

# COMPREHENSIVE MOCK EXAM 6

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Supplemental to: Neonatology Board Review 2026-2027

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*American Board of Pediatrics Neonatal-Perinatal Medicine subspecialty certification examination — 200 single-best-answer questions, A-D options, domain-weighted to the approved ABP blueprint*

## Board-Review Questions

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1. A 29-year-old at 29 weeks' gestation is admitted with preterm labor and intact membranes. Delivery is considered likely within 48 hours. Which antenatal intervention most directly lowers the newborn's risk of respiratory distress syndrome and intraventricular hemorrhage?

- A. Administer maternal furosemide to reduce fetal lung fluid and prevent respiratory distress.
- B. Give maternal vitamin K to increase fetal pulmonary compliance before preterm delivery.
- C. Begin maternal indomethacin solely to stimulate fetal surfactant secretion before birth.
- D. Give a course of antenatal betamethasone to accelerate fetal organ maturation.

2. A term newborn is apneic after initial warming, positioning, drying, and stimulation. The heart rate is 80/min. What is the next priority?

- A. Observe for another minute because a heart rate above 60/min excludes the need for ventilation.
- B. Begin effective positive-pressure ventilation and assess for chest movement and heart-rate response.
- C. Start chest compressions immediately before providing assisted ventilation.
- D. Give intravenous epinephrine before attempting positive-pressure ventilation.

3. A 26-week infant on day 4 has a large ductus arteriosus on echocardiography but stable blood pressure, improving respiratory support, and no evidence of systemic hypoperfusion. What is the most appropriate general approach?

- A. Use conservative observation rather than routine prophylactic or early closure solely because the ductus is patent.
- B. Perform immediate surgical ligation because any large PDA in an extremely preterm infant worsens long-term outcome.
- C. Begin scheduled indomethacin now because persistent ductal patency on day 4 is itself sufficient indication for pharmacologic closure.
- D. Arrange urgent transcatheter closure during the first week before assessing the infant's clinical trajectory.

**4.** A very preterm neonate has poor lung compliance and diffuse alveolar collapse immediately after delivery; radiography shows a fine granular pattern and low lung expansion. Which physiologic defect is primarily responsible?

- A.** Deficient pulmonary surfactant causes high alveolar surface tension and widespread atelectasis.
- B.** Meconium obstruction produces patchy air trapping and chemical pneumonitis in the preterm lung.
- C.** Pulmonary lymphatic obstruction causes chylous filling of alveoli immediately after delivery.
- D.** Delayed absorption of fetal lung fluid causes isolated interstitial edema with normal surfactant function.

**5.** A hypotonic newborn has a flat facial profile, epicanthal folds, a wide sandal gap, and an endocardial cushion defect. Which chromosomal abnormality most directly explains this phenotype?

- A.** Deletion 22q11.2 with conotruncal heart disease and hypocalcemia.
- B.** Trisomy 21 caused by an extra copy of chromosome 21.
- C.** Trisomy 18 with the characteristic pattern of clenched overlapping fingers.
- D.** Monosomy X with neonatal lymphedema and left-sided cardiac obstruction.

**6.** A 980-g infant is clinically stable and the mother is expressing milk. Which enteral feeding source is preferred as the foundation of nutrition?

- A.** Standard term formula because it provides sufficient protein and minerals without fortification.
- B.** Sterile water with parenteral nutrition until the infant reaches 34 weeks postmenstrual age.
- C.** Mother's own milk, appropriately fortified as needed for very-low-birth-weight nutrition.
- D.** Unfortified donor milk even when an adequate supply of mother's own milk is available.

7. A term newborn's serum creatinine is measured 6 hours after birth and is similar to the mother's creatinine. How should this value be interpreted?

- A. Creatinine does not cross the placenta, so any similarity to the maternal value proves neonatal renal failure.
- B. The value is determined primarily by neonatal muscle injury during delivery rather than placental equilibration.
- C. The newborn value immediately represents the infant's steady-state glomerular filtration rate independent of maternal levels.
- D. Early neonatal creatinine partly reflects maternal creatinine transferred across the placenta.

8. A 3-hour-old large-for-gestational-age infant of a diabetic mother is asymptomatic but has a very low plasma glucose on confirmatory testing. Which physiologic factor most strongly predisposes this infant to hypoglycemia?

- A. Increased ketone production that consumes circulating glucose during the first hours.
- B. Persistent hyperinsulinemia after interruption of the transplacental maternal glucose supply.
- C. Inadequate fetal insulin production caused by chronic maternal hyperglycemia.
- D. Excess neonatal glucagon secretion that drives glucose into peripheral tissues.

9. A 25-week infant has lower passive systemic antibody levels than a term infant despite a healthy mother. Which immunologic fact best explains this difference?

- A. Maternal IgM is the principal placentally transferred immunoglobulin and transfer stops after 20 weeks.
- B. Most placental transfer of maternal IgG occurs during the third trimester.
- C. Preterm infants actively destroy maternal IgG in the placenta before it enters fetal blood.
- D. Secretory IgA crosses the placenta mainly during labor and supplies the newborn's systemic antibody pool.

**10.** A 12-hour-old preterm infant develops respiratory distress, temperature instability, and hypotension after prolonged rupture of membranes and maternal intrapartum fever. Which pathogens are among the leading causes of early-onset neonatal sepsis?

- A.** *Bordetella pertussis* and *Mycoplasma pneumoniae* acquired mainly through school-age siblings.
- B.** *Clostridioides difficile* and rotavirus acquired from prolonged enteral antibiotic exposure.
- C.** Coagulase-negative staphylococci and *Candida* species acquired primarily from long-term central catheters.
- D.** Group B *Streptococcus* and *Escherichia coli* acquired around the time of birth.

**11.** On day 12, a very preterm infant who recently advanced feeds becomes lethargic with temperature instability and a tense abdomen. Abdominal imaging reveals intramural bowel gas and portal venous gas. Which disease process is most likely?

- A.** Pyloric stenosis, which causes nonbilious projectile emesis without bloody stools or pneumatosis.
- B.** Spontaneous intestinal perforation, which more often presents as an isolated perforation without diffuse pneumatosis.
- C.** Hirschsprung disease, which causes distal obstruction but not typical pneumatosis in a preterm infant on feeds.
- D.** Necrotizing enterocolitis with inflammatory intestinal injury and pneumatosis in a preterm infant.

**12.** A healthy 38-week newborn is preparing for discharge. Which approach best reflects modern prevention of severe hyperbilirubinemia?

- A.** Use visual inspection alone because transcutaneous or serum bilirubin testing is unnecessary in healthy term infants.
- B.** Measure bilirubin before discharge and plan follow-up according to gestational age, risk factors, and how close the value is to the treatment threshold.
- C.** Schedule follow-up at a fixed routine interval after discharge without incorporating the bilirubin's distance from the treatment threshold.
- D.** Begin phototherapy before discharge for breastfeeding-associated bilirubin rise even when the bilirubin remains well below the treatment threshold.

**13.** On the second day of life, a vigorous term infant develops crops of red papules and small pustules over the trunk. A smear from a pustule contains numerous eosinophils, and the palms and soles are unaffected. What is the most likely diagnosis?

- A.** Staphylococcal scalded skin syndrome, which causes tender erythema and superficial exfoliation.
- B.** Erythema toxicum neonatorum, a benign self-limited newborn eruption.
- C.** Congenital candidiasis, which can cause diffuse pustules including palms and soles and may occur at birth.
- D.** Disseminated neonatal herpes, which can produce vesicles and systemic illness and requires urgent antiviral therapy.

