin to increase central chemoreceptor sensitivity
- D.** Phenobarbital to stimulate medullary ventilation

**51.** A newborn has jaundice, hepatomegaly, vomiting after milk feeds, hypoglycemia, and Escherichia coli sepsis. Which inborn error should be considered urgently?

- A.** Medium-chain acyl-CoA dehydrogenase deficiency
- B.** Maple syrup urine disease
- C.** Phenylketonuria
- D.** Classic galactosemia

**52.** A mother reports frequent exposure to cat litter and undercooked meat during pregnancy. Her newborn has ventriculomegaly, chorioretinal inflammation, and intracranial calcifications that are not limited to the periventricular region. Which congenital infection is most likely?

- A.** Rubella
- B.** Toxoplasmosis
- C.** Syphilis
- D.** Cytomegalovirus

**53.** A 25-week infant is receiving supplemental oxygen for evolving chronic lung disease. Which principle best minimizes oxygen-related injury while maintaining safety?

- A.** Use a fixed high oxygen concentration regardless of saturation
- B.** Avoid monitoring because alarm limits increase oxygen exposure
- C.** Keep saturation at 100% continuously to prevent all hypoxemic events
- D.** Use pulse oximetry to titrate oxygen within a prescribed target range

**54.** On day 9, a 46,XX newborn with clitoromegaly presents in shock. Laboratory testing shows sodium 124 mEq/L, potassium 7.1 mEq/L, low cortisol, and markedly elevated 17-hydroxyprogesterone. Which disorder is most likely?

- A.** Salt-wasting 21-hydroxylase-deficient congenital adrenal hyperplasia
- B.** Primary hyperaldosteronism causing sodium retention
- C.** Central diabetes insipidus causing free-water loss
- D.** Congenital hypothyroidism without adrenal failure

**55.** During normal transition after birth, what change promotes functional constriction of the ductus arteriosus?

- A.** Increasing pulmonary vascular resistance during lung aeration
- B.** Falling arterial oxygen tension together with rising prostaglandin levels
- C.** Rising arterial oxygen tension together with falling circulating prostaglandin levels
- D.** Persistent placental flow maintaining low systemic vascular resistance

**56.** A 27-week infant has frequent gastric residuals but a soft abdomen, normal stooling, stable vital signs, and no radiographic signs of necrotizing enterocolitis. What is the best interpretation?

- A.** Residuals always indicate congenital bowel obstruction
- B.** Gastric residual volume alone is an imperfect marker of feeding intolerance
- C.** Residuals prove cow-milk protein allergy even before milk exposure
- D.** Any residual proves necrotizing enterocolitis and requires surgery

**57.** A 2-hour-old newborn has weak femoral pulses, metabolic acidosis, and worsening perfusion as the ductus begins to constrict. Critical left-sided obstructive heart disease is suspected. What therapy should be started immediately?

- A.** Furosemide as the sole treatment for systemic hypoperfusion
- B.** Adenosine to slow atrioventricular conduction
- C.** Indomethacin to accelerate ductal closure
- D.** Prostaglandin E1 infusion to maintain ductal patency

**58.** A 3-day-old infant develops poor feeding, abnormal tone, and a sweet odor to the urine. Plasma amino-acid testing shows elevated branched-chain amino acids. Which diagnosis is most likely?

- A.** Maple syrup urine disease
- B.** Galactosemia
- C.** Tyrosinemia type I
- D.** Urea-cycle defect

**59.** An extremely preterm infant's incubator is left open during a procedure. The axillary temperature falls, glucose consumption rises, and oxygen demand increases despite no change in lung disease. Which response to cold stress explains these changes?

- A.** Decreased oxygen consumption because brown fat is inactive
- B.** Increased oxygen and glucose consumption from nonshivering thermogenesis
- C.** Reduced risk of hypoglycemia because metabolic demand falls
- D.** Suppression of catecholamine release and lipolysis

**60.** A male newborn has a distended bladder, bilateral hydroureteronephrosis, and a weak urinary stream. Which obstructive lesion is most likely?

- A.** Multicystic dysplastic kidney
- B.** Renal vein thrombosis
- C.** Posterior urethral valves
- D.** Unilateral ureteropelvic junction obstruction

**61.** A preterm infant is receiving prolonged exclusive breast-milk feeding. Which micronutrient commonly requires supplementation because body stores are limited and needs are high?

- A.** Daily high-dose vitamin K supplementation
- B.** Routine supplemental elemental iron
- C.** Routine fluoride during the first week of life
- D.** High-dose oral vitamin A for every growing preterm infant

**62.** The parents of a hospitalized newborn have limited English proficiency. A complex consent discussion is planned. Which communication strategy is best?

- A.** Speak more loudly in English without interpretation
- B.** Provide the consent form only and omit verbal discussion
- C.** Rely on an older sibling to interpret complex risk information
- D.** Use a qualified medical interpreter

**63.** A preterm infant develops early-onset sepsis after prolonged rupture of membranes. Blood culture grows a gram-negative bacillus that is a leading cause of early sepsis in very preterm infants. Which organism is most likely?

- A.** *Candida albicans*
- B.** *Staphylococcus epidermidis*
- C.** *Bordetella pertussis*
- D.** *Escherichia coli*

**64.** A diagnostic test with unchanged sensitivity and specificity is introduced into a population in which the target disease is much more common. What generally happens to the test's positive predictive value?

- A.** The positive predictive value generally increases
- B.** It becomes identical to sensitivity
- C.** It necessarily falls to zero
- D.** It is completely independent of disease prevalence

**65.** A term infant with a moderate ventricular septal defect is asymptomatic at birth but develops tachypnea and poor weight gain several weeks later. Why do symptoms often emerge after the immediate newborn period?

- A.** The foramen ovale enlarges and creates severe right-to-left shunting
- B.** Systemic vascular resistance progressively falls below fetal levels
- C.** The ductus venosus reopens and diverts blood away from the liver
- D.** Pulmonary vascular resistance falls, increasing left-to-right shunt volume

**66.** A newborn has cyanosis and diminished pulmonary blood flow. Echocardiography shows severe pulmonary stenosis and a large ventricular septal defect. Which bedside intervention may transiently improve a hypercyanotic spell by increasing systemic vascular resistance?

- A.** Placing the infant flat with legs extended to reduce venous return
- B.** Knee-chest positioning while definitive support is organized
- C.** Closing any ductal connection immediately with indomethacin
- D.** Giving a pure arterial vasodilator to lower systemic resistance

**67.** A term infant has persistent hypoglycemia, micropenis, and cholestasis. Which endocrine problem should be considered?

- A.** Primary hyperthyroidism
- B.** Isolated aldosterone excess
- C.** Transient neonatal hyperparathyroidism
- D.** Congenital hypopituitarism

**68.** A 27-week infant has a continuous murmur, bounding pulses, widened pulse pressure, and increasing ventilator requirements during the second week. Which lesion is most likely?

- A.** Coarctation of the aorta with ductal closure
- B.** Hemodynamically important patent ductus arteriosus
- C.** Critical pulmonary valve stenosis
- D.** Tetralogy of Fallot with reduced pulmonary flow

**69.** A 3-week-old infant has progressive nonbilious projectile vomiting, remains hungry after emesis, and has hypochloremic metabolic alkalosis. Which diagnosis is most likely?

- A.** Malrotation with midgut volvulus
- B.** Duodenal atresia
- C.** Necrotizing enterocolitis
- D.** Hypertrophic pyloric stenosis

**70.** A 24-week infant requires mechanical ventilation. Which ventilator-related exposure is most directly associated with volutrauma?

- A.** Low airway pressure causing direct alveolar rupture
- B.** Low inspired oxygen concentration causing oxidative injury
- C.** Excessive tidal volume causing overdistension of aerated lung units
- D.** Short expiratory time causing only systemic hypotension

**71.** An infant with an imperforate anus is also found to have a tracheoesophageal fistula, unilateral renal agenesis, and a hypoplastic radius. Which association should be suspected and prompt evaluation for vertebral and cardiac defects?

- A.** Beckwith-Wiedemann syndrome
- B.** CHARGE syndrome
- C.** Turner syndrome
- D.** VACTERL association

**72.** A newborn has sepsis and meningitis. The maternal history includes a febrile gastrointestinal illness during pregnancy. Which organism classically crosses the placenta and can cause neonatal sepsis?

- A.** *Helicobacter pylori*
- B.** *Mycoplasma pneumoniae*
- C.** *Listeria monocytogenes*
- D.** *Clostridioides difficile*

**73.** A newborn has unilateral leukocoria on examination. Which diagnosis must be excluded urgently because delayed recognition can threaten both vision and life?

- A.** Retinoblastoma involving the affected eye
- B.** Physiologic retinal vascularization
- C.** Transient neonatal pustular melanosis
- D.** Nasolacrimal duct obstruction

**74.** A term small-for-gestational-age infant is plethoric, jittery, and has a venous hematocrit of 72%. Which syndrome is most likely?

- A.** Vitamin K deficiency bleeding with coagulopathy
- B.** Neonatal polycythemia with hyperviscosity
- C.** Congenital factor VIII deficiency, or hemophilia A
- D.** Anemia of prematurity with low erythropoietin

**75.** A preterm infant has frequent desaturations during feeds but is otherwise stable. Chest radiography is unchanged, and episodes occur with coughing and milk in the pharynx. Which problem should be considered?

- A.** Ductal-dependent pulmonary blood flow triggered by sucking
- B.** New respiratory distress syndrome from recurrent surfactant deficiency
- C.** Swallowing dysfunction with aspiration during oral feeding
- D.** Tension pneumothorax occurring only during each bottle feed

**76.** A term infant requires positive-pressure ventilation at birth but has no known cyanotic heart disease. What initial oxygen concentration is generally appropriate?

- A.** Withhold oxygen and ventilation until pulse oximetry provides a reading
- B.** Begin with 100% oxygen for every term infant requiring ventilation
- C.** Begin with room air and titrate oxygen to preductal saturation targets
- D.** Begin with 60% oxygen and avoid subsequent titration

77. A newborn's heart rate remains below 60/min despite effective ventilation and coordinated compressions. Vascular access is available through an umbilical venous catheter. Which medication is indicated?

- A. Adenosine given for presumed sinus bradycardia
- B. Epinephrine given by the intravenous route
- C. Indomethacin given to increase systemic vascular resistance
- D. Atropine given routinely for birth-related bradycardia

78. A former 24-week infant is now 37 weeks postmenstrual age and still requires supplemental oxygen after months of mechanical and noninvasive respiratory support. Chest imaging shows coarse interstitial change and areas of hyperinflation. Which chronic pulmonary disorder best fits this course?

- A. Transient tachypnea of the newborn from retained fetal lung fluid
- B. Acute meconium aspiration syndrome beginning at 36 weeks postmenstrual age
- C. Isolated congenital lobar emphysema acquired from oxygen therapy
- D. Bronchopulmonary dysplasia related to arrested lung development and injury

79. A newborn delivered after shoulder dystocia holds the affected arm adducted and internally rotated with the elbow extended. The grasp reflex is intact. Which lesion is most likely?

- A. Isolated radial nerve injury
- B. Upper brachial plexus injury involving C5-C6
- C. Spinal muscular atrophy
- D. Lower brachial plexus injury involving C8-T1

**80.** A NICU team wants to improve admission temperatures in extremely preterm infants. They test a small change for one week, review the results, modify the process, and test again. Which quality-improvement method does this illustrate?

- A.** A retrospective case report
- B.** Iterative Plan-Do-Study-Act quality-improvement cycles
- C.** A fixed protocol that cannot be modified after implementation
- D.** A single blinded randomized trial as the only acceptable method

**81.** A 25-week infant has a routine cranial ultrasound on day 7 that shows echogenic blood confined to the germinal matrix without ventricular dilatation. Which diagnosis best describes this finding?

- A.** Cystic periventricular leukomalacia
- B.** Cerebellar hypoplasia
- C.** Posthemorrhagic hydrocephalus
- D.** Grade I germinal-matrix hemorrhage

**82.** A pregnant patient at 28 weeks is likely to deliver within 24 hours because of preterm labor. Which antenatal intervention most directly lowers the newborn's risk of respiratory distress syndrome?

- A.** Routine maternal oxygen therapy until delivery
- B.** Immediate maternal indomethacin solely to stimulate surfactant release
- C.** Maternal loop diuretic treatment to reduce fetal lung water
- D.** Antenatal corticosteroid treatment to accelerate fetal lung maturation

**83.** A preterm infant on glucocorticoids develops sudden abdominal distention and free intraperitoneal air but has little bowel-wall pneumatosis and less systemic inflammation than expected for necrotizing enterocolitis. Which diagnosis is most likely?