**14.** A 4-day-old exclusively breastfed infant who did not receive prophylaxis at birth develops oozing from venipuncture sites and gastrointestinal bleeding. PT is markedly prolonged. Which diagnosis is most likely?

- A.** Disseminated intravascular coagulation from sepsis despite an otherwise well infant and normal platelet count.
- B.** Hemophilia A causing isolated factor VIII deficiency with a normal prothrombin time.
- C.** Neonatal alloimmune thrombocytopenia causing severe thrombocytopenia rather than isolated PT prolongation.
- D.** Vitamin K deficiency bleeding due to inadequate vitamin K-dependent clotting factor activity.

**15.** A 39-week infant has moderate encephalopathy after a sentinel hypoxic event. At 3 hours of age the infant has abnormal tone, weak suck, and seizures. Which therapy offers proven neuroprotection when eligibility criteria are met?

- A.** Initiate whole-body therapeutic hypothermia within 6 hours and continue cooling for 72 hours with specialized monitoring.
- B.** Delay cooling until MRI confirms injury because treatment is most effective after the first 24 hours.
- C.** Defer cooling in this term infant and reserve therapeutic hypothermia for selected infants born before 34 weeks.
- D.** Use intermittent ice packs to lower core temperature below 31°C without continuous monitoring.

**16.** At a developmental visit, an 8-month-old former 28-week infant is being compared with term-born peers for sitting and reaching skills. The clinician subtracts the 12 weeks of prematurity. Which age should guide interpretation of early milestones?

- A.** Use chronological age for milestone comparison because correction for prematurity is unnecessary after NICU discharge.
- B.** Use a corrected age of about 5 months for early developmental assessment.
- C.** Subtract the NICU length of stay rather than the number of weeks born before term.
- D.** Use the gestational age at birth as the developmental age until school entry.

**17.** A NICU team is reviewing which infants need scheduled retinal examinations before discharge. Which infant has gestational age and birth weight that independently place the infant within routine U.S. ROP screening thresholds?

- A.** A 33-week infant weighing 2300 g with an uncomplicated course and no additional ROP risk factors.
- B.** An infant born at 29 weeks with a birth weight of 1280 g.
- C.** A healthy infant born at 35 weeks weighing 2600 g with no oxygen exposure or unstable course.
- D.** A term infant weighing 3400 g who had brief transient tachypnea and room-air recovery.

**18.** Why do many hydrophilic medications have a larger apparent volume of distribution per kilogram in very preterm infants than in older children?

- A.** Preterm infants have substantially more body fat, which expands distribution of water-soluble drugs.
- B.** Stronger neonatal albumin binding reduces the free fraction and drives hydrophilic drug distribution into tissues.
- C.** Preterm infants have a higher proportion of total body and extracellular water.
- D.** Reduced GFR decreases renal clearance and therefore directly lowers a hydrophilic drug's apparent volume of distribution.

**19.** Parents at 22 weeks' gestation ask about resuscitation options after counseling reveals a wide range of possible outcomes and substantial prognostic uncertainty. Which approach is most appropriate?

- A.** Use shared decision-making that explains prognosis and uncertainty, explores family values, and presents medically reasonable intensive and comfort-focused options.
- B.** Avoid discussing comfort-focused care because offering it automatically constitutes abandonment.
- C.** Defer detailed goals-of-care counseling until after delivery so decisions can be based on the newborn's initial appearance.
- D.** Present one institutional survival percentage and require intensive care regardless of parental goals.

**20.** In a randomized trial, NEC occurs in 12% of control infants and 8% of infants receiving an intervention. What is the absolute risk reduction?

- A.** 20 percentage points because the two event rates should be added before subtraction.
- B.** 33 percentage points, calculated as the relative reduction rather than the absolute difference.
- C.** 50 percentage points because the event rate fell from 12 to 8.
- D.** An absolute risk reduction of 4 percentage points (12% minus 8%).

**21.** A 29-week newborn has a heart rate above 100/min and is breathing spontaneously but has grunting, retractions, and an increasing oxygen requirement. Which delivery-room respiratory support strategy best limits avoidable intubation?

- A.** Plan immediate endotracheal ventilation based on gestational age before a CPAP trial.
- B.** Use supplemental oxygen alone without distending pressure because CPAP worsens functional residual capacity.
- C.** Provide early nasal CPAP while monitoring work of breathing, oxygen need, and gas exchange.
- D.** Observe without positive-pressure support until a blood gas documents respiratory acidosis.

**22.** A severely growth-restricted infant has a small jaw, prominent occiput, short sternum, and persistent flexion of the hands with the index finger overlapping the third digit. The feet have a convex rocker-bottom appearance. Which diagnosis is most likely?

- A.** Turner syndrome, which more often causes lymphedema, webbed neck, and coarctation.
- B.** Beckwith-Wiedemann syndrome, which causes overgrowth, macroglossia, and omphalocele.
- C.** Trisomy 13, which more often features holoprosencephaly, clefting, and postaxial polydactyly.
- D.** Trisomy 18 (Edwards syndrome), matching the overlapping-finger and rocker-bottom-foot phenotype.

**23.** A VLBW infant cannot receive enough mother's own milk. Which alternative is generally favored to reduce necrotizing enterocolitis risk when available?

- A.** High-calorie term formula without considering donor milk because formula lowers NEC risk.
- B.** Pasteurized donor human milk, with appropriate fortification for growth.
- C.** Unmodified cow's milk because intact bovine protein promotes intestinal adaptation in preterm infants.
- D.** Clear glucose-electrolyte solution as the sole enteral feeding until discharge.

- 24.** A preterm infant with recent fluid losses has oliguria, rising blood urea nitrogen, cool extremities, and a low fractional excretion of sodium. Which renal process is most likely?
- A.** Prerenal kidney dysfunction from reduced renal perfusion with intact tubular sodium conservation.
  - B.** Established acute tubular necrosis with impaired sodium reabsorption and typically higher fractional sodium excretion.
  - C.** Posterior urethral valves producing bilateral hydronephrosis despite the low fractional excretion of sodium.
  - D.** Renal salt wasting from immature tubules despite clinical evidence of intravascular depletion.
- 25.** A newborn screen shows a markedly elevated TSH and low thyroxine in an infant who appears clinically well. What is the most important management principle?
- A.** Start thyroid hormone replacement promptly while confirming the diagnosis because early treatment protects neurodevelopment.
  - B.** Repeat thyroid testing over the first year before starting therapy because asymptomatic neonatal TSH elevation rarely affects neurodevelopment.
  - C.** Use antithyroid medication because elevated TSH indicates neonatal hyperthyroidism.
  - D.** Wait for clinical signs such as macroglossia and prolonged jaundice before initiating therapy.
- 26.** A breastfed newborn receives substantial mucosal immune protection despite relatively low systemic absorption of milk antibodies. Which immunoglobulin is most important in this role?
- A.** IgD is the major antibody in colostrum and directly activates neonatal complement in the bloodstream.
  - B.** IgM from human milk crosses the intact neonatal gut in large amounts and becomes the dominant serum immunoglobulin.
  - C.** IgE in human milk provides broad antibacterial neutralization in the intestinal lumen.
  - D.** Secretory IgA in human milk coats mucosal surfaces and helps limit pathogen adherence.