- A.** Hirschsprung disease
- B.** Pyloric stenosis
- C.** Spontaneous intestinal perforation
- D.** Uncomplicated feeding intolerance

**84.** A cyanotic newborn has a single loud second heart sound. Echocardiography shows one large arterial trunk arising from the ventricles and supplying systemic, pulmonary, and coronary circulations. What is the diagnosis?

- A.** Pulmonary atresia with intact ventricular septum
- B.** Isolated coarctation of the aorta
- C.** Persistent truncus arteriosus
- D.** d-Transposition of the great arteries

**85.** A preterm infant has a large unilateral intraventricular hemorrhage followed by a persistent echogenic lesion in the adjacent periventricular white matter. Which mechanism best explains this lesion?

- A.** Primary bacterial abscess formation
- B.** Venous infarction from impaired drainage adjacent to the hemorrhage
- C.** Global cortical necrosis caused by isolated hypoglycemia
- D.** Congenital absence of the cerebral hemisphere

**86.** An infant born to a mother with untreated positive syphilis serology develops hepatomegaly, persistent nasal discharge, painful limitation of limb movement, and a desquamating rash on the palms and soles. Which infection is most likely?

- A.** Neonatal candidiasis
- B.** Congenital syphilis
- C.** Congenital toxoplasmosis
- D.** Congenital rubella

**87.** A term infant has progressive respiratory distress. Imaging shows marked hyperinflation of one upper lobe with compression of adjacent lung but no pleural air. Which congenital lesion is most likely?

- A.** Bilateral pulmonary hypoplasia
- B.** Diffuse respiratory distress syndrome
- C.** Congenital lobar overinflation
- D.** Tension pneumothorax

**88.** A term infant is pale and hypotensive after an emergency delivery for a large placental abruption. Cord hemoglobin is markedly reduced. Which mechanism best explains the newborn's shock?

- A.** Isolated neonatal sepsis beginning before membrane rupture
- B.** Physiologic fall in hemoglobin expected after the first month
- C.** Chronic neonatal iron deficiency developing during labor
- D.** Acute fetal blood loss associated with placental separation

**89.** An observational cohort comparing an exposure with no exposure finds a relative risk of 0.70 for an adverse neonatal outcome, with a 95% confidence interval from 0.50 to 0.98. Which statement correctly interprets this estimate?

- A.** The association is compatible with reduced risk because the 95% confidence interval excludes 1
- B.** The confidence interval means 95% of individual patients have risks between 0.50 and 0.98
- C.** The result proves causation regardless of study design
- D.** There is no possible treatment effect because the point estimate is below 1

**90.** A 29-week infant is receiving full volumes of human milk but has poor protein and mineral intake for growth. What is the usual nutritional strategy?

- A.** Dilute the milk with sterile water to reduce osmolality
- B.** Add a human-milk fortifier to increase nutrient density
- C.** Replace all human milk with fruit juice
- D.** Stop enteral feeding and use parenteral glucose alone

**91.** A very preterm infant is receiving human milk with fortifier. Which nutrient pair is particularly important for supporting skeletal mineralization?

- A.** Copper and selenium alone
- B.** Calcium and phosphorus
- C.** Sodium and chloride alone
- D.** Vitamin K and folate alone

**92.** A newborn has an absent red reflex in one eye. The cornea is clear, and a white opacity is visible behind the pupil. Which next step is most appropriate?

- A.** Urgent ophthalmologic evaluation for congenital cataract
- B.** Routine reassessment at the 2-year well-child visit
- C.** No follow-up because unilateral findings do not affect vision
- D.** Empiric topical antibiotic treatment only

**93.** A former 25-week infant develops persistent systemic hypertension after prolonged umbilical arterial catheterization. Which complication should be considered first?

- A.** Isolated patent ductus arteriosus causing elevated diastolic pressure
- B.** Pulmonary vein stenosis causing systemic arterial hypertension
- C.** Physiologic postnatal sodium loss causing chronic hypertension
- D.** Renal arterial thrombosis causing renovascular hypertension

**94.** Parents decline a clearly life-saving treatment for their newborn because of beliefs that would expose the infant to a high likelihood of preventable death. What is the neonatologist's primary ethical obligation?

- A.** Secretly provide treatment without documenting the disagreement
- B.** Protect the infant's best interests while seeking ethics and legal support as needed
- C.** Accept every parental refusal regardless of the degree of preventable harm
- D.** Discharge the infant immediately without further discussion

**95.** A newborn has a midline abdominal-wall defect at the base of the umbilical cord with viscera contained within a translucent sac. Which additional evaluation is particularly important?

- A.** Delay examination until after complete enteral feeding is established
- B.** Screen only for pyloric stenosis
- C.** Assessment for associated cardiac and chromosomal anomalies
- D.** Assume the defect is isolated and avoid further evaluation

**96.** How does intensive phototherapy primarily reduce unconjugated bilirubin in the newborn?

- A.** It permanently increases hepatic UDP-glucuronosyltransferase gene expression within minutes
- B.** It converts bilirubin into more water-soluble photoisomers that can be excreted without conjugation
- C.** It increases red-cell production to dilute bilirubin
- D.** It mechanically removes bilirubin through the skin

**97.** A 29-week infant on advancing feeds develops recurrent apnea, metabolic acidosis, abdominal wall erythema, and bloody stool. Abdominal imaging shows intramural bowel gas and portal venous gas. Which disease process is most likely?

- A.** Necrotizing enterocolitis
- B.** Uncomplicated gastroesophageal reflux
- C.** Physiologic meconium passage
- D.** Hypertrophic pyloric stenosis

**98.** A newly placed umbilical venous catheter courses cephalad on radiography but its tip projects within the liver rather than near the diaphragm. What is the main concern?

- A.** The catheter is ideally positioned for hyperosmolar parenteral nutrition
- B.** The finding confirms correct placement in the superior vena cava
- C.** The catheter is in the descending aorta and should be advanced further
- D.** The tip is in the portal system and is unsafe for central hyperosmolar infusion

**99.** A previously well newborn becomes lethargic after a period of poor intake. Laboratory studies show hypoketotic hypoglycemia. Which metabolic defect is most consistent with this pattern?

- A.** Classic galactosemia after complete lactose avoidance
- B.** Medium-chain acyl-CoA dehydrogenase deficiency
- C.** Classic phenylketonuria from phenylalanine hydroxylase deficiency
- D.** Congenital primary hypothyroidism

**100.** An extremely preterm infant has recurrent apnea of prematurity and is started on caffeine citrate. Which mechanism contributes most directly to caffeine's respiratory-stimulant effect?

- A.** Selective beta-1 adrenergic blockade
- B.** Direct surfactant replacement
- C.** Adenosine-receptor antagonism
- D.** Irreversible opioid-receptor blockade

**101.** A 28-week infant is growing poorly despite adequate calculated calories. The infant has chronic lung disease and marked tachypnea. Why may energy needs be higher than expected?

- A.** Supplemental oxygen prevents any additional caloric need
- B.** Tachypnea lowers metabolic demand below basal levels
- C.** Increased work of breathing raises energy expenditure
- D.** Chronic lung disease eliminates all protein requirements

**102.** A newborn with hypoplastic left heart syndrome becomes gray and acidotic as the ductus constricts. Which circulatory pathway is essential for systemic perfusion before surgery?

- A.** Coronary venous blood flowing directly into the descending aorta
- B.** Right-ventricular output crossing the ductus into the systemic circulation
- C.** Pulmonary venous blood bypassing the atria through the ductus venosus
- D.** Left-ventricular output crossing an intact aortic valve into the systemic circulation

**103.** A newborn is encased in thick plate-like scales with deep fissures, ectropion, and respiratory compromise from restricted chest movement. Which disorder is most likely?

- A.** Harlequin ichthyosis
- B.** Nevus simplex
- C.** Miliaria rubra
- D.** Erythema toxicum neonatorum

**104.** A growth-restricted preterm infant born to a mother with severe hypertension has neutropenia on the first day but is clinically well. Which association is most likely?

- A.** Transient neonatal neutropenia associated with maternal hypertensive disease
- B.** Congenital HIV infection as the only explanation
- C.** Late-onset catheter sepsis before catheter placement
- D.** Physiologic neutrophilia of labor misreported as neutropenia

**105.** A preterm infant on prolonged parenteral nutrition has hypertriglyceridemia after lipid advancement. Which response is most appropriate?

- A.** Add free water only and ignore the triglyceride concentration
- B.** Stop all protein because amino acids cause isolated hypertriglyceridemia
- C.** Reassess lipid dose and clearance before further advancement
- D.** Increase lipid infusion immediately to improve triglyceride clearance

**106.** A fetus with known gastroschisis is delivered at term. The exposed bowel is edematous, and the infant is rapidly losing heat and fluid while the operating room is prepared. Which delivery-room management bundle is most appropriate?

- A.** Cover bowel in sterile plastic, decompress stomach, give IV fluids, and call surgery
- B.** Attempt forceful bedside reduction without monitoring perfusion
- C.** Apply dry gauze tightly and begin immediate enteral feeding
- D.** Leave the bowel exposed to air to reduce edema

**107.** After the initial steps of resuscitation, a newly born infant remains apneic with a heart rate of 80/min. What should be done next?

- A.** Give intravenous epinephrine before establishing ventilation
- B.** Start chest compressions before attempting ventilation
- C.** Begin effective positive-pressure ventilation and assess chest movement
- D.** Observe for another minute because the heart rate exceeds 60/min

**108.** A 38-week infant delivered by elective cesarean without labor develops tachypnea shortly after birth. Oxygen need is modest, chest radiography shows prominent perihilar streaking, and symptoms improve over 36 hours. What is the most likely diagnosis?

- A.** Respiratory distress syndrome from severe surfactant deficiency
- B.** Bacterial pneumonia with progressive parenchymal destruction
- C.** Transient tachypnea of the newborn from delayed lung-fluid clearance
- D.** Congenital diaphragmatic hernia with pulmonary hypoplasia

**109.** A newborn who did not receive vitamin K prophylaxis develops gastrointestinal and umbilical bleeding. PT is markedly prolonged and platelets are normal. Which diagnosis is most likely?

- A.** Fetal and neonatal alloimmune thrombocytopenia
- B.** Disseminated intravascular coagulation
- C.** Congenital thrombocytopenia-absent radius syndrome
- D.** Vitamin K deficiency bleeding

**110.** An infant with rapidly rising unconjugated bilirubin becomes lethargic, hypotonic, and develops a high-pitched cry. The bilirubin is at the exchange transfusion threshold despite intensive phototherapy. What is the most appropriate next step?

- A.** Prepare for urgent exchange transfusion while continuing intensive phototherapy
- B.** Discharge with routine follow-up because jaundice is physiologic
- C.** Give only additional enteral water to lower the bilirubin
- D.** Stop phototherapy because neurologic signs indicate treatment failure

**111.** A fetus has prolonged severe oligohydramnios after very early rupture of membranes. The newborn has small lungs and severe respiratory failure at birth. What mechanism best explains the respiratory disease?

- A.** Isolated neonatal aspiration of amniotic fluid at delivery
- B.** Primary pulmonary venous hypertension from left-sided obstruction
- C.** Pulmonary hypoplasia from impaired lung growth in a low-fluid environment
- D.** Excess surfactant production causing alveolar overdistension

**112.** On day 2, a vigorous term infant develops fleeting erythematous patches containing tiny pustules. The palms and soles are spared, and a smear from a pustule contains many eosinophils. Which benign neonatal eruption is most likely?

- A.** Staphylococcal scalded skin syndrome
- B.** Congenital candidiasis with systemic illness
- C.** Disseminated herpes simplex infection
- D.** Erythema toxicum neonatorum

**113.** A newborn has hypertelorism, low-set ears, a webbed neck, pectus deformity, and pulmonary valve stenosis. Karyotype is 46,XY. Which syndrome is most likely?

- A.** Turner syndrome
- B.** Noonan syndrome
- C.** Beckwith-Wiedemann syndrome
- D.** Trisomy 13

**114.** A term infant has persistent conjugated hyperbilirubinemia, pale stools, and dark urine. Which diagnosis must be excluded promptly because earlier intervention improves outcome?

- A.** Biliary atresia
- B.** ABO hemolytic disease
- C.** Breast-milk jaundice
- D.** Physiologic unconjugated jaundice

**115.** A newborn screen shows absent T-cell receptor excision circles. The infant has persistent lymphopenia. Which disorder is the screening program designed to detect?

- A.** Selective IgA deficiency
- B.** Complement C5 deficiency
- C.** Severe combined immunodeficiency
- D.** Chronic granulomatous disease

**116.** In a trial, the rate of a poor outcome is 20% with standard therapy and 10% with a new therapy. What is the number needed to treat to prevent one additional poor outcome?

- A.** 5
- B.** 10
- C.** 2
- D.** 20

**117.** A jaundiced preterm infant requires treatment for a suspected serious bacterial infection. Which cephalosporin is generally avoided in young neonates because of bilirubin displacement and potentially dangerous calcium-ceftriaxone precipitation?

- A.** Cephalexin
- B.** Cefazolin
- C.** Ceftriaxone
- D.** Cefepime in every circumstance

**118.** A preterm infant dependent on parenteral nutrition develops direct hyperbilirubinemia after several weeks. Which complication is most likely?

- A.** Physiologic unconjugated jaundice of the first week
- B.** Breastfeeding jaundice from low milk intake
- C.** Isolated hemolysis from ABO incompatibility
- D.** Parenteral-nutrition-associated cholestasis

**119.** A newborn has profound hypotonia, facial diplegia, respiratory failure, and difficulty feeding. The mother has a history of grip myotonia and early cataracts. Which diagnosis best explains the infant's presentation?

- A.** Congenital myotonic dystrophy
- B.** Transient hypermagnesemia from maternal therapy only
- C.** Duchenne muscular dystrophy
- D.** Isolated neonatal botulism

**120.** A male neonate develops a large cephalohematoma and prolonged oozing after heel sticks. Platelets and PT are normal, the aPTT is prolonged, and several maternal male relatives have excessive bleeding. Which inherited disorder is most likely?

- A.** Congenital factor VIII deficiency, or hemophilia A
- B.** Disseminated intravascular coagulation
- C.** Fetal and neonatal alloimmune thrombocytopenia
- D.** Vitamin K deficiency bleeding

**121.** A male newborn has severe hypotonia, tongue fasciculations, weak cry, and absent deep tendon reflexes. Genetic testing confirms biallelic loss of SMN1. Which disorder is present?

- A.** Prader-Willi syndrome
- B.** Hypoxic-ischemic encephalopathy
- C.** Congenital myotonic dystrophy
- D.** Spinal muscular atrophy

**122.** A stable 27-week infant is starting enteral feeds. Which milk source is preferred when available because it is associated with lower rates of important preterm intestinal morbidity?

- A.** The infant's own mother's milk
- B.** Routine sterile water feeds before any milk
- C.** Unmodified cow's milk intended for older infants
- D.** Standard term formula as the preferred first choice

**123.** A new bedside test for neonatal sepsis is positive in 90 of 100 infants who truly have sepsis. Which test characteristic is 90% in this example?

- A.** Positive predictive value
- B.** Specificity
- C.** Negative predictive value
- D.** Sensitivity of the test

**124.** A newborn fails the hearing screen and has microcephaly, thrombocytopenia, hepatosplenomegaly, and periventricular calcifications on cranial imaging. Which congenital infection is the leading diagnosis?

- A.** Cytomegalovirus
- B.** Parvovirus B19
- C.** Toxoplasma gondii
- D.** Congenital syphilis

**125.** A study concludes that two treatments differ when in truth there is no real difference. Which statistical error has occurred?

- A.** Selection bias only
- B.** Type II error
- C.** A type I, or alpha, error
- D.** Confounding by indication only

**126.** A newborn receives a continuous morphine infusion after surgery and becomes increasingly sedated when renal and hepatic function deteriorate. Which developmental pharmacology principle is most relevant?

- A.** Neonates always eliminate morphine faster than older children
- B.** Immature and impaired clearance can prolong opioid exposure in neonates
- C.** Protein binding alone guarantees rapid drug removal
- D.** Morphine has no active metabolites and is independent of organ function

**127.** A very preterm infant later develops bilateral cystic lesions in the periventricular white matter. Which long-term motor outcome is most strongly associated with this injury pattern?

- A.** Isolated lower motor neuron weakness
- B.** Progressive muscular dystrophy
- C.** Spastic diplegic cerebral palsy
- D.** Pure sensory neuropathy

**128.** In a randomized trial, several participants cross over from their assigned treatment to the other group. The primary analysis keeps participants in the groups to which they were originally randomized. What is this analysis called?

- A.** Intention-to-treat analysis
- B.** Case-control matching
- C.** Per-protocol analysis
- D.** Ecologic analysis

**129.** A breastfed preterm infant reaches full enteral feeds but has inadequate weight gain. Which assessment is most useful before simply increasing fluid volume?

- A.** Stop fortification because fortified milk cannot support growth
- B.** Review actual energy, protein, and mineral intake relative to growth trajectory
- C.** Increase free water without evaluating nutrient density
- D.** Assume all poor growth reflects congenital infection

**130.** Serum immunoglobulin testing at birth shows much less IgG in a 25-week infant than in a term newborn, despite normal maternal IgG levels. Which feature of placental antibody transport best explains the difference?

- A.** Preterm infants catabolize all transferred IgG before birth
- B.** Placental IgG transfer increases markedly during the third trimester
- C.** Maternal IgG synthesis stops during preterm labor
- D.** Maternal IgG cannot cross the placenta at any gestational age

**131.** A preterm infant is transitioning from parenteral to enteral nutrition. Which finding best supports advancing enteral intake?

- A.** Complete absence of bowel sounds in a deteriorating infant
- B.** One isolated green gastric residual regardless of the rest of the examination
- C.** Stable abdominal examination and overall clinical tolerance over time
- D.** New abdominal discoloration with systemic instability

**132.** A neonate requires diagnostic lumbar puncture. Which interspace is generally chosen to minimize risk of spinal-cord injury?

- A.** Upper thoracic T2-T3 interspace
- B.** Lower lumbar L3-L4 or L4-L5 interspace
- C.** High cervical C1-C2 interspace
- D.** Lower thoracic T8-T9 interspace

**133.** A newborn has rhizomelic shortening of the limbs, frontal bossing, and midface hypoplasia. Which skeletal dysplasia is most likely?

- A.** Achondroplasia due to an FGFR3-related skeletal dysplasia
- B.** Thanatophoric dysplasia with a narrow thorax
- C.** Osteogenesis imperfecta type II
- D.** Campomelic dysplasia with bowed long bones

**134.** A newborn is delivered to a mother who is hepatitis B surface antigen positive. What preventive strategy should be provided promptly after birth?

- A.** Hepatitis B vaccine plus hepatitis B immune globulin at separate sites
- B.** Give hepatitis A vaccine because it cross-protects against hepatitis B
- C.** Delay all hepatitis B prophylaxis until the 2-month visit
- D.** Give hepatitis B immune globulin alone without vaccination

**135.** Humidity in the incubator of a 24-week infant is inadvertently kept too low. Over 48 hours the infant loses excessive weight and develops hypernatremia despite adequate urine output. Which physiologic property is the major contributor?

- A.** Complete absence of evaporative water loss
- B.** High transepidermal and other insensible water losses
- C.** Obligate water retention caused by mature skin
- D.** Exceptionally effective renal concentrating ability

**136.** A term infant exposed to chronic maternal fentanyl use develops tremors, high-pitched crying, loose stools, and difficulty consoling over the first 2 days. Which diagnosis best fits this pattern?

- A.** Congenital hypothyroidism with reduced sympathetic activity
- B.** Early bacterial meningitis without systemic inflammation
- C.** Hypoxic-ischemic encephalopathy with evolving cerebral edema
- D.** Neonatal opioid withdrawal syndrome

**137.** A fetus is noted to have persistent bradycardia. After birth, the ECG shows complete atrioventricular block. The mother is asymptomatic but has anti-Ro antibodies. What is the mechanism?

- A.** Fetal hyperkalemia from maternal diet causing fixed conduction block
- B.** Transplacental autoantibody injury to the fetal conduction system
- C.** Transplacental thyroid hormone causing atrioventricular nodal fibrosis
- D.** Maternal beta-blocker exposure causing permanent atrioventricular scarring

**138.** After a central nervous system infection, a ventilated neonate develops serum sodium of 124 mEq/L and low serum osmolality. Urine remains inappropriately concentrated with substantial urinary sodium excretion. Which disorder best explains this pattern?

- A.** Syndrome of inappropriate antidiuretic hormone secretion
- B.** Renal salt wasting with maximally dilute urine
- C.** Osmotic diuresis from severe hyperglycemia
- D.** Central diabetes insipidus

**139.** A newborn develops marked unconjugated hyperbilirubinemia after exposure to an oxidant drug. The blood smear shows bite cells, and direct antiglobulin testing is negative. Which diagnosis is most likely?

- A.** G6PD deficiency with oxidative hemolysis
- B.** Uncomplicated physiologic neonatal jaundice
- C.** Rh alloimmune hemolysis with antibody-coated cells
- D.** Biliary atresia with conjugated hyperbilirubinemia

**140.** A 24-week infant with a central venous catheter develops late-onset sepsis. Blood cultures repeatedly grow coagulase-negative staphylococci. What is the most likely source?

- A.** Food-borne maternal listeriosis at delivery
- B.** Catheter-associated infection involving skin flora
- C.** Transplacental infection acquired before rupture of membranes
- D.** Congenital viral infection causing bacterial culture positivity

**141.** A growth-restricted preterm infant receives rapidly advanced calories after a period of undernutrition and develops hypophosphatemia. Which process is most likely?

- A.** Refeeding-related intracellular phosphate shift during anabolic metabolism
- B.** Excess surfactant therapy causing renal phosphate wasting
- C.** Primary hyperparathyroidism causing isolated phosphate loss
- D.** Vitamin K deficiency causing phosphate sequestration

**142.** A term infant with sepsis has cool extremities, prolonged capillary refill, rising lactate, and low urine output. Which finding most directly indicates inadequate systemic oxygen delivery?

- A.** A single normal systolic blood pressure measurement
- B.** A soft systolic murmur without metabolic acidosis
- C.** Progressively rising serum lactate with worsening end-organ perfusion
- D.** A transient increase in heart rate during handling

**143.** The parents of an infant with a lethal condition and severe suffering ask whether burdensome intensive care can be redirected toward comfort. Which approach best reflects shared decision-making?

- A.** Make the decision solely on predicted hospital cost
- B.** Discuss prognosis, goals, values, and reasonable options with the family and interdisciplinary team
- C.** Require maximal life-prolonging treatment regardless of the infant's condition
- D.** Avoid discussing uncertainty because it may distress the parents

**144.** A 39-week infant is 3 hours old after placental abruption and prolonged resuscitation. The infant has lethargy, hypotonia, weak reflexes, and recurrent seizures consistent with moderate hypoxic-ischemic encephalopathy. Which intervention has the strongest evidence for reducing death or major neurodevelopmental disability?

- A.** Routine prophylactic phenobarbital in the absence of seizures
- B.** Whole-body therapeutic hypothermia started promptly
- C.** High-dose corticosteroids for cerebral edema
- D.** Induced hyperthermia to improve cerebral perfusion

**145.** A term newborn has severe hypoxemia from persistent pulmonary hypertension. Which physiologic change would be expected to worsen right-to-left extrapulmonary shunting?

- A.** A fall in pulmonary vascular resistance after lung recruitment
- B.** A rise in systemic vascular resistance with improved perfusion
- C.** Improved alveolar oxygenation with correction of acidosis
- D.** An increase in pulmonary vascular resistance relative to systemic resistance

**146.** A term infant with neonatal encephalopathy has an arterial carbon dioxide tension that is repeatedly very low because of excessive ventilation. Which neurologic concern makes marked hypocapnia undesirable?

- A.** Prevention of all seizure activity
- B.** Reduced cerebral blood flow from cerebral vasoconstriction
- C.** Increased cerebral blood flow from obligatory vasodilation
- D.** Direct stimulation of cerebrospinal-fluid production

**147.** Immediately after cord clamping, a macrosomic infant of a mother with poorly controlled diabetes develops symptomatic hypoglycemia. The infant's insulin and C-peptide levels remain inappropriately high. Which mechanism accounts for the glucose fall?

- A.** Persistent fetal hyperinsulinemia after interruption of maternal glucose delivery
- B.** Congenital absence of hepatic glycogen stores despite macrosomia
- C.** Failure of pancreatic beta cells to respond to maternal hyperglycemia
- D.** Excess neonatal glucagon secretion after placental separation

**148.** A very preterm infant begins parenteral nutrition on the first day of life. Why are amino acids provided early rather than waiting several days?

- A.** To limit negative nitrogen balance and support protein accretion
- B.** To eliminate the need for all future enteral protein
- C.** To close a patent ductus arteriosus through nitrogen loading
- D.** To prevent every episode of neonatal hypoglycemia

**149.** A newborn has a sharply demarcated area of absent skin on the scalp without surrounding inflammation. The infant is otherwise well. Which diagnosis is most likely?

- A.** Transient neonatal pustular melanosis
- B.** Aplasia cutis congenita
- C.** Seborrheic dermatitis
- D.** Erythema toxicum

**150.** A vigorous extremely preterm infant is breathing spontaneously immediately after birth and can remain close to the mother while thermal measures are provided. In the absence of a need for urgent resuscitation, which approach to the umbilical cord is generally favored?