**27.** At 12 days of age, an infant develops focal seizures and rapidly rising aminotransferases; clustered vesicles are then found on the scalp. Which treatment should be started before virologic confirmation returns?

- A.** Intravenous acyclovir for suspected neonatal herpes simplex virus infection.
- B.** Oral amoxicillin alone because skin vesicles and hepatitis are typical of group B streptococcal infection.
- C.** Intravenous ganciclovir because HSV and congenital CMV have identical acute treatment.
- D.** No antiviral therapy until culture confirmation because empiric acyclovir worsens neonatal outcomes.

**28.** A 24-week infant on day 6 suddenly develops abdominal distention and free intraperitoneal air but has little preceding systemic illness and no pneumatosis. Which diagnosis is most likely?

- A.** Spontaneous intestinal perforation causing an isolated early focal bowel perforation.
- B.** Malrotation with volvulus, which typically causes bilious emesis and ischemia in a more proximal obstructive pattern.
- C.** Meconium ileus, which presents from birth with distal obstruction rather than acute focal perforation after several days.
- D.** Classic necrotizing enterocolitis with diffuse intestinal inflammation and pneumatosis as a defining feature.

**29.** A 2-day-old infant develops rapidly rising unconjugated bilirubin after exposure to an oxidant drug. The infant has anemia and bite cells on smear. Which diagnosis is most likely?

- A.** Breast-milk jaundice causing intravascular hemolysis and Heinz-body formation.
- B.** Biliary atresia causing hemolysis and unconjugated bilirubin after oxidant exposure.
- C.** Glucose-6-phosphate dehydrogenase deficiency with oxidant-triggered hemolysis.
- D.** Physiologic jaundice causing bite cells and acute anemia from oxidative injury.

**30.** During the first newborn examination, several flaccid vesiculopustules are already ruptured and dark macules remain beneath a thin rim of scale. The baby is afebrile and feeds normally. Which benign eruption best explains these findings?

- A.** Erythema toxicum, which usually arises after birth on an erythematous base and lacks persistent hyperpigmented macules.
- B.** Neonatal HSV, which produces grouped vesicles and may progress to CNS or disseminated disease.
- C.** Bullous impetigo, which is an infectious blistering disorder rather than a benign congenital pustulosis.
- D.** Transient neonatal pustular melanosis, a benign pustular eruption present at birth.

**31.** A term infant is plethoric, lethargic, and hypoglycemic with a venous hematocrit of 70%. Which diagnosis best explains the symptoms?

- A.** Vitamin K deficiency causing bleeding with a normal or reduced hematocrit.
- B.** Hereditary spherocytosis causing hemolysis and reticulocytosis rather than plethoric hyperviscosity.
- C.** Neonatal polycythemia with hyperviscosity impairing microvascular flow.
- D.** Anemia of prematurity causing reduced oxygen-carrying capacity and low hematocrit.

**32.** A 25-week infant has a routine cranial ultrasound on day 7 showing blood confined to the germinal matrix with a small amount extending into the lateral ventricle but no ventricular dilation. Which injury is being described?

- A.** Hydranencephaly, which represents near-complete absence of cerebral hemispheres.
- B.** Periventricular leukomalacia, which is a white-matter ischemic injury rather than acute ventricular blood.
- C.** A low-grade germinal matrix-intraventricular hemorrhage of prematurity.
- D.** Subdural hemorrhage from traumatic delivery, which lies outside the brain parenchyma and ventricles.

**33.** An ELBW infant is preparing for discharge after a complicated NICU course. Which follow-up plan best addresses neurodevelopmental risk?

- A.** Use standard primary-care follow-up and postpone standardized developmental assessment until school-entry age.
- B.** Enroll in high-risk infant follow-up with standardized developmental surveillance, hearing and vision follow-up, and early-intervention referral when indicated.
- C.** Wait until a major delay is obvious before involving therapy services because early intervention does not affect function.
- D.** Focus solely on weight gain because neurologic and sensory outcomes are unrelated to extreme prematurity.

**34.** A premature infant is diagnosed with type 1 retinopathy of prematurity in zone I. What is the management principle?

- A.** Defer further retinal examinations until vascularization is complete because examination itself can aggravate neovascularization.
- B.** Use systemic corticosteroids as definitive therapy because ROP is primarily an inflammatory retinal disease.
- C.** Arrange prompt treatment by an experienced ophthalmologist, using retinal laser or anti-VEGF therapy according to disease features and follow-up capability.
- D.** Observe until retinal detachment develops because treatment before stage 5 increases visual loss.

**35.** An aminoglycoside is prescribed for a preterm newborn. Why are dosing intervals often longer than in older infants?

- A.** Hepatic sequestration prolongs aminoglycoside exposure, making once-daily dosing necessary despite normal renal elimination.
- B.** Immature glomerular filtration slows renal elimination and prolongs the drug's half-life.
- C.** Neonatal hepatic CYP induction rapidly metabolizes aminoglycosides, requiring longer intervals to prevent low peaks.
- D.** Aminoglycosides are eliminated mainly through bile, which is absent in preterm infants.

**36.** A parent with limited English proficiency must provide consent for a high-risk procedure. A bilingual relative offers to translate. What is the best communication approach?

- A.** Use the bilingual relative as the primary interpreter because familiarity with the family improves accuracy for procedural consent.
- B.** Proceed without interpretation if the consent form has already been signed in English.
- C.** Ask the parent to rely on written discharge material because spoken interpretation can introduce bias.
- D.** Use a qualified medical interpreter and communicate directly with the parent while confirming understanding.

**37.** A therapy lowers the risk of severe BPD from 20% to 15%. What is the number needed to treat, using the absolute risk difference?

- A.** 25 infants, because the relative risk reduction is 25%.
- B.** 100 infants, because the absolute reduction is 5 percentage points.
- C.** 5 infants, because the treated event rate is 15% and the control rate is 20%.
- D.** 20 infants need treatment to prevent one additional event.

**38.** A pregnant patient at 27 weeks has severe preeclampsia and will undergo delivery for worsening maternal status. Which maternal medication is given primarily for fetal neuroprotection when very preterm birth is imminent?

- A.** Start prolonged maternal oxygen therapy to prevent cerebral palsy in the preterm infant.
- B.** Administer antenatal digoxin to reduce neonatal intraventricular hemorrhage after birth.
- C.** Administer magnesium sulfate before delivery when there is time to do so.
- D.** Give maternal sodium bicarbonate to prevent fetal metabolic acidosis during delivery.

**39.** After 30 seconds of demonstrably effective positive-pressure ventilation through an advanced airway, a newborn's heart rate remains 50/min. What should the team do next?

- A.** Stop ventilation and provide chest compressions alone at the pediatric basic life support rate.
- B.** Continue ventilation alone for several additional minutes because compressions are contraindicated in newborns.
- C.** Begin coordinated chest compressions with ventilation using a 3:1 compression-to-ventilation ratio.
- D.** Administer sodium bicarbonate before starting compressions to correct presumed metabolic acidosis.

**40.** A 25-week infant remains ventilator-dependent at 3 weeks with pulmonary overcirculation and systemic steal attributed to a persistent hemodynamically significant PDA. Which strategy is reasonable after individualized assessment?

- A.** Use prophylactic acetaminophen indefinitely because long courses improve neurodevelopmental outcome.
- B.** Proceed directly to open surgical ligation without considering medical or transcatheter options.
- C.** Continue observation without considering closure after 14 days because late medical or procedural treatment has no role in persistent hemodynamically significant PDA.
- D.** Consider a medical closure attempt such as ibuprofen, with procedural closure reserved for selected persistent cases.