- A.** Milk the intact cord repeatedly as the routine approach for all extremely preterm infants
- B.** Delay cord clamping briefly while providing routine thermal support
- C.** Clamp immediately solely because the infant is preterm
- D.** Keep the cord unclamped until the placenta is delivered regardless of clinical status

**151.** A spontaneously breathing 28-week infant has respiratory distress syndrome but is hemodynamically stable. Which initial respiratory strategy can help establish functional residual capacity while avoiding immediate intubation?

- A.** Routine chest compressions synchronized with spontaneous breaths
- B.** Negative-pressure abdominal binding
- C.** Immediate long-term invasive ventilation regardless of work of breathing
- D.** Nasal continuous positive airway pressure

**152.** A neonate with severe lung disease has persistent oxygen desaturation. The pulse-oximeter waveform is good, but arterial blood gas shows adequate PaO<sub>2</sub> while the pulse oximeter reads unexpectedly low. Which hemoglobin abnormality can produce this discrepancy?

- A.** Fetal hemoglobin predominance alone
- B.** Clinically significant methemoglobinemia
- C.** Physiologic anemia of infancy
- D.** Mild iron deficiency without dyshemoglobinemia

**153.** A fetus has a large left congenital diaphragmatic hernia with liver herniation into the chest. Which prenatal consequence most strongly predicts severe postnatal respiratory disease?

- A.** Primary tracheal stenosis without pulmonary vascular involvement
- B.** Excessive alveolar number causing immediate hyperinflation
- C.** Pulmonary hypoplasia with abnormal pulmonary vascular development
- D.** Isolated surfactant overproduction with normal lung size

**154.** A newborn has superficial pustules present at birth that rupture easily and leave hyperpigmented macules with a collarette of scale. The infant is otherwise healthy. Which diagnosis is most likely?

- A.** Transient neonatal pustular melanosis
- B.** Epidermolysis bullosa
- C.** Neonatal herpes simplex virus infection
- D.** Bullous impetigo

**155.** A monochorionic diamniotic twin pregnancy develops oligohydramnios and growth restriction in one twin and polyhydramnios with cardiomegaly in the other. What is the underlying mechanism?

- A.** Two separate placentas with independent maternal vascular malperfusion
- B.** Discordant fetal insulin secretion caused by maternal diabetes
- C.** Maternal alloimmunization affecting only the larger twin
- D.** Unbalanced placental vascular anastomoses causing net transfusion between twins

**156.** A ventilated neonate suddenly has pink frothy secretions from the endotracheal tube, worsening oxygenation, and diffuse new alveolar opacities. Which diagnosis is most likely?

- A.** Pulmonary hemorrhage
- B.** Isolated upper-airway laryngomalacia
- C.** Transient tachypnea of the newborn
- D.** Simple apnea of prematurity

**157.** A very preterm infant with a prior large intraventricular hemorrhage has progressively increasing ventricular size on serial cranial ultrasound. Head growth accelerates over several days. Which complication is most likely?

- A.** Isolated craniosynostosis
- B.** Posthemorrhagic ventricular dilatation
- C.** Benign enlargement of the subarachnoid spaces
- D.** Congenital microcephaly

**158.** A 27-week infant with low-volume reticulogranular lungs has escalating oxygen needs and is intubated after CPAP fails. Which treatment acts directly at the alveolar air-liquid interface to reduce surface tension?

- A.** Inhaled nitric oxide as routine treatment for surfactant deficiency
- B.** Prostaglandin E1 to maintain pulmonary blood flow
- C.** Systemic bicarbonate to increase alveolar surface tension
- D.** Intratracheal exogenous surfactant replacement

**159.** A ventilated extremely preterm infant has repeated episodes of hypocarbia with PaCO<sub>2</sub> values in the low 20s mm Hg. Why is this undesirable?

- A.** Marked hypocarbia can reduce cerebral blood flow and contribute to brain injury
- B.** Hypocarbia reliably increases cerebral blood flow and prevents ischemia
- C.** Low PaCO<sub>2</sub> has no important physiologic effect in preterm infants
- D.** Hypocarbia prevents all forms of ventilator-induced lung injury

**160.** A term infant has early unconjugated hyperbilirubinemia and anemia. The direct antiglobulin test is negative, and the blood smear shows numerous spherocytes. The father underwent splenectomy for chronic hemolytic anemia. Which diagnosis is most likely?

- A.** Vitamin K deficiency bleeding
- B.** Anemia of prematurity
- C.** ABO immune hemolysis
- D.** Hereditary spherocytosis

**161.** A 31-week infant is delivered after maternal fever, uterine tenderness, fetal tachycardia, and prolonged rupture of membranes. Which fetal exposure best links this history to early neonatal illness?

- A.** Isolated maternal hyperthyroidism without placental inflammation
- B.** An intra-amniotic inflammatory and infectious process
- C.** Maternal anti-Ro antibody transfer causing isolated heart block
- D.** A chronic fetomaternal hemorrhage without membrane rupture

**162.** A newborn with prenatally diagnosed congenital diaphragmatic hernia has severe respiratory distress immediately after birth. Which initial respiratory strategy is most appropriate?

- A.** Endotracheal intubation and gastric decompression while avoiding routine bag-mask ventilation
- B.** Immediate oral feeding to decompress the stomach
- C.** Prolonged vigorous bag-mask ventilation to inflate both lungs and the stomach
- D.** Routine chest compressions before assessing heart rate

**163.** A newborn has a lumbosacral myelomeningocele. Before definitive neurosurgical closure, which immediate bedside measure is most appropriate?

- A.** Begin vigorous passive range-of-motion exercises across the lesion
- B.** Apply povidone-iodine repeatedly to desiccate exposed neural tissue
- C.** Place the infant supine directly on the lesion
- D.** Cover the lesion with a sterile moist nonadherent dressing and avoid pressure on it

**164.** A premature infant undergoing retinopathy screening is found to have plus disease with retinal changes meeting treatment criteria. Which general management principle is most appropriate?

- A.** Treat only if the red reflex becomes absent
- B.** Delay all intervention until the infant reaches term-corrected age regardless of progression
- C.** Arrange prompt treatment by an ophthalmologist experienced in retinopathy of prematurity
- D.** Stop ophthalmologic examinations because spontaneous regression is guaranteed

**165.** A term infant with severe birth asphyxia develops oliguria, rising creatinine, hyperkalemia, and fluid overload. Which diagnosis is most likely?

- A.** Acute kidney injury from hypoxic-ischemic renal injury
- B.** Physiologic postnatal diuresis
- C.** Central diabetes insipidus
- D.** Isolated nephrogenic salt wasting without renal injury

**166.** A fetus develops hydrops and severe anemia in a pregnancy complicated by maternal red-cell alloimmunization. Which fetal test is commonly used noninvasively to screen for significant anemia?

- A.** Maternal serum alpha-fetoprotein concentration
- B.** Umbilical artery resistance index alone
- C.** Middle cerebral artery peak systolic velocity Doppler
- D.** Fetal ductus venosus A-wave direction alone

**167.** Prenatal imaging shows alobar holoprosencephaly and a congenital heart defect. At birth, the infant has a midline cleft lip, postaxial polydactyly, and severe growth restriction. Which chromosomal abnormality is most likely?

- A.** Trisomy 13
- B.** 47,XXY
- C.** Trisomy 18
- D.** Monosomy X

**168.** A term newborn has diffuse petechiae and a platelet count of 12,000/ $\mu$ L but is otherwise well. The mother's platelet count is normal. Which diagnosis should be strongly considered?

- A.** Fetal and neonatal alloimmune thrombocytopenia
- B.** Maternal immune thrombocytopenia with severe maternal thrombocytopenia
- C.** Anemia of prematurity
- D.** Physiologic neonatal thrombocytopenia

**169.** A preterm infant has repeated episodes of eye deviation and subtle bicycling movements. The bedside examination cannot determine whether these events are epileptic. Which test is most useful for confirming neonatal seizures?

- A.** Routine skull radiography
- B.** A single serum prolactin level
- C.** Continuous video electroencephalographic monitoring
- D.** Amplitude-integrated EEG alone as a definitive test for all events

**170.** A 9-day-old infant develops clustered vesicles on the scalp and around the eye. The mother had no recognized genital lesions at delivery. Which infection must be treated urgently while testing is performed?

- A.** Transient neonatal pustular melanosis
- B.** Neonatal herpes simplex virus infection
- C.** Erythema toxicum neonatorum
- D.** Miliaria from heat exposure

**171.** A newborn has multiple fractures, blue sclerae, and severe bone demineralization without a history of birth trauma sufficient to explain the injuries. Which diagnosis is most likely?

- A.** Congenital hypothyroidism
- B.** Physiologic osteopenia of the term newborn
- C.** Osteogenesis imperfecta
- D.** Achondroplasia

**172.** A newborn has recurrent inspiratory stridor that worsens when supine and improves when prone. Oxygenation and feeding are otherwise adequate. What is the most likely diagnosis?

- A.** Tension pneumothorax causing unilateral absent breath sounds
- B.** Bilateral choanal atresia causing expiratory obstruction
- C.** Laryngomalacia with dynamic supraglottic collapse
- D.** Respiratory distress syndrome from surfactant deficiency

**173.** A post-term infant is born through thick meconium-stained fluid and later develops coarse breath sounds, patchy atelectasis, hyperinflation, and hypoxemia. Which mechanism contributes to this lung disease?

- A.** Pulmonary hypoplasia from prolonged oligohydramnios
- B.** Airway obstruction with ball-valve air trapping plus chemical pneumonitis
- C.** Pure cardiogenic edema from isolated left ventricular failure
- D.** Uniform alveolar collapse caused only by absent surfactant synthesis

**174.** At follow-up, a former extremely preterm infant has normal head growth but poor social communication, delayed language, and repetitive behaviors. Which principle is most appropriate for developmental surveillance?

- A.** A normal neonatal cranial ultrasound eliminates the need for developmental follow-up
- B.** Motor assessment alone is sufficient through school age
- C.** Developmental testing should be deferred until adolescence
- D.** Former very preterm infants require longitudinal screening across multiple developmental domains

**175.** A 26-week infant develops grunting and retractions within minutes of birth. Chest radiography shows diffuse reticulogranular opacities and low lung volumes. What is the central pathophysiologic defect?

- A.** Insufficient pulmonary surfactant causing alveolar instability and low compliance
- B.** Delayed absorption of fetal lung fluid with preserved surfactant function
- C.** Fixed pulmonary venous obstruction producing hydrostatic edema
- D.** Primary upper-airway collapse from bilateral vocal cord paralysis

**176.** A newborn with persistent abdominal distention undergoes contrast enema showing a narrow distal rectosigmoid segment with proximal dilatation. Rectal suction biopsy demonstrates absence of ganglion cells. Which diagnosis is established?

- A.** Gastroesophageal reflux
- B.** Hirschsprung disease
- C.** Duodenal atresia with no distal bowel gas
- D.** Hypertrophic pyloric stenosis

**177.** Following severe sepsis and shock, a preterm infant begins oozing from an umbilical catheter site and heel punctures. Platelets and fibrinogen are low, PT and aPTT are prolonged, and fibrin-degradation products are elevated. Which hemostatic disorder is present?

- A.** Disseminated intravascular coagulation
- B.** Isolated hemophilia A
- C.** Fetal and neonatal alloimmune thrombocytopenia
- D.** Vitamin K deficiency without consumption

**178.** A term newborn has rhythmic limb movements that continue despite gentle restraint and are accompanied by eye deviation. Which feature most strongly favors an epileptic seizure over neonatal jitteriness?

- A.** The movements are stimulus sensitive
- B.** The movements stop with flexion of the extremity
- C.** The movements are not suppressed by holding the limb
- D.** The infant has a normal gaze during the event

**179.** A former 25-week infant has poor linear growth, low serum phosphorus, and elevated alkaline phosphatase while receiving prolonged nutritional support. Which diagnosis is most likely?

- A.** Congenital hyperparathyroidism as the routine cause
- B.** Metabolic bone disease of prematurity
- C.** Osteopetrosis from excessive mineral intake
- D.** Vitamin K deficiency without bone involvement

**180.** A newborn with severe hypoxic brain injury develops marked hyponatremia, high serum osmolality, very dilute urine, and polyuria. Which disorder should be suspected?

- A.** Syndrome of inappropriate antidiuretic hormone secretion
- B.** Acute glomerulonephritis
- C.** Congenital nephrotic syndrome with water retention
- D.** Central diabetes insipidus

**181.** A term infant has severe hypoxemia with a preductal oxygen saturation of 94% in the right hand and a postductal saturation of 82% in a foot. Echocardiography shows no structural heart defect. What process best explains the differential saturation?

- A.** A small isolated ventricular septal defect without pulmonary hypertension
- B.** Premature closure of the foramen ovale with normal pulmonary pressure
- C.** Right-to-left ductal shunting from persistent pulmonary hypertension
- D.** Left-to-right ductal shunting caused by low pulmonary vascular resistance

**182.** A very preterm infant who received prolonged loop-diuretic therapy develops echogenic foci in the renal pyramids on ultrasonography. Which complication is most likely?

- A.** Renal agenesis
- B.** Renal vein thrombosis
- C.** Nephrocalcinosis
- D.** Posterior urethral valves

**183.** A large-for-gestational-age newborn has recurrent hypoglycemia that requires a progressively high glucose infusion rate. Insulin remains detectable during a low plasma glucose concentration. Which mechanism is most likely?

- A.** Complete absence of pancreatic beta-cell function
- B.** Inappropriate insulin secretion suppressing ketone and glucose production
- C.** Isolated cortisol excess increasing hepatic glucose output
- D.** Appropriate fasting adaptation with high ketone production

**184.** A newborn with pulmonary atresia has rapidly worsening cyanosis as the ductus constricts. Which physiologic goal is most urgent?

- A.** Reduce atrial mixing by closing the foramen ovale
- B.** Preserve ductal blood flow to the pulmonary arteries
- C.** Lower systemic venous return with aggressive diuresis
- D.** Accelerate ductal closure to reduce pulmonary overcirculation

**185.** An extremely preterm infant receiving prolonged broad-spectrum antibiotics and parenteral nutrition develops persistent thrombocytopenia and sepsis. Blood cultures grow yeast. Which infection is most likely?

- A.** Invasive candidiasis
- B.** Early-onset group B streptococcal disease
- C.** Respiratory syncytial virus bronchiolitis
- D.** Congenital rubella

**186.** A preterm infant receiving gentamicin for suspected sepsis has worsening renal function. Which pharmacologic approach is most appropriate?

- A.** Increase dosing frequency because renal failure accelerates gentamicin elimination
- B.** Use therapeutic drug monitoring and adjust the dosing interval for reduced clearance
- C.** Ignore serum concentrations because aminoglycoside toxicity is unrelated to exposure
- D.** Switch automatically to an oral aminoglycoside formulation

**187.** A stable newborn has a regular narrow-complex tachycardia at 260/min. Vagal maneuvers fail, blood pressure is preserved, and there is no evidence of pre-excitation on the monitor. What is the next acute therapy?

- A.** Oral digoxin as the immediate first intervention during the episode
- B.** Intravenous epinephrine to increase atrioventricular conduction
- C.** Indomethacin to eliminate a ductal trigger for the tachycardia
- D.** Rapid intravenous adenosine for supraventricular tachycardia

**188.** A mechanically ventilated 24-week infant suddenly requires higher pressures. Chest radiography shows branching radiolucent streaks tracking along bronchovascular bundles and small cyst-like air collections within the lung. Which air-leak disorder is most likely?

- A.** Transient tachypnea from retained fetal lung fluid
- B.** Diffuse pulmonary hemorrhage without an air-leak pattern
- C.** Pulmonary interstitial emphysema from air dissecting into the interstitium
- D.** Pulmonary edema caused by nephrotic syndrome

**189.** A newborn is delivered to a mother who developed varicella from 5 days before to 2 days after delivery. Why is this timing particularly concerning?

- A.** Varicella cannot be transmitted around the time of birth
- B.** Only maternal disease in the first trimester can affect the newborn
- C.** Maternal varicella in this interval guarantees lifelong infant immunity
- D.** High viral exposure may occur before adequate protective maternal antibody is transferred

**190.** A ventilated preterm infant suddenly becomes bradycardic and profoundly hypoxemic. Breath sounds are absent on the right and the right chest is hyperexpanded. What is the immediate concern?

- A.** Apnea of prematurity with symmetric aeration
- B.** Left-to-right patent ductus shunting without lung asymmetry
- C.** Tension pneumothorax causing acute cardiorespiratory compromise
- D.** Gradual bronchopulmonary dysplasia without an acute air leak

**191.** A preterm infant has frequent apnea and bradycardia. During one event there are no abnormal movements, and continuous EEG shows no ictal activity. Which interpretation is most appropriate?

- A.** Normal EEG during the event proves the infant has no neurologic disease of any kind
- B.** Every apnea episode in a preterm infant is an electrographic seizure
- C.** Apnea alone requires long-term antiseizure medication
- D.** Most apnea of prematurity is not caused by epileptic seizures

**192.** A dysmorphic newborn has multiple congenital anomalies, but a standard karyotype is normal and there is no obvious single-gene syndrome. Which test is commonly the next genomic study for detecting submicroscopic copy-number changes?

- A.** Repeat newborn metabolic screening alone
- B.** Direct Coombs testing
- C.** Chromosomal microarray analysis
- D.** Serum alpha-fetoprotein measurement

**193.** A term infant develops severe cyanosis, pulmonary edema, and respiratory distress soon after birth. Echocardiography shows all pulmonary veins draining to a vertical vein with obstruction before reaching the systemic venous circulation. What is the diagnosis?

- A.** Isolated pulmonary valve stenosis
- B.** Obstructed total anomalous pulmonary venous return
- C.** Unobstructed atrial septal defect
- D.** Coarctation with normal pulmonary venous drainage

**194.** A screening test is negative in 950 of 1,000 infants who do not have the disease. Which test characteristic is 95%?

- A.** Prevalence
- B.** Specificity
- C.** Sensitivity
- D.** Relative risk

**195.** A newborn screen later shows elevated thyroid-stimulating hormone, and confirmatory testing reveals low free thyroxine with high TSH. What is the most likely diagnosis?

- A.** Central hypothyroidism from pituitary failure
- B.** Physiologic euthyroid transition
- C.** Primary congenital hypothyroidism
- D.** Neonatal Graves disease

**196.** A newborn has micrognathia, glossoptosis, and a U-shaped cleft palate with intermittent upper-airway obstruction. Which sequence is most likely?

- A.** Pierre Robin sequence
- B.** VACTERL association
- C.** Beckwith-Wiedemann syndrome
- D.** Turner syndrome

**197.** A newborn with persistent hyperinsulinemic hypoglycemia has macroglossia, hemihyperplasia, and enlarged abdominal organs. The care team discusses later embryonal-tumor surveillance. Which overgrowth syndrome is most likely?

- A.** Smith-Lemli-Opitz syndrome
- B.** Trisomy 18
- C.** Beckwith-Wiedemann syndrome
- D.** Cri-du-chat syndrome

**198.** A ventilated newborn suddenly becomes hypotensive and severely hypoxemic. Breath sounds are absent on the right, and transillumination is strongly positive. There is no time to obtain radiography. What should be done first?

- A.** Begin enteral feeding to reduce agitation
- B.** Immediate decompression of the suspected tension pneumothorax
- C.** Wait for a chest radiograph before intervening
- D.** Increase positive end-expiratory pressure substantially

**199.** An unvaccinated mother had a febrile maculopapular illness during the first trimester. Her infant is born with a congenital cataract, patent ductus arteriosus, and sensorineural deafness. Which congenital infection best explains the pattern?

- A.** Parvovirus B19
- B.** Cytomegalovirus
- C.** Congenital rubella
- D.** Herpes simplex virus

**200.** Prenatal growth restriction and choroid plexus cysts are followed by birth findings of a prominent occiput, micrognathia, clenched fists with overlapping digits, and congenital heart disease. Which chromosomal disorder is most likely?

- A.** Trisomy 18 syndrome
- B.** Beckwith-Wiedemann syndrome
- C.** Turner syndrome
- D.** Trisomy 21

### Answers

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**1. C.** Thyroid-stimulating receptor antibodies can persist after definitive maternal treatment and cross the placenta. A mother can therefore be clinically hypothyroid after ablation while her infant develops transient antibody-mediated hyperthyroidism.

**2. B.** Maternal hypertensive disease and placental insufficiency are associated with fetal growth restriction and transient neonatal cytopenias, including thrombocytopenia. The mechanism reflects chronic placental stress and altered fetal hematopoiesis rather than acute neonatal infection in an otherwise well infant.

**3. A.** Newborns are preferential nasal breathers. Bilateral choanal atresia can cause cyclic cyanosis that improves with crying, and inability to pass a catheter through the nares supports the diagnosis.

**4. A.** Extremely preterm infants lose heat rapidly because of thin skin, high surface-area-to-mass ratio, and large evaporative losses. A polyethylene wrap or bag used under radiant heat limits evaporation while resuscitative care proceeds.

**5. C.** During resuscitation, a low umbilical venous catheter can provide rapid emergency venous access when inserted only far enough for free blood return. If continued central use is needed, definitive tip position must be confirmed.

**6. C.** Emergency treatment needed to prevent death or serious harm may proceed when a legal surrogate is unavailable and delay would endanger the child. Documentation and continued efforts to contact the parents remain important.

**7. A.** Bilious vomiting in a newborn is concerning for intestinal obstruction. Malrotation with midgut volvulus can compromise mesenteric blood flow rapidly and requires urgent surgical evaluation and prompt diagnostic imaging without delaying definitive care.

- 8. B.** Automated auditory brainstem response assesses neural transmission through the auditory pathway and is preferred for NICU populations at increased risk for auditory neuropathy. Failed screening requires timely diagnostic follow-up.
- 9. B.** High-frequency oscillatory ventilation uses very rapid small-volume oscillations around a controlled mean airway pressure. It can be used as a rescue strategy when conventional ventilation cannot provide adequate gas exchange without potentially injurious settings.
- 10. A.** The classic triad of renal vein thrombosis is hematuria, thrombocytopenia, and an enlarged kidney, although all features are not always present. Risk factors include dehydration, sepsis, polycythemia, and maternal diabetes.
- 11. C.** Preterm kidneys have limited tubular sodium reabsorptive capacity and a reduced ability to conserve sodium. Renal sodium losses can therefore contribute to hyponatremia and poor growth after the early diuretic phase.
- 12. C.** The combination of characteristic facial features, hypotonia, and an atrioventricular septal defect is classic for trisomy 21. Confirmation is by cytogenetic testing, which also distinguishes free trisomy from translocation or mosaicism.
- 13. D.** Reversed end-diastolic flow in a growth-restricted fetus is a marker of high placental vascular resistance. A small infarcted placenta is characteristic of maternal vascular malperfusion, often associated with hypertensive disease or other causes of uteroplacental insufficiency.
- 14. D.** CHARGE is an acronym reflecting coloboma, heart disease, choanal atresia, growth or developmental abnormalities, genital anomalies, and ear abnormalities. Airway and feeding difficulties are common neonatal management issues.
- 15. A.** A high umbilical arterial catheter position is typically in the descending thoracic aorta above major abdominal branches and below the aortic arch, approximately T6-T9 on radiography.
- 16. B.** When mother's own milk is unavailable, pasteurized donor human milk is commonly preferred for very preterm infants because human-milk feeding is associated with a lower risk of necrotizing enterocolitis than formula feeding. Fortification may still be needed for growth.
- 17. C.** Benign neonatal sleep myoclonus consists of myoclonic jerks during sleep that cease with arousal and are not associated with epileptiform EEG activity. Recognition prevents unnecessary antiseizure treatment.
- 18. D.** Perinatal arterial ischemic stroke commonly presents with focal neonatal seizures. MRI with diffusion-weighted imaging is the preferred study for defining acute ischemic brain injury and vascular territory.

- 19. A.** Serum creatinine in the first day or two reflects both maternal creatinine and the newborn's evolving renal function. Trends over time, urine output, gestational age, and clinical context are more informative than a single early value.
- 20. D.** Chyle contains triglyceride-rich lymph and becomes characteristically milky after enteral fat intake begins. Thoracic surgery can disrupt lymphatic channels and produce a neonatal chylothorax.
- 21. A.** Gastroesophageal reflux is common in infants because of liquid feeds, supine positioning, and immature gastrointestinal function. Treatment decisions should be driven by clinically important complications rather than visible regurgitation alone.
- 22. C.** Trophic or minimal enteral feeds provide small amounts of milk to stimulate gastrointestinal maturation while nutrition is advanced cautiously. They are not intended to supply the infant's complete caloric needs immediately.
- 23. D.** When very preterm delivery is anticipated, antenatal magnesium sulfate is used for fetal neuroprotection and reduces the risk of cerebral palsy. It is not an antimicrobial treatment, a glucose-stabilizing therapy, or a method of closing the ductus.
- 24. A.** The four defining features are a ventricular septal defect, overriding aorta, right ventricular outflow obstruction, and right ventricular hypertrophy. The degree of neonatal cyanosis depends mainly on the severity of right ventricular outflow obstruction.
- 25. D.** Anemia of prematurity reflects low endogenous erythropoietin response, shortened red-cell lifespan, rapid growth, and phlebotomy losses. The gradual decline with a relatively low reticulocyte response differs from acute hemorrhage or brisk hemolysis.
- 26. B.** Infants of diabetic mothers are at increased risk for early neonatal hypocalcemia, in part because of altered parathyroid adaptation. Jitteriness, tremors, or seizures can be presenting manifestations when ionized calcium is low.
- 27. A.** Hyperammonemia can stimulate central hyperventilation and produce respiratory alkalosis, a useful clue to a urea-cycle defect. In contrast, many organic acidemias more often present with a high-anion-gap metabolic acidosis.
- 28. C.** Early-onset GBS disease typically presents in the first day with sepsis, pneumonia, or meningitis after vertical exposure. Late line-associated and fungal pathogens are less consistent with illness beginning only hours after birth.
- 29. C.** Current neonatal resuscitation practice does not recommend routine tracheal suction solely because a nonvigorous infant was born through meconium. If the infant is apneic or bradycardic, establishing ventilation promptly is the priority, with airway suction reserved for suspected obstruction.