**41.** A breastfed term newborn has lost 13% of birth weight and is lethargic with serum sodium of 158 mEq/L. What is the most likely underlying problem?

- A.** Congenital adrenal hyperplasia causing sodium loss, hyperkalemia, and volume depletion.
- B.** Hypernatremic dehydration from inadequate milk intake with proportionally greater water than sodium loss.
- C.** Renal salt wasting causing hyponatremia from excessive urinary sodium loss.
- D.** Syndrome of inappropriate antidiuretic hormone secretion causing water retention and concentrated hyponatremia.

- 42.** A 46,XX newborn with virilized external genitalia later develops weight loss, vomiting, hyperkalemia, and hyponatremia; the newborn screen shows markedly elevated 17-hydroxyprogesterone. Which enzyme deficiency is most likely?
- A.** 11-beta-hydroxylase deficiency causing mineralocorticoid excess with hypertension rather than salt wasting.
  - B.** 21-hydroxylase deficiency causing salt-wasting congenital adrenal hyperplasia.
  - C.** 17-alpha-hydroxylase deficiency causing undervirilization with hypertension and hypokalemia.
  - D.** Thyroid peroxidase deficiency causing hypothyroidism without adrenal salt loss.
- 43.** A newborn has persistent lymphopenia, chronic diarrhea, oral candidiasis, and severe viral and bacterial infections. Which primary immune defect should be urgently considered?
- A.** Hereditary spherocytosis, which causes hemolysis but does not produce persistent lymphopenia.
  - B.** Complement C5 deficiency alone, which predisposes mainly to *Neisseria* rather than broad opportunistic infection.
  - C.** Severe combined immunodeficiency with profound T-cell dysfunction and variable B-cell impairment.
  - D.** Selective IgA deficiency, which rarely presents in the newborn with life-threatening opportunistic infections.
- 44.** A growth-restricted newborn has petechiae, jaundice, hepatosplenomegaly, periventricular calcifications, and sensorineural hearing loss. Which congenital infection is most likely?
- A.** Congenital syphilis, which causes snuffles, rash, and long-bone abnormalities rather than periventricular calcifications.
  - B.** Congenital toxoplasmosis, which more classically causes diffuse intracranial calcifications, hydrocephalus, and chorioretinitis.
  - C.** Congenital cytomegalovirus infection with the characteristic periventricular calcification pattern.
  - D.** Congenital rubella, which more typically causes cataracts, hearing loss, and patent ductus arteriosus.

**45.** A 5-week-old infant has persistent jaundice with dark urine, pale acholic stools, hepatomegaly, and elevated direct bilirubin. Which diagnosis must be evaluated urgently?

- A.** Breast-milk jaundice causing isolated unconjugated hyperbilirubinemia with normally pigmented stools.
- B.** Biliary atresia causing progressive extrahepatic obstruction and cholestasis.
- C.** Physiologic jaundice persisting normally for several months with direct hyperbilirubinemia.
- D.** G6PD deficiency causing acholic stools by obstructing the extrahepatic bile ducts.

**46.** A 3-week-old infant has jaundice and a direct bilirubin concentration that is clearly elevated. What is the most important interpretation?

- A.** Breast-milk jaundice typically causes isolated direct hyperbilirubinemia with pale stools.
- B.** Use intensive phototherapy as the primary treatment for direct hyperbilirubinemia while deferring hepatobiliary evaluation.
- C.** Direct bilirubin elevation is a normal extension of physiologic jaundice during the first two months.
- D.** Conjugated hyperbilirubinemia is pathologic and requires evaluation for cholestasis and hepatobiliary disease.

**47.** A hospitalized newborn develops fever, diffuse tender erythema, superficial blistering, and a positive Nikolsky sign. Mucous membranes are relatively spared. Which diagnosis is most likely?

- A.** Incontinentia pigmenti, which follows a staged Blaschko-line eruption rather than acute febrile desquamation.
- B.** Staphylococcal scalded skin syndrome from exfoliative toxin-producing *Staphylococcus aureus*.
- C.** Epidermolysis bullosa from inherited skin fragility without fever or toxin-mediated tenderness.
- D.** Erythema toxicum neonatorum, which is benign and does not cause diffuse exfoliation.

**48.** A firstborn term infant has diffuse petechiae at birth and a platelet count of 12,000/ $\mu$ L. The mother has a normal platelet count and no autoimmune disease. Which diagnosis is most likely?

- A.** Maternal immune thrombocytopenia, which usually affects a mother who also has thrombocytopenia or a known autoimmune history.
- B.** Neonatal alloimmune thrombocytopenia from maternal antibodies against a paternally inherited fetal platelet antigen.
- C.** Vitamin K deficiency, which impairs coagulation factors but does not usually produce profound isolated thrombocytopenia.
- D.** Hemophilia B, which causes factor IX deficiency with normal platelet numbers.

**49.** A former 27-week infant later develops spastic diplegia. Neonatal imaging had shown cystic injury adjacent to the lateral ventricles. Which diagnosis best explains the pattern?

- A.** Subarachnoid hemorrhage from birth trauma, which does not classically cause symmetric periventricular cystic injury.
- B.** Neonatal meningitis limited to the meninges without white-matter injury.
- C.** Periventricular leukomalacia affecting vulnerable preterm cerebral white matter.
- D.** Basal ganglia injury from acute profound asphyxia, which more often causes dyskinetic motor abnormalities.

**50.** A former extremely preterm toddler has persistent scissoring of the legs, toe walking, and increased lower-extremity tone more than upper-extremity tone. Which motor diagnosis is most likely?

- A.** Benign hypotonia of infancy, which produces decreased rather than increased muscle tone.
- B.** Peripheral neuropathy causing areflexia and flaccid weakness rather than spasticity.
- C.** Spastic diplegic cerebral palsy, often associated with preterm white-matter injury.
- D.** Isolated developmental coordination disorder, which does not cause fixed hypertonia and scissoring.

**51.** A newborn has a dense unilateral congenital cataract obscuring the visual axis. Why is prompt ophthalmologic management important?

- A.** Congenital cataracts routinely clear spontaneously by 6 months, so early treatment risks unnecessary surgery.
- B.** The main danger is retinal detachment within hours of birth rather than deprivation amblyopia.
- C.** Defer cataract treatment until visual maturation is established because early infancy contributes little to cortical visual development.
- D.** Early visual deprivation can cause irreversible amblyopia during a critical period of visual development.

**52.** A newborn receives chloramphenicol at doses appropriate for an older child and develops abdominal distention, vomiting, cyanosis, hypotension, and gray discoloration. What pharmacologic problem is responsible?

- A.** Immature hepatic glucuronidation and renal clearance cause toxic chloramphenicol accumulation, producing gray baby syndrome.
- B.** High neonatal albumin binding prevents chloramphenicol distribution and causes isolated tissue hypoxia.
- C.** Rapid renal elimination causes a withdrawal syndrome when chloramphenicol levels fall too quickly.
- D.** Excess neonatal CYP3A4 activity converts chloramphenicol into a toxic metabolite too rapidly.

**53.** A critically ill newborn has a poor prognosis and ongoing invasive treatments are no longer achieving the agreed goals of care. Which ethical principle best supports discussing withdrawal of burdensome life-sustaining treatment?