- 30. D.** Carbon dioxide elimination depends mainly on effective alveolar minute ventilation. Increasing effective tidal volume or respiratory rate raises minute ventilation and generally lowers PaCO<sub>2</sub>, whereas changing inspired oxygen alone does not directly correct hypercapnia.
- 31. C.** Maternal IgG antibodies against A or B antigens can cross the placenta and cause neonatal hemolysis, most often when the mother is group O. A positive direct antiglobulin test supports antibody-coated infant red cells.
- 32. C.** Chest compressions are indicated when the heart rate remains below 60/min despite effective ventilation. Because ventilation failure is the commonest cause of neonatal bradycardia, compressions should not precede confirmation that ventilation is effective.
- 33. A.** Ten percent dextrose contains 100 mg/mL. At 4 mL/kg/h, the infant receives 400 mg/kg/h; dividing by 60 minutes gives a glucose infusion rate of about 6.7 mg/kg/min.
- 34. D.** Parenteral-nutrition-associated liver disease is multifactorial. Prematurity, sepsis, prolonged parenteral nutrition, and absence of enteral feeding all contribute by altering bile flow, metabolism, and intestinal-liver signaling.
- 35. C.** Fluid management in extremely preterm infants is dynamic. Serial weight, electrolytes, urine output, humidity, phototherapy, respiratory support, and clinical status should guide adjustments rather than a fixed formula alone.
- 36. A.** Prader-Willi syndrome often presents in the neonatal period with marked hypotonia and feeding difficulty, despite the later development of hyperphagia. The disorder reflects absent expression of paternally derived genes in the 15q11-q13 region.
- 37. B.** A poor heart-rate response with absent chest movement indicates ineffective ventilation. The priority is to correct mask seal, airway position, obstruction, pressure, or airway access until ventilation moves the chest before escalating to compressions or medications.
- 38. B.** Inability to pass a tube into the stomach with excessive secretions and aspiration symptoms strongly suggests esophageal atresia. Many affected infants also have a distal tracheoesophageal fistula that permits gas to enter the stomach.
- 39. D.** The lung point is the transition between sliding lung and a region without sliding and is highly specific for pneumothorax when present. Absence of lung sliding alone is less specific because other conditions can also abolish sliding.
- 40. D.** Meconium ileus results from inspissated intestinal contents and is strongly associated with cystic fibrosis. The distal small bowel is obstructed by thick meconium, and contrast enema may show a microcolon.

- 41. C.** Newborn cardiac arrest is most often asphyxial, so ventilation is emphasized during cardiopulmonary resuscitation. Coordinated neonatal CPR therefore uses a 3:1 compression-to-ventilation ratio, producing 90 compressions and 30 breaths per minute.
- 42. B.** In d-transposition, pulmonary and systemic circulations run largely in parallel, so survival depends on mixing. A restrictive atrial septum can cause severe hypoxemia; balloon atrial septostomy enlarges the interatrial communication and improves mixing.
- 43. D.** A 22q11.2 deletion can disrupt pharyngeal pouch development, causing conotruncal cardiac disease, thymic hypoplasia, and hypoparathyroidism with hypocalcemia. The severity of immune dysfunction varies among affected infants.
- 44. A.** Current risk-stratified management gives antiretroviral prophylaxis to all infants with in utero or intrapartum HIV exposure. With sustained maternal suppression defining low transmission risk, zidovudine alone for a short course is appropriate, whereas high-risk exposure calls for multidrug presumptive therapy.
- 45. A.** Physiologic jaundice reflects increased bilirubin production, immature hepatic conjugation, and enhanced enterohepatic circulation. It typically appears after the first day and resolves as feeding and hepatic handling mature.
- 46. D.** Critical coarctation may be masked while the ductus remains open and then present with shock as ductal flow declines. Upper-extremity hypertension relative to the legs and diminished femoral pulses are key bedside clues.
- 47. D.** Neutrophilic CSF pleocytosis with low glucose and high protein in a symptomatic neonate is strongly concerning for bacterial meningitis. Neonatal meningitis requires prompt systemic antimicrobial therapy because neurologic injury can progress rapidly.
- 48. C.** Turner syndrome can present in the newborn period with peripheral lymphedema, webbed neck, shield chest, and left-sided cardiac lesions such as coarctation. Karyotyping typically demonstrates monosomy X or a related X-chromosome abnormality.
- 49. C.** Parvovirus B19 has tropism for erythroid progenitor cells and can suppress fetal erythropoiesis. Severe anemia may lead to high-output cardiac failure and nonimmune hydrops fetalis.
- 50. A.** Apnea of prematurity reflects immature respiratory control after secondary causes have been excluded. Caffeine is a methylxanthine that stimulates respiratory drive and improves diaphragmatic function, reducing clinically important apnea in preterm infants.
- 51. D.** Classic galactosemia can present after lactose exposure with liver dysfunction, hypoglycemia, vomiting, and a characteristic susceptibility to *E. coli* sepsis. Galactose and lactose should be removed while confirmatory testing is pursued.

- 52. B.** The classic triad of congenital toxoplasmosis includes chorioretinitis, hydrocephalus, and intracranial calcifications. CMV more characteristically produces periventricular calcifications and microcephaly rather than hydrocephalus.
- 53. D.** Both hypoxemia and hyperoxemia can be harmful in very preterm infants. Continuous pulse oximetry with a prescribed saturation target and appropriate alarm limits allows oxygen to be adjusted rather than delivered at an unnecessarily high fixed concentration.
- 54. A.** Classic 21-hydroxylase deficiency impairs cortisol and aldosterone synthesis and increases androgen production. Salt-wasting crisis produces hyponatremia, hyperkalemia, dehydration, and shock, while affected 46,XX infants may have virilized external genitalia.
- 55. C.** Lung aeration raises oxygen tension and placental separation lowers circulating prostaglandin levels. These changes favor ductal smooth-muscle constriction and functional closure in the normal newborn.
- 56. B.** Preterm gastric emptying is variable, and isolated residual volume is a nonspecific finding. Feeding decisions should integrate the abdominal examination, emesis, stool pattern, systemic signs, and imaging rather than treating residual volume alone as proof of disease.
- 57. D.** Ductal-dependent systemic blood flow can deteriorate rapidly as the ductus arteriosus constricts. Prostaglandin E1 maintains or reopens the ductus and stabilizes systemic perfusion while definitive cardiac anatomy and treatment are arranged.
- 58. A.** Maple syrup urine disease results from deficient branched-chain alpha-ketoacid dehydrogenase activity. Accumulation of leucine and related metabolites produces encephalopathy and the characteristic odor in the neonatal period.
- 59. B.** Newborns rely heavily on nonshivering thermogenesis in brown adipose tissue. Cold stress increases oxygen and glucose consumption and can worsen hypoglycemia, metabolic acidosis, and respiratory instability.
- 60. C.** Posterior urethral valves are a major cause of lower urinary tract obstruction in male newborns. Severe obstruction can produce bladder distention, bilateral hydroureteronephrosis, renal dysplasia, and oligohydramnios before birth.
- 61. B.** Preterm infants have lower iron stores because much fetal iron accretion occurs late in gestation, and rapid growth increases requirements. Supplemental iron is commonly needed once enteral feeding is established, with timing individualized to transfusion history and clinical status.
- 62. D.** Qualified medical interpreters improve accuracy, confidentiality, and informed decision-making. Family members, especially children, should not routinely be used to interpret complex medical information when professional interpretation is available.

- 63. D.** *E. coli* is a major cause of early-onset neonatal sepsis, particularly among preterm and very-low-birth-weight infants. The timing and maternal membrane rupture support vertical acquisition rather than a later catheter-associated infection.
- 64. A.** Positive predictive value depends on disease prevalence as well as test performance. When prevalence rises, a positive result is more likely to represent true disease, so positive predictive value generally increases.
- 65. D.** Pulmonary vascular resistance is relatively high at birth, limiting left-to-right flow across a ventricular septal defect. As resistance falls over the first weeks, pulmonary overcirculation increases and heart-failure symptoms may become apparent.
- 66. B.** Increasing systemic vascular resistance reduces right-to-left shunting across the ventricular septal defect in tetralogy physiology. Knee-chest positioning is a temporizing maneuver while oxygenation, volume status, and definitive management are addressed.
- 67. D.** The combination of persistent hypoglycemia, micropenis, and cholestasis can signal congenital hypopituitarism. Deficiencies of cortisol and growth hormone impair glucose homeostasis, while genital findings suggest gonadotropin deficiency.
- 68. B.** A significant patent ductus arteriosus produces left-to-right shunting after pulmonary vascular resistance falls. Bounding pulses, widened pulse pressure, a characteristic murmur, and worsening pulmonary overcirculation are classic findings.
- 69. D.** Hypertrophic pyloric stenosis produces gastric outlet obstruction, causing progressive nonbilious projectile vomiting and a characteristic hypochloremic metabolic alkalosis. Bilious emesis would instead suggest obstruction distal to the ampulla and requires urgent evaluation.
- 70. C.** Volutrauma refers to injury from excessive tidal stretch and overdistension. Preterm lungs are especially vulnerable because disease is heterogeneous, so apparently modest global tidal volumes can still overdistend the most compliant regions.
- 71. D.** VACTERL describes the nonrandom co-occurrence of vertebral, anorectal, cardiac, tracheoesophageal, renal, and limb anomalies. It is a clinical association rather than a single uniform genetic diagnosis, so evaluation should search systematically for each component.
- 72. C.** *Listeria* can cause maternal febrile illness, placental infection, preterm birth, fetal loss, and neonatal sepsis or meningitis. Transplacental or peripartum acquisition makes the maternal gastrointestinal-like illness an important clue.
- 73. A.** Leukocoria is an abnormal white pupillary reflex. Retinoblastoma is an important vision- and life-threatening cause and warrants urgent ophthalmologic evaluation, although congenital cataract and other disorders are also possible.

- 74. B.** A venous hematocrit above about 65% defines neonatal polycythemia and can increase blood viscosity. Symptomatic infants may have neurologic, metabolic, gastrointestinal, or cardiorespiratory manifestations.
- 75. C.** Preterm infants may have immature suck-swallow-breathe coordination and oropharyngeal dysphagia. Coughing, desaturation, and feeding-associated events raise concern for aspiration rather than a new diffuse pulmonary disorder.
- 76. C.** For term and late-preterm newborns requiring respiratory support, resuscitation can generally start with room air and oxygen is then titrated using preductal saturation targets. Routine exposure to 100% oxygen is avoided because hyperoxia can be harmful.
- 77. B.** Persistent severe bradycardia after effective ventilation and compressions is an indication for epinephrine. The intravenous route is preferred once reliable umbilical venous access is available because drug delivery is more predictable than endotracheal administration.
- 78. D.** Bronchopulmonary dysplasia is a chronic lung disorder of prematurity associated with disrupted alveolar and vascular development plus injury from oxygen, inflammation, and respiratory support. Persistent oxygen or respiratory-support need near 36 weeks postmenstrual age is central to contemporary severity assessment.
- 79. B.** The classic Erb pattern after shoulder dystocia reflects upper brachial plexus injury, usually C5-C6. The arm is adducted and internally rotated with an extended elbow, while hand grasp may remain intact.
- 80. B.** Plan-Do-Study-Act cycles use iterative small tests of change: plan an intervention, do it, study the results, and act on what was learned. This approach is widely used in quality improvement.
- 81. D.** Grade I intraventricular hemorrhage is confined to the germinal matrix. The highly vascular germinal matrix of very preterm infants is particularly vulnerable to fluctuations in cerebral blood flow.
- 82. D.** Antenatal corticosteroids accelerate structural and biochemical maturation of the fetal lung and increase surfactant production. Their neonatal benefits also extend beyond respiratory distress syndrome, but lung maturation is the mechanism most directly relevant here.
- 83. C.** Spontaneous intestinal perforation usually affects extremely preterm infants and often presents as an isolated focal perforation with pneumoperitoneum. It differs from necrotizing enterocolitis, which more often features diffuse intestinal inflammation and pneumatosis.
- 84. C.** Persistent truncus arteriosus results from failure of conotruncal septation, leaving a single arterial trunk that supplies the pulmonary, systemic, and coronary circulations. A ventricular septal defect is usually present beneath the common trunk.