- A.** Comfort medication should be withheld after withdrawal because symptom relief could unintentionally shorten life.
- B.** Any treatment that can prolong biologic life must continue regardless of suffering or achievable goals.
- C.** Treatment may be forgone when its burdens outweigh expected benefits and the plan remains centered on the infant's best interests and family values.
- D.** Withdrawing an intervention is ethically different from withholding the same intervention even when the intent is identical.

**54.** A screening assay is designed to miss very few affected newborns, even at the cost of some false-positive results. Which clinical interpretation follows from this test characteristic?

- A.** A positive result necessarily proves disease regardless of specificity or pretest probability.
- B.** High sensitivity makes a positive result highly predictive of disease even when the underlying prevalence is very low.
- C.** A negative result can be useful for ruling out disease when sensitivity is sufficiently high and the test is applied appropriately.
- D.** Sensitivity describes the proportion of healthy infants who correctly test negative.

**55.** A fetus with severe early-onset growth restriction has a small abdominal circumference, relatively preserved head circumference, and abnormal umbilical artery Doppler flow. Which mechanism best explains this growth pattern?

- A.** A primary skeletal dysplasia selectively limits abdominal growth while preserving placental flow.
- B.** Placental insufficiency causes preferential redistribution of blood flow toward the brain.
- C.** Maternal hyperthyroidism causes isolated hepatic enlargement with reduced head growth.
- D.** A fetal insulin excess causes proportionate overgrowth of the abdomen and subcutaneous tissues.

**56.** A newborn receiving chest compressions and effective ventilation still has a heart rate below 60/min. Umbilical venous access has been obtained. Which medication is indicated?

- A.** Give calcium gluconate routinely because hypocalcemia is the usual cause of delivery-room bradycardia.
- B.** Give atropine to reverse presumed vagal bradycardia before continuing ventilation.
- C.** Give epinephrine by the intravascular route while continuing coordinated ventilation and compressions.
- D.** Give adenosine because severe neonatal bradycardia is usually an atrioventricular nodal reentry rhythm.

**57.** A term newborn has severe cyanosis that improves little with oxygen. Chest radiograph shows a narrow mediastinum, and echocardiography confirms transposition of the great arteries with restrictive atrial mixing. What is the most urgent intervention?

- A.** Give indomethacin to close the ductus and reduce pulmonary overcirculation before transfer.
- B.** Restrict fluids and wait for spontaneous closure of the foramen ovale to improve oxygenation.
- C.** Administer a beta blocker to reduce infundibular obstruction as the primary therapy.
- D.** Start prostaglandin E1 and arrange urgent balloon atrial septostomy to improve mixing.

**58.** A 28-week infant on CPAP has escalating oxygen need and work of breathing consistent with surfactant-deficient RDS. Which intervention most directly treats the underlying deficiency?

- A.** Administer exogenous surfactant using the least invasive effective technique available for the infant.
- B.** Give systemic furosemide to remove alveolar surfactant inhibitors before continuing CPAP.
- C.** Start inhaled nitric oxide because pulmonary vascular resistance is the primary lesion in RDS.
- D.** Increase inspired oxygen alone because oxygen replacement corrects the missing phospholipid layer.

**59.** A newborn has microphthalmia, a cleft lip and palate, postaxial polydactyly, and a severe forebrain malformation. Which diagnosis is most likely?

- A.** Achondroplasia with rhizomelic limb shortening and macrocephaly.
- B.** Trisomy 18 with overlapping fingers and rocker-bottom feet but fewer midline clefting defects.
- C.** Trisomy 13 (Patau syndrome) with characteristic midline defects and postaxial polydactyly.
- D.** Noonan syndrome with pulmonic stenosis, hypertelorism, and webbed neck.

**60.** A 27-week infant is tolerating increasing volumes of human milk but growth and serum phosphorus are suboptimal. What is the most appropriate nutritional adjustment?

- A.** Dilute the milk with sterile water to reduce renal solute load and improve mineral retention.
- B.** Add a human-milk fortifier to increase protein, calcium, phosphorus, and energy delivery.
- C.** Pause enteral feeds and meet nutrition needs with intravenous dextrose until serum phosphorus normalizes.
- D.** Replace human milk with an unfortified term formula because fortification increases NEC risk.

**61.** Prenatal history includes frequent handling of cat litter and undercooked meat. Postnatal imaging shows ventriculomegaly with scattered intracranial calcifications, and ophthalmologic examination reveals chorioretinitis. Which congenital infection is most likely?

- A.** Congenital rubella, which is associated with cataracts and patent ductus arteriosus.
- B.** Congenital syphilis, which is associated with rash, hepatosplenomegaly, and long-bone disease.
- C.** Congenital toxoplasmosis, classically associated with hydrocephalus, chorioretinitis, and diffuse calcifications.
- D.** Congenital CMV, which more often produces periventricular calcifications and microcephaly.

**62.** A term newborn develops bilious emesis and abdominal pain. Upper gastrointestinal contrast study shows an abnormal position of the duodenojejunal junction and a corkscrew appearance. What is the most urgent diagnosis?

- A.** Intestinal malrotation with midgut volvulus causing acute bilious obstruction and threatened bowel ischemia.
- B.** Uncomplicated gastroesophageal reflux, which does not cause bilious vomiting or bowel ischemia.
- C.** Hypertrophic pyloric stenosis, which causes nonbilious emesis and typically presents later in infancy.
- D.** Hirschsprung disease, which causes distal functional obstruction without a corkscrew upper-GI pattern.

**63.** A newborn with severe unconjugated hyperbilirubinemia is treated with intensive phototherapy. How does phototherapy lower bilirubin?

- A.** Light directly increases hepatic UDP-glucuronosyltransferase production within minutes and permanently cures enzyme deficiency.
- B.** Light binds albumin and prevents bilirubin from leaving the intravascular compartment.
- C.** Light transforms conjugated bilirubin into unconjugated bilirubin for renal excretion.
- D.** Light converts bilirubin into more water-soluble photoisomers that can be excreted without hepatic conjugation.

**64.** Removal of routine monitoring adhesive causes new erosions on a neonate's skin, and gentle friction produces additional blisters. The parents report similar blistering with minor trauma in multiple family members. Which diagnosis best fits?

- A.** Transient neonatal pustular melanosis, which leaves hyperpigmented macules rather than trauma-induced blisters.
- B.** Physiologic desquamation, which does not cause deep blisters or a familial pattern of mechanical fragility.
- C.** Epidermolysis bullosa, an inherited disorder of skin structural proteins causing mechanical fragility.
- D.** Staphylococcal scalded skin syndrome, which is an acute toxin-mediated illness with diffuse tenderness and systemic signs.

**65.** A mother with immune thrombocytopenia has a platelet count of 45,000/ $\mu$ L near delivery. Her newborn has moderate thrombocytopenia without other abnormalities. Which mechanism is most likely?

- A.** Transplacental maternal antiplatelet IgG is causing immune destruction of neonatal platelets.
- B.** Vitamin K deficiency has reduced platelet production in the neonatal bone marrow.
- C.** Maternal platelet transfusion across the placenta has diluted the infant's circulating platelet pool.
- D.** An inherited thrombopoietin deficiency in the infant is the expected mechanism when maternal immune thrombocytopenia is present.

**66.** A term neonate has recurrent electrographic seizures after an acute brain injury. Glucose and electrolytes are normal. Which medication is commonly used as first-line treatment for neonatal seizures?