- 85. B.** A periventricular hemorrhagic infarction associated with severe IVH is thought to result from obstruction of terminal venous drainage and venous infarction, rather than simple extension of blood into previously normal tissue.
- 86. B.** Early congenital syphilis can cause rhinitis, hepatosplenomegaly, rash including palms and soles, anemia or thrombocytopenia, and characteristic bone lesions. Maternal serology and infant evaluation guide treatment.
- 87. C.** Congenital lobar overinflation produces hyperexpansion of an affected lobe and compresses adjacent normal lung. The presence of intrapulmonary markings within the hyperlucent lobe helps distinguish it from a pneumothorax.
- 88. D.** A major placental abruption can cause acute fetal blood loss as well as hypoxic-ischemic stress. Profound pallor, hypotension, and a low hemoglobin at birth support hemorrhagic shock rather than a normal postnatal change in hemoglobin.
- 89. A.** For a relative measure such as relative risk, 1 is the null value. A 95% confidence interval that excludes 1 is conventionally statistically significant at the two-sided 0.05 level, but observational association does not by itself establish causation.
- 90. B.** Human milk alone may not meet the protein, calcium, phosphorus, and other nutrient needs of a very preterm infant at usual volumes. Fortification increases nutrient density while preserving the advantages of human milk.
- 91. B.** Most fetal calcium and phosphorus accretion occurs during the third trimester. Preterm infants therefore require adequate enteral or parenteral delivery of both minerals to reduce the risk of metabolic bone disease.
- 92. A.** An absent red reflex may indicate a congenital cataract or another vision-threatening disorder. Early ophthalmologic evaluation is essential because prolonged visual deprivation can cause irreversible amblyopia.
- 93. D.** Umbilical arterial catheters can be associated with aortic or renal arterial thrombosis. Renal ischemia activates the renin-angiotensin system and is an important cause of neonatal renovascular hypertension.
- 94. B.** Parental authority is substantial but not unlimited when refusal creates a serious risk of preventable harm to a child. The clinical team should communicate respectfully, involve ethics resources, and seek legal protection when necessary to safeguard the infant.
- 95. C.** Omphalocele is frequently associated with other congenital anomalies, particularly cardiac defects and chromosomal or syndromic conditions. Protecting the sac and evaluating associated abnormalities are priorities before definitive repair.

- 96. B.** Phototherapy changes bilirubin molecules in the skin into structural and configurational photoisomers that are more readily excreted. This process bypasses the infant's limited hepatic conjugation capacity.
- 97. A.** Pneumatosis intestinalis in a systemically ill preterm infant with abdominal distention and bloody stool is highly characteristic of necrotizing enterocolitis. The disease represents intestinal injury with inflammation, bacterial involvement, and variable necrosis.
- 98. D.** An umbilical venous catheter should traverse the ductus venosus toward the inferior vena cava-right atrial junction. A tip within the liver or portal system is malpositioned and can cause hepatic or portal complications, especially with hyperosmolar infusions.
- 99. B.** Fatty-acid oxidation defects such as MCAD deficiency impair the ability to generate energy and ketones during fasting. Hypoglycemia with inappropriately low ketone production is therefore an important clue.
- 100. C.** Caffeine is a methylxanthine that antagonizes adenosine receptors. It increases respiratory drive, improves diaphragmatic contractility, and is standard pharmacologic therapy for apnea of prematurity.
- 101. C.** Infants with chronic lung disease may expend substantial energy on breathing and may also have fluid restrictions that limit feed volume. Nutrient density often must be optimized to support growth without excessive fluid.
- 102. B.** In hypoplastic left heart syndrome, the left ventricle and ascending aorta cannot provide adequate systemic output. Systemic perfusion depends on right-ventricular output reaching the aorta through a patent ductus arteriosus.
- 103. A.** Harlequin ichthyosis is a severe disorder of epidermal barrier formation that produces massive hyperkeratotic plates and fissures. Fluid loss, infection, temperature instability, feeding difficulty, and respiratory restriction make intensive supportive care essential.
- 104. A.** Maternal hypertension and placental insufficiency can be associated with transient neonatal neutropenia, particularly in growth-restricted preterm infants. Clinical context remains important because infection can also cause abnormal neutrophil counts.
- 105. C.** Preterm infants have limited lipid clearance, especially during illness. Significant hypertriglyceridemia should prompt reassessment of lipid delivery and clinical factors before advancing the infusion further.
- 106. A.** Gastroschisis leaves bowel exposed without a protective sac, causing heat and fluid losses and risk of injury. Initial care includes bowel protection, thermoregulation, gastric decompression, fluid resuscitation, and prompt surgical management.

- 107. C.** In neonatal resuscitation, apnea or a heart rate below 100/min after initial steps calls for positive-pressure ventilation. Ventilation is the most important intervention because bradycardia at birth is usually secondary to inadequate gas exchange.
- 108. C.** Transient tachypnea is common after delivery without labor because catecholamine-mediated and mechanical lung-fluid clearance may be delayed. Mild oxygen need, perihilar streaking, and rapid improvement distinguish it from more persistent parenchymal disease.
- 109. D.** Vitamin K is required for gamma-carboxylation of several coagulation factors. Deficiency causes impaired secondary hemostasis, often with a prolonged PT before the aPTT, while platelet count remains normal.
- 110. A.** Neurologic signs suggest acute bilirubin encephalopathy, and a bilirubin at the exchange threshold requires escalation of care. Exchange transfusion rapidly lowers bilirubin and removes circulating antibodies in immune hemolysis when present.
- 111. C.** Adequate amniotic fluid and fetal breathing movements contribute to normal lung expansion and growth. Severe prolonged oligohydramnios early in gestation can produce pulmonary hypoplasia and profound respiratory failure after birth.
- 112. D.** Erythema toxicum is a benign, self-limited eruption that commonly appears after the first day of life in otherwise well term infants. Systemic illness, grouped vesicles, or diffuse skin tenderness would point toward infectious or toxin-mediated disease instead.
- 113. B.** Noonan syndrome can resemble Turner syndrome phenotypically but occurs in either sex and usually has a normal sex-chromosome complement. Pulmonary valve stenosis and hypertrophic cardiomyopathy are important cardiac associations.
- 114. A.** Acholic stools and direct hyperbilirubinemia indicate cholestasis, not physiologic jaundice. Biliary atresia requires timely diagnosis because outcomes after portoenterostomy are better when surgery occurs early.
- 115. C.** T-cell receptor excision circles are generated during normal T-cell development. Very low or absent values identify infants with profound T-cell lymphopenia, including severe combined immunodeficiency, before serious opportunistic infections occur.
- 116. B.** The absolute risk reduction is 0.20 minus 0.10, or 0.10. The number needed to treat is the reciprocal of the absolute risk reduction:  $1/0.10 = 10$ .
- 117. C.** Ceftriaxone is generally avoided in neonates, especially those with hyperbilirubinemia or exposure to intravenous calcium, because of bilirubin displacement and the risk of calcium-ceftriaxone precipitation.

**118. D.** Prolonged parenteral nutrition, lack of enteral stimulation, prematurity, and infection all increase the risk of cholestasis. A rising direct bilirubin after weeks of parenteral nutrition is not typical physiologic jaundice.

**119. A.** Congenital myotonic dystrophy can cause severe neonatal hypotonia, respiratory insufficiency, and feeding problems. The maternal history of myotonia is a major diagnostic clue because congenital disease is usually transmitted from an affected mother.

**120. A.** Hemophilia A is an X-linked factor VIII deficiency. Affected newborns can bleed after procedures; the typical screening pattern is an isolated prolonged aPTT with normal PT and platelet count.

**121. D.** Biallelic SMN1 loss causes spinal muscular atrophy, a lower motor neuron disorder that can present in infancy with profound weakness, hypotonia, tongue fasciculations, and absent reflexes. Early recognition is important because disease-modifying therapy is time sensitive.

**122. A.** Mother's own milk is the preferred enteral nutrition for very preterm infants because it is associated with lower rates of necrotizing enterocolitis and provides immunologic and developmental benefits. Donor human milk is commonly used when maternal milk is unavailable.

**123. D.** Sensitivity is the proportion of individuals with the disease who test positive: true positives divided by all diseased individuals. It describes how well a test detects disease when disease is actually present.

**124. A.** Congenital CMV commonly causes growth restriction, petechiae, hepatosplenomegaly, thrombocytopenia, microcephaly, and periventricular calcifications. Sensorineural hearing loss may be present at birth or develop later.

**125. C.** A type I error is a false-positive conclusion: rejecting the null hypothesis when it is actually true. The alpha level is the prespecified probability of this error under the null hypothesis.

**126. B.** Drug clearance in newborns is influenced by immature hepatic metabolism and renal excretion, and critical illness can reduce clearance further. Sedatives and analgesics therefore require careful titration and clinical monitoring.

**127. C.** Cystic periventricular leukomalacia injures descending corticospinal fibers near the lateral ventricles. The legs are often affected more than the arms, producing a characteristic risk for spastic diplegia.

**128. A.** Intention-to-treat analysis preserves the benefits of randomization by analyzing participants according to original assignment, regardless of crossover or adherence. It is commonly used for the primary analysis of randomized trials.

**129. B.** Growth failure in the NICU requires assessment of delivered calories, protein, minerals, feeding losses, illness, and fluid restrictions. Increasing fluid alone may worsen respiratory or cardiac problems without correcting inadequate nutrient density.

**130. B.** Active placental transfer of maternal IgG is greatest late in gestation. Extremely preterm birth therefore interrupts a large fraction of normal antibody acquisition and contributes to vulnerability to infection.

**131. C.** Feed advancement should be guided by the infant's overall clinical tolerance, including abdominal findings, stooling, emesis, and systemic stability. New abdominal discoloration or systemic illness would instead raise concern for intestinal pathology.

**132. B.** The spinal cord usually terminates above the lower lumbar region. Lumbar puncture in neonates is generally performed at L3-L4 or L4-L5 while maintaining careful positioning and monitoring.

**133. A.** Achondroplasia produces disproportionate short stature with rhizomelic limb shortening, frontal bossing, and midface hypoplasia. The thorax is generally not as severely constricted as in lethal skeletal dysplasias.

**134. A.** Infants born to mothers with hepatitis B infection need both active and passive immunoprophylaxis after birth. Timely hepatitis B vaccine and hepatitis B immune globulin substantially reduce perinatal transmission risk.

**135. B.** Extremely preterm infants have a large surface-area-to-mass ratio and highly permeable skin, producing substantial insensible water loss. Humidity and careful fluid adjustment are therefore especially important early after birth.

**136. D.** Autonomic and gastrointestinal hyperactivity with tremors and irritability after chronic opioid exposure is typical of neonatal opioid withdrawal. Hypoxic-ischemic encephalopathy and hypothyroidism more often produce depressed tone or activity rather than this withdrawal pattern.

**137. B.** Maternal anti-Ro/SSA and anti-La/SSB antibodies can cross the placenta and injure the fetal atrioventricular conduction system. Congenital complete heart block may therefore occur even when the mother has no previously diagnosed autoimmune disease.

**138. A.** SIADH causes water retention with hypo-osmolar hyponatremia, while urine remains inappropriately concentrated and urine sodium is often elevated. Central nervous system and pulmonary disease are common neonatal triggers.

**139. A.** G6PD deficiency can cause acute oxidative hemolysis after specific exposures or illness, leading to a rapid bilirubin rise. A negative direct antiglobulin test helps distinguish it from immune-mediated hemolysis.