- A.** Use oral carbamazepine as the routine initial agent for acute neonatal seizures after metabolic causes are excluded.
- B.** Diazepam by repeated intramuscular injections because prolonged sedation improves neurodevelopment.
- C.** Phenobarbital, while the underlying cause is simultaneously identified and treated.
- D.** No antiseizure medication because electrographic seizures without major motor activity are harmless.

**67.** A preterm infant has normal tone at term-equivalent age but remains at high risk for later language and executive-function difficulties. What does this illustrate about neurodevelopmental prognosis?

- A.** Reserve structured developmental surveillance for infants who show major motor abnormalities before discharge.
- B.** Preterm neurodevelopmental problems are limited to motor function and do not affect learning or behavior.
- C.** A normal early motor examination does not exclude later cognitive, language, attention, or executive-function difficulties.
- D.** Normal tone at term-equivalent age strongly predicts normal later cognition and behavior, so specialized follow-up can be discontinued.

**68.** A NICU graduate who required intensive care for more than 5 days needs newborn hearing screening. Which test strategy is preferred to detect auditory neuropathy as well as cochlear hearing loss?

- A.** Visual reinforcement audiometry before discharge because it is validated in preterm newborns.
- B.** Automated auditory brainstem response rather than relying only on otoacoustic emissions.
- C.** Behavioral response to hand clapping because physiologic hearing tests are inaccurate in newborns.
- D.** Tympanometry alone because middle-ear compliance identifies neural hearing disorders.

**69.** Which property makes ceftriaxone generally avoided in many neonates, especially those with hyperbilirubinemia or receiving calcium-containing intravenous solutions?

- A.** It can displace bilirubin from albumin and can precipitate with intravenous calcium.
- B.** It causes severe neonatal hypoglycemia by directly stimulating pancreatic insulin release.
- C.** It irreversibly blocks pulmonary surfactant production and causes acute respiratory distress syndrome.
- D.** It is not active against any neonatal bacterial pathogen and therefore has no clinical use.

**70.** A NICU quality review finds that families from certain neighborhoods have less access to follow-up and higher missed-appointment rates. Which response best addresses the problem?

- A.** Exclude families with missed appointments from outcome measurement to avoid biasing the program's results.
- B.** Keep identical appointment and referral processes across neighborhoods so the health system does not treat groups differently.
- C.** Attribute the disparity primarily to parental adherence and intensify counseling rather than altering system processes.
- D.** Examine structural barriers such as transportation, language access, scheduling, and referral processes and redesign care with affected families.

**71.** A diagnostic test has 95% specificity. What does this number describe?

- A.** The disease prevalence is 5% in the population being tested.
- B.** Among infants without the disease, 95% will have a negative test result.
- C.** Among infants with the disease, 95% will have a positive test result.
- D.** Specificity of 95% means that about 95% of positive tests are true positives across clinical settings.

**72.** A term infant born to a mother with poorly controlled pregestational diabetes is plethoric and macrosomic. At 2 hours of life the infant is jittery with a plasma glucose of 24 mg/dL. What is the principal pathophysiologic cause?

- A.** Abrupt loss of maternal insulin causes transient neonatal insulin deficiency and hyperglycemia.
- B.** Reduced fetal glycogen stores from chronic maternal hypoglycemia limit hepatic glucose production.
- C.** Placental glucagon transfer suppresses neonatal ketogenesis and causes early postnatal hypoglycemia.
- D.** Persistent fetal hyperinsulinemia continues after the maternal glucose supply is interrupted at birth.

**73.** A nonvigorous term infant is born through meconium-stained fluid and has poor respiratory effort. What should happen first after the initial steps?

- A.** Provide chest compressions before any ventilation because meconium exposure implies severe asphyxia.
- B.** Delay respiratory support while obtaining a chest radiograph to confirm meconium aspiration.
- C.** Prioritize assisted ventilation rather than delaying ventilation for routine tracheal suctioning.
- D.** Perform immediate routine tracheal suctioning until no meconium is recovered, then assess breathing.

**74.** A neonate becomes pale with weak femoral pulses and metabolic acidosis at 2 days of age as the ductus begins to close. Upper-extremity pulses remain stronger. Which treatment should be started while definitive anatomy is evaluated?

- A.** Use adenosine because the pulse discrepancy indicates supraventricular tachycardia.
- B.** Begin prostaglandin E1 to restore ductal support of systemic blood flow.
- C.** Give indomethacin to accelerate ductal closure and reduce left-to-right shunting.
- D.** Start inhaled nitric oxide because the presentation is most consistent with isolated pulmonary vascular disease.

**75.** Several hours after an elective cesarean birth, a near-term infant remains tachypneic but needs little supplemental oxygen. The lungs are mildly hyperinflated with fissural fluid, and symptoms begin to improve during the first day. Which diagnosis is most likely?

- A.** Transient tachypnea of the newborn from delayed clearance of fetal lung fluid.
- B.** Respiratory distress syndrome from severe primary surfactant deficiency in a term infant.
- C.** Meconium aspiration syndrome despite clear amniotic fluid and no airway obstruction.
- D.** Congenital lobar emphysema causing focal hyperinflation and mediastinal shift.

**76.** A female newborn has puffy hands and feet, a broad shield chest, a webbed neck, and diminished femoral pulses. Which genetic diagnosis should be suspected?

- A.** Prader-Willi syndrome, which causes neonatal hypotonia and feeding difficulty without lymphedema.
- B.** Turner syndrome from complete or partial monosomy X with congenital lymphedema and coarctation risk.
- C.** Trisomy 13, which commonly causes polydactyly and severe midline malformations.
- D.** Klinefelter syndrome, which usually presents later in phenotypic males with hypogonadism.

**77.** A 25-week infant cannot yet tolerate full enteral nutrition. Which parenteral-nutrition principle is most appropriate early after birth?

- A.** Provide amino acids and energy early while advancing enteral feeds as clinical tolerance permits.
- B.** Postpone amino-acid provision until substantial enteral volumes are tolerated because early parenteral amino acids increase catabolism.
- C.** Withhold amino acids for the first week because preterm kidneys cannot excrete nitrogen at any dose.
- D.** Use dextrose alone until birth weight is regained, then begin protein after two weeks.

**78.** A 25-week infant in the second postnatal week has hyponatremia despite appropriate fluid balance. Urine sodium is elevated and there is no edema. What is the most likely mechanism?

- A.** Excess aldosterone activity causing inappropriate renal sodium retention and dilutional hyponatremia.
- B.** Renal sodium wasting from tubular immaturity in an extremely preterm infant.
- C.** Complete absence of glomerular filtration causing no urinary sodium excretion.
- D.** Hypertonic dehydration from insufficient free water intake, which should raise rather than lower serum sodium.

**79.** A preterm infant becomes cold after transport and then develops increased oxygen consumption and hypoglycemia. Which mechanism links cold stress to these metabolic problems?

- A.** Shivering is the dominant neonatal heat source and directly causes metabolic alkalosis.
- B.** Nonshivering thermogenesis increases catecholamine-driven metabolism and glucose and oxygen use.
- C.** Peripheral vasodilation during cold stress increases heat retention but lowers glucose use.
- D.** Cold exposure suppresses brown-fat metabolism, reducing oxygen consumption while causing hyperglycemia.

**80.** A newborn with a conotruncal heart defect and hypocalcemia has a very small thymus and low T-cell numbers. Which immunologic problem is most likely?

- A.** T-cell deficiency from thymic hypoplasia associated with 22q11.2 deletion syndrome.
- B.** Transient low maternal IgG transfer caused by post-term gestation.
- C.** Neutrophil oxidative-burst failure from chronic granulomatous disease.
- D.** Isolated terminal complement deficiency presenting predominantly with recurrent meningococcal infection.

**81.** A former 25-week infant at 6 weeks of age has a slowly falling hemoglobin, low reticulocyte response, and frequent prior phlebotomy losses but no hemolysis. What is the main mechanism?

- A.** Anemia of prematurity from low endogenous erythropoietin response compounded by phlebotomy and rapid growth.
- B.** Autoimmune hemolytic anemia from maternal IgM antibodies persisting for several months.
- C.** Vitamin K deficiency causing chronic red-cell destruction without bleeding.
- D.** Polycythemia caused by excessive erythropoietin production after preterm birth.

**82.** A term infant develops focal clonic movements of the right arm at 12 hours of age. MRI shows an acute left middle cerebral artery infarction. Which diagnosis is most likely?

- A.** Benign sleep myoclonus occurring during sleep and stopping with arousal despite the associated MRI lesion.
- B.** Generalized hypocalcemic jitteriness, which is stimulus-sensitive and lacks a focal arterial lesion.
- C.** Perinatal arterial ischemic stroke presenting with focal neonatal seizures.
- D.** Apnea of prematurity, which occurs in preterm infants and does not cause focal cortical infarction.

**83.** A former VLBW infant has a normal newborn hearing screen but had prolonged aminoglycoside exposure and meningitis. Which follow-up principle is appropriate?

- A.** Defer formal hearing testing until school age because early amplification has little effect on language outcomes.
- B.** No further hearing assessment is needed because a passed newborn screen excludes future sensorineural hearing loss.
- C.** Rely on parental observation rather than formal audiology because behavioral hearing tests are unreliable in former preterm infants.
- D.** Continue audiologic surveillance because later-onset or progressive hearing loss can occur despite an initial pass.

**84.** A newborn becomes cyanotic during quiet breathing and feeding but pinks up when crying. Both nares resist catheter passage. Which diagnosis should be suspected?

- A.** Laryngomalacia, which causes inspiratory stridor rather than inability to pass a nasal catheter.
- B.** Pierre Robin sequence, which causes tongue-base airway obstruction from micrognathia rather than posterior nasal blockage.
- C.** Bilateral choanal atresia producing posterior nasal obstruction in a newborn.
- D.** Vocal-cord paralysis, which causes stridor and weak cry but does not obstruct both nasal passages.

**85.** Investigators plan a trial to show that a less invasive surfactant method is not unacceptably worse than standard therapy. Before enrollment, which design element must be defined to make a valid noninferiority conclusion?

- A.** A requirement that the new method achieve statistical superiority before noninferiority can be assessed.
- B.** Treat confidence intervals as optional because noninferiority conclusions depend primarily on the P value.
- C.** A post hoc margin chosen after results are known so the new treatment meets the desired conclusion.
- D.** A clinically justified noninferiority margin defining the largest acceptable loss of efficacy.

**86.** A RhD-negative gravida has a rising anti-D titer and fetal middle cerebral artery Doppler shows increased peak systolic velocity. Which fetal complication is being screened for by this Doppler finding?

- A.** Fetal hyperviscosity caused by chronic maternal hypoxemia without hemolysis.
- B.** Fetal anemia from maternal IgG-mediated red-cell destruction.
- C.** Fetal polycythemia caused by placental transfusion from the maternal circulation.
- D.** Fetal thrombocytopenia caused by maternal antibodies against platelet antigens.

**87.** A 39-week newborn requires positive-pressure ventilation at birth. Which initial oxygen concentration is appropriate for routine resuscitation of a term infant?

- A.** Begin resuscitation with 100% oxygen in a ventilated term newborn and titrate down after the heart rate improves.
- B.** Start with 60% oxygen and maintain that fixed concentration for the first 10 minutes.
- C.** Avoid supplemental oxygen monitoring because pulse oximetry is unreliable during neonatal transition.
- D.** Start with room air and titrate oxygen according to preductal saturation and clinical response.

**88.** A newborn has a single loud second heart sound, a systolic ejection murmur, profound cyanosis, and echocardiography shows pulmonary atresia with an intact ventricular septum. Which immediate therapy is most important?

- A.** Begin digoxin alone because improved right-ventricular contractility will open the atretic valve.
- B.** Treat with diuretics as the primary intervention because pulmonary edema is the dominant cause of cyanosis in this lesion.
- C.** Close the ductus pharmacologically because pulmonary blood flow is excessive in this lesion.
- D.** Maintain ductal patency with prostaglandin E1 to provide pulmonary blood flow.

**89.** After delivery through heavily meconium-stained fluid, a post-term newborn requires escalating oxygen and develops an air-leak syndrome. Radiographs show uneven aeration with coarse bilateral opacities and hyperexpanded regions. Which diagnosis best explains the lung disease?

- A.** Primary pulmonary lymphangiectasia causing uniform bilateral pleural effusions at birth.
- B.** Meconium aspiration syndrome with airway obstruction and chemical pneumonitis.
- C.** Transient tachypnea caused solely by delayed fetal lung-fluid absorption after elective cesarean delivery.
- D.** Classic preterm respiratory distress syndrome with diffuse low-volume reticulogranular lungs.

**90.** A newborn has truncus arteriosus, hypocalcemic seizures, a small thymic shadow, and characteristic facial features. Which genetic lesion is most likely?

- A.** A 22q11.2 deletion affecting pharyngeal arch development.
- B.** A paternal 15q11-q13 deletion causing Prader-Willi syndrome.
- C.** An FGFR3 activating variant causing achondroplasia.
- D.** A SMN1 deletion causing spinal muscular atrophy.

**91.** A preterm infant on prolonged fat-free parenteral nutrition develops dermatitis, thrombocytopenia, poor growth, and abnormal essential fatty acid indices. Which deficiency best explains the syndrome?

- A.** Copper excess causing dermatitis and thrombocytopenia during lipid restriction.
- B.** Sodium deficiency causing a characteristic scaly rash and altered triene-to-tetraene ratio.
- C.** Essential fatty acid deficiency from inadequate lipid provision.
- D.** Vitamin B12 deficiency causing isolated macrocytic anemia without skin or fatty-acid abnormalities.

**92.** A newborn has abdominal distention, a poor urinary stream, a palpable bladder, bilateral hydronephrosis, and a thick-walled bladder on ultrasound. Which diagnosis is most likely?

- A.** Renal vein thrombosis, which causes renal enlargement and hematuria rather than a distended obstructed bladder.
- B.** Bilateral renal agenesis, which produces absent kidneys and an empty rather than distended bladder.
- C.** Autosomal recessive polycystic kidney disease, which causes enlarged echogenic kidneys without outlet obstruction.
- D.** Posterior urethral valves causing lower urinary tract obstruction in a male infant.

**93.** A newborn has seizures from hypocalcemia that improve only after a low serum magnesium level is corrected. Why can hypomagnesemia make hypocalcemia refractory?

- A.** Magnesium deficiency increases calcitonin breakdown and causes severe hypercalcemia.
- B.** Low magnesium directly binds ionized calcium in plasma and prevents renal excretion.
- C.** Magnesium deficiency causes vitamin K antagonism that lowers serum calcium.
- D.** Magnesium deficiency impairs parathyroid hormone secretion and action.