- 140. B.** Coagulase-negative staphylococci are common skin organisms and major causes of late-onset sepsis in very preterm infants with indwelling vascular catheters. Repeated positive cultures with compatible illness support true infection rather than simple contamination.
- 141. A.** Rapid transition from catabolism to anabolism can drive phosphate, potassium, and magnesium into cells. Growth-restricted preterm infants with limited stores are especially vulnerable to refeeding-like electrolyte abnormalities when nutrition is advanced aggressively.
- 142. C.** Shock is a state of inadequate tissue oxygen delivery, not simply a low blood pressure. Rising lactate together with oliguria and delayed capillary refill indicates impaired end-organ perfusion even if a single blood pressure measurement remains within a population reference range.
- 143. B.** Shared decision-making integrates the best available medical information with the family's values and the infant's best interests. When life-sustaining treatment is nonbeneficial or disproportionately burdensome, a comfort-focused plan can be ethically appropriate.
- 144. B.** For eligible term or near-term infants with moderate or severe hypoxic-ischemic encephalopathy, therapeutic hypothermia initiated within the accepted early treatment window reduces death or major neurodevelopmental disability. Supportive care includes careful control of ventilation, perfusion, glucose, and seizures.
- 145. D.** Right-to-left shunting through the ductus arteriosus or foramen ovale is favored when pulmonary vascular resistance remains high relative to systemic resistance. Hypoxemia, acidosis, and poor lung recruitment can all perpetuate this maladaptive state.
- 146. B.** Marked hypocapnia causes cerebral vasoconstriction and can reduce cerebral blood flow. In infants with brain injury, ventilation is therefore adjusted to avoid both significant hypocapnia and severe hypercapnia unless a specific indication exists.
- 147. A.** Chronic fetal exposure to maternal hyperglycemia stimulates fetal beta-cell hyperplasia and hyperinsulinemia. After delivery, placental glucose delivery stops abruptly while insulin remains high, producing early neonatal hypoglycemia.
- 148. A.** Extremely preterm infants have limited nutrient stores and quickly develop protein catabolism if amino acids are withheld. Early amino-acid provision reduces negative nitrogen balance and supports growth while enteral feeding is established.
- 149. B.** Aplasia cutis congenita is a focal absence of skin, most commonly on the scalp. Small superficial lesions may heal with local care, whereas larger or membranous lesions require evaluation for deeper defects and associated anomalies.
- 150. B.** For preterm infants who do not require immediate resuscitation, a short period of delayed cord clamping can improve placental transfusion and reduce transfusion needs. Immediate clamping is reserved for situations in which maternal or neonatal circumstances require it.

- 151. D.** Continuous positive airway pressure helps maintain end-expiratory lung volume and prevent alveolar collapse in a spontaneously breathing preterm infant. It can reduce the need for invasive ventilation when gas exchange and respiratory effort remain adequate.
- 152. B.** Methemoglobin absorbs light differently from oxyhemoglobin and can cause pulse-oximeter readings that remain near the mid-80s despite a relatively adequate arterial oxygen tension. Co-oximetry is required to identify the dyshemoglobin directly.
- 153. C.** Congenital diaphragmatic hernia compresses the developing lungs, producing pulmonary hypoplasia and abnormal pulmonary vasculature. Postnatal illness therefore reflects both limited gas-exchange surface area and a high risk of pulmonary hypertension.
- 154. A.** Transient neonatal pustular melanosis is a benign eruption often present at birth. Fragile superficial pustules leave characteristic hyperpigmented macules and a rim of scale without systemic illness.
- 155. D.** Twin-to-twin transfusion syndrome occurs in monochorionic pregnancies because placental vascular anastomoses create chronic net blood flow from the donor to the recipient. The donor becomes volume depleted while the recipient develops volume loading and cardiac strain.
- 156. A.** Blood-tinged or frankly bloody endotracheal secretions with acute respiratory deterioration and diffuse opacities are characteristic of pulmonary hemorrhage. It may occur in critically ill preterm infants and can be associated with hemodynamic instability or a significant ductal shunt.
- 157. B.** Blood and inflammatory debris after intraventricular hemorrhage can impair cerebrospinal-fluid circulation and resorption. Progressive ventricular enlargement after IVH is posthemorrhagic ventricular dilatation and requires close serial assessment.
- 158. D.** Exogenous surfactant lowers alveolar surface tension and improves compliance in respiratory distress syndrome. It treats the central deficiency rather than merely changing vascular tone or systemic acid-base status.
- 159. A.** Carbon dioxide is an important regulator of cerebral vascular tone. Excessive hypocarbia causes cerebral vasoconstriction and has been associated with adverse neurologic outcomes, so ventilation should avoid unnecessary overventilation.
- 160. D.** Hereditary spherocytosis can present in the newborn period with Coombs-negative hemolytic anemia and jaundice. A family history of hemolytic anemia or splenectomy and spherocytes on smear support the diagnosis.
- 161. B.** Maternal fever, uterine tenderness, fetal tachycardia, and prolonged membrane rupture are classic features of intra-amniotic infection. The fetus may be exposed to both microorganisms and inflammatory mediators, increasing risk for early-onset infection and systemic illness.

- 162. A.** In congenital diaphragmatic hernia, bag-mask ventilation can distend intrathoracic bowel and worsen lung compression. Initial stabilization therefore emphasizes endotracheal ventilation, gastric decompression, gentle ventilatory pressures, and management of pulmonary hypertension before surgical repair.
- 163. D.** An open myelomeningocele should be protected from trauma, drying, and contamination with a sterile moist nonadherent dressing while pressure on the sac is avoided. Early multidisciplinary neurosurgical evaluation is required.
- 164. C.** Treatment-requiring retinopathy of prematurity can progress rapidly. Once accepted treatment criteria are met, prompt retinal therapy by an experienced ophthalmologist is indicated rather than waiting for symptoms or term age.
- 165. A.** The kidney is highly vulnerable to systemic hypoperfusion during asphyxia. Oliguria, rising creatinine, hyperkalemia, and fluid accumulation support acute kidney injury rather than a normal postnatal adaptation.
- 166. C.** Severe fetal anemia lowers blood viscosity and increases cardiac output, producing an increased middle cerebral artery peak systolic velocity. This Doppler measurement is a standard noninvasive way to identify fetuses at risk for clinically important anemia.
- 167. A.** Holoprosencephaly, midline facial anomalies, polydactyly, and major congenital heart disease are classic features of trisomy 13. The combination strongly points to a chromosomal disorder rather than an isolated limb anomaly.
- 168. A.** Severe isolated thrombocytopenia in an otherwise well term infant with a normal maternal platelet count suggests fetal and neonatal alloimmune thrombocytopenia. Maternal antibodies target paternally inherited fetal platelet antigens and can cause intracranial hemorrhage.
- 169. C.** Continuous video EEG is the reference method for confirming neonatal electrographic seizures and correlating them with clinical events. Many neonatal seizures are electrographic-only, while many abnormal movements are nonepileptic.
- 170. B.** Neonatal HSV may occur even when maternal infection was unrecognized. Vesicular lesions in a neonate warrant urgent evaluation for disseminated, central nervous system, or skin-eye-mouth disease and prompt empiric acyclovir while diagnostic studies are obtained.
- 171. C.** Osteogenesis imperfecta results from defects in type I collagen and can present with fractures, bone fragility, deformity, and blue sclerae. Severe forms may be apparent prenatally or immediately after birth.
- 172. C.** Laryngomalacia is the most common cause of congenital stridor and typically worsens when supine, feeding, or agitated. Mild cases have preserved oxygenation and growth and improve as the laryngeal structures mature.

**173. B.** Meconium aspiration can obstruct airways, causing regional atelectasis and ball-valve air trapping, while also producing inflammatory chemical pneumonitis and surfactant dysfunction. The resulting radiograph is often heterogeneous rather than uniformly low-volume.

**174. D.** Prematurity confers risk across motor, cognitive, language, behavioral, hearing, vision, and social-emotional domains. Longitudinal surveillance and standardized screening are needed even when neonatal imaging is reassuring.

**175. A.** Respiratory distress syndrome in a very preterm infant results primarily from surfactant deficiency. High surface tension causes end-expiratory alveolar collapse, low functional residual capacity, poor compliance, and increased work of breathing.

**176. B.** Hirschsprung disease results from absence of enteric ganglion cells in the distal bowel, causing functional obstruction. Delayed meconium passage and explosive stool after rectal examination are classic clues.

**177. A.** Disseminated intravascular coagulation causes widespread coagulation activation with consumption of platelets and clotting factors, secondary fibrinolysis, and bleeding. The combination of prolonged clotting times, low fibrinogen, thrombocytopenia, and elevated D-dimer is typical.

**178. C.** Jitteriness is typically stimulus sensitive and can be suppressed by passive flexion or holding the limb. Sustained rhythmic activity that cannot be suppressed, especially with ocular or autonomic changes, is more concerning for seizure.

**179. B.** Metabolic bone disease of prematurity reflects inadequate mineral accretion in an infant who missed much of third-trimester calcium and phosphorus transfer. Low phosphorus and elevated alkaline phosphatase are common biochemical clues.

**180. D.** Central diabetes insipidus results from inadequate antidiuretic hormone release, causing inability to concentrate urine. Polyuria with dilute urine despite hypernatremia and high serum osmolality is the key pattern.

**181. C.** A higher right-hand than foot saturation reflects deoxygenated blood entering the descending aorta through a right-to-left ductal shunt. In a structurally normal heart, this pattern strongly supports persistent pulmonary hypertension of the newborn.

**182. C.** Nephrocalcinosis occurs disproportionately in very preterm infants and is associated with factors such as loop-diuretic exposure, mineral imbalance, and prolonged intensive care. Ultrasound typically shows increased echogenicity in the medullary pyramids.

**183. B.** Hyperinsulinemic hypoglycemia is suggested when insulin action is inappropriate at the time of hypoglycemia, especially with suppressed ketones and free fatty acids and a high glucose requirement. Persistent cases require evaluation beyond the transient physiology of an infant of a diabetic mother.

- 184. B.** In pulmonary atresia, pulmonary blood flow may depend on the ductus arteriosus. Maintaining ductal patency with prostaglandin E1 is therefore urgent while anatomy and definitive intervention are planned.
- 185. A.** Extreme prematurity, central access, parenteral nutrition, and broad-spectrum antibiotic exposure are important risk factors for invasive candidiasis. Persistent thrombocytopenia and sepsis despite antibacterial therapy should heighten suspicion for fungal infection.
- 186. B.** Gentamicin is cleared primarily by the kidneys and has a narrow therapeutic window. Neonatal renal function and gestational age affect clearance, so serum concentration monitoring and interval adjustment help limit nephrotoxicity and ototoxicity.
- 187. D.** A regular narrow-complex tachycardia near 250 to 300/min in a stable neonate is typical of supraventricular tachycardia. After vagal maneuvers fail, rapid intravenous adenosine can transiently block atrioventricular nodal conduction and terminate AV node-dependent reentry.
- 188. C.** Pulmonary interstitial emphysema occurs when ventilating pressure causes alveolar rupture and air dissects into the pulmonary interstitium. It is most common in very preterm infants with fragile lungs receiving positive-pressure ventilation.
- 189. D.** Maternal varicella immediately around delivery creates a period in which the infant can be exposed to virus before the mother has generated and transferred enough protective antibody. These infants require prompt risk assessment for postexposure management.
- 190. C.** Sudden deterioration with unilateral absent breath sounds and hyperexpansion strongly suggests a tension pneumothorax. In an unstable infant, decompression should not be delayed for a radiograph if clinical suspicion is high.
- 191. D.** Apnea of prematurity is primarily related to immature respiratory control. Seizures can occasionally cause apnea, but a typical apnea-bradycardia event without ictal EEG activity should not be labeled a seizure.
- 192. C.** Chromosomal microarray can detect clinically important deletions and duplications that are below the resolution of a conventional karyotype. It is commonly used early in the evaluation of an infant with unexplained multiple congenital anomalies.
- 193. B.** Obstructed total anomalous pulmonary venous return causes pulmonary venous hypertension, pulmonary edema, and severe hypoxemia. Unlike many cyanotic lesions, pulmonary blood flow cannot decompress normally because venous return itself is obstructed.
- 194. B.** Specificity is the proportion of individuals without the disease who test negative: true negatives divided by all nondiseased individuals.
- 195. C.** Primary thyroid gland failure produces low circulating thyroxine and a compensatory rise in TSH. Prompt treatment is crucial because thyroid hormone is essential for early brain development.

**196. A.** Pierre Robin sequence consists of mandibular hypoplasia leading to posterior displacement of the tongue and upper-airway obstruction; a U-shaped cleft palate is common. Airway and feeding assessment are central to management.

**197. C.** Beckwith-Wiedemann syndrome is an overgrowth disorder associated with macroglossia, abdominal-wall defects, organomegaly, and neonatal hypoglycemia from hyperinsulinism. Affected children also require tumor-surveillance planning after the newborn period.

**198. B.** Tension pneumothorax is a clinical emergency. In a rapidly deteriorating infant with convincing findings, immediate needle decompression is indicated, followed by definitive pleural drainage when needed; treatment should not be delayed for imaging.

**199. C.** Congenital rubella classically combines cataracts, congenital heart disease such as patent ductus arteriosus, and sensorineural hearing loss. Early gestational infection carries the greatest risk of major fetal anomalies.

**200. A.** Trisomy 18 commonly presents with severe fetal growth restriction, prominent occiput, micrognathia, clenched hands with overlapping fingers, rocker-bottom feet, and congenital heart disease. The phenotype is distinct from the macrosomia of Beckwith-Wiedemann syndrome.