**94.** A newborn develops complete heart block and a photosensitive annular rash. The mother is asymptomatic but has anti-Ro/SSA antibodies. Which process is responsible?

- A.** Maternal IgM anti-Ro crossing the placenta and directly infecting cardiac conduction tissue.
- B.** Passive transplacental autoantibody-mediated neonatal lupus.
- C.** A congenital complement deficiency producing immune-complex deposition predominantly in the skin.
- D.** An intrinsic neonatal anti-Ro autoimmune response generated de novo during the first days of life.

**95.** A mother recalls a febrile rash illness during early pregnancy. Her newborn has a cloudy lens examination, failed hearing screen, and a continuous cardiac murmur. Which congenital infection best unifies these findings?

- A.** Neonatal HSV, which usually presents with vesicles, encephalitis, or disseminated hepatitis.
- B.** Congenital CMV, which more often causes periventricular calcifications and petechiae.
- C.** Congenital toxoplasmosis, which classically causes chorioretinitis rather than congenital cataracts.
- D.** Congenital rubella syndrome with cataracts, hearing loss, and patent ductus arteriosus.

**96.** A term infant with persistent abdominal distention has not stoolled normally since birth. Contrast enema demonstrates a narrow distal rectosigmoid segment with proximal colonic dilation. Which diagnosis is most likely?

- A.** Meconium ileus from cystic fibrosis, which typically shows inspissated ileal contents and microcolon.
- B.** Pyloric stenosis from hypertrophy of the pyloric muscle causing nonbilious emesis at several weeks.
- C.** Hirschsprung disease from congenital absence of enteric ganglion cells in the distal bowel.
- D.** Biliary atresia causing cholestasis and acholic stools rather than distal functional obstruction.

**97.** Parents of a former 25-week infant ask why early-intervention services were recommended before a definite developmental diagnosis was established. What is the best explanation?

- A.** Therapy should be postponed because infant neuroplasticity makes intervention ineffective during the first two years.
- B.** Delay developmental services until cerebral palsy or intellectual disability has been formally diagnosed.
- C.** Early-intervention programs are used mainly to assign a permanent disability label before developmental testing is possible.
- D.** Early therapy can support emerging motor, feeding, communication, and family needs while development is still rapidly changing.

**98.** A neonate repeatedly obstructs the upper airway when supine. Examination shows a markedly recessed mandible, posterior displacement of the tongue, and a broad U-shaped palatal cleft. Which developmental sequence is most likely?

- A.** Beckwith-Wiedemann syndrome, which causes macroglossia and overgrowth rather than micrognathia.
- B.** CHARGE syndrome, which is defined by coloboma, choanal atresia, ear anomalies, and other systemic findings rather than isolated micrognathia sequence.
- C.** Treacher Collins syndrome, which can cause micrognathia and cleft palate but does not explain the classic Pierre Robin sequence.
- D.** Pierre Robin sequence with micrognathia, glossoptosis, airway obstruction, and cleft palate.

**99.** In a randomized trial, several infants cross over from their assigned respiratory strategy to the alternative strategy. Which analysis best preserves the benefit of randomization for the primary comparison?

- A.** Per-protocol analysis that excludes crossovers and protocol deviations before estimating the treatment effect.
- B.** As-treated analysis that groups infants according to the therapy they ultimately received.
- C.** Intention-to-treat analysis based on the group to which each infant was originally randomized.
- D.** A case-control analysis matching infants after outcomes are known.

**100.** A mother with active Graves disease was treated during pregnancy. Her newborn is tachycardic, irritable, poorly gaining weight, and has a goiter despite the mother's own thyroid function being controlled. Which mechanism is most likely?

- A.** Maternal levothyroxine accumulates in fetal tissues and acts as a long-lived thyroid agonist after birth.
- B.** Placental deiodinase failure causes neonatal TSH resistance and secondary hyperthyroidism.
- C.** Transplacental thyroid-stimulating immunoglobulins are activating the infant's TSH receptor.
- D.** Maternal TSH crosses the placenta and directly stimulates the neonatal thyroid for several weeks.

**101.** A 27-week infant needs respiratory support in the delivery room. Which approach to initial oxygen is most appropriate?

- A.** Use a fixed 60% oxygen concentration initially and maintain it until a blood gas confirms adequate oxygenation.
- B.** Use 100% oxygen until the infant is intubated because extremely preterm infants cannot tolerate lower concentrations.
- C.** Use room air without pulse oximetry because supplemental oxygen is contraindicated before NICU admission.
- D.** Use a blended low oxygen concentration and titrate to preductal saturation targets rather than starting at 100%.

**102.** A cyanotic newborn has a harsh systolic murmur. During an episode of intense crying, oxygen saturation falls abruptly and the infant becomes deeply cyanotic. Which bedside maneuver can improve the physiology of a hypercyanotic spell in tetralogy of Fallot?

- A.** Lower systemic vascular resistance with a vasodilator to increase right-to-left shunting across the VSD.
- B.** Give indomethacin immediately to close the ductus and eliminate pulmonary steal.
- C.** Increase systemic vascular resistance with a knee-chest position while calming the infant.
- D.** Place the infant flat with legs extended to decrease venous return to the right ventricle.

**103.** A ventilated preterm infant suddenly becomes bradycardic and hypotensive with asymmetric chest movement and absent breath sounds on the right. Transillumination is bright on the right side. What is the immediate priority?

- A.** Give surfactant through the endotracheal tube because acute unilateral air loss indicates recurrent RDS.
- B.** Perform urgent needle decompression for a suspected tension pneumothorax before waiting for radiography.
- C.** Obtain a chest radiograph before any intervention because decompression is contraindicated without imaging.
- D.** Increase positive end-expiratory pressure to recruit the poorly ventilated right lung before further evaluation.

**104.** A newborn has hypertelorism, low-set ears, a webbed neck, pectus deformity, and valvar pulmonary stenosis. Which diagnosis is most likely?

- A.** Trisomy 18, which causes severe growth restriction and overlapping fingers.
- B.** Noonan syndrome, commonly associated with RAS-MAPK pathway variants.
- C.** Turner syndrome from sex-chromosome loss, which would better explain coarctation and neonatal lymphedema.
- D.** Williams syndrome, which is more associated with supravalvar aortic stenosis and hypercalcemia.

**105.** A former 24-week infant has rising alkaline phosphatase, low serum phosphorus, and radiographic demineralization. Which nutritional problem is most likely?

- A.** Metabolic bone disease of prematurity from inadequate calcium and phosphorus accretion.
- B.** Hypervitaminosis D causing excessive mineral deposition and increased bone density.
- C.** Iron deficiency causing osteopenia through reduced hemoglobin production.
- D.** Vitamin K deficiency causing reduced bone density as the primary neonatal manifestation.

**106.** An infant of a diabetic mother becomes dehydrated and then develops gross hematuria, thrombocytopenia, and an enlarged flank mass. Which diagnosis should be suspected?

- A.** Renal vein thrombosis causing renal enlargement, hematuria, and thrombocytopenia.
- B.** Prerenal azotemia causing a flank mass from renal enlargement and venous occlusion.
- C.** Minimal-change nephrotic syndrome causing hematuria and thrombocytopenia immediately after birth.
- D.** Posterior urethral valves causing isolated bladder outlet obstruction without thrombocytopenia.

**107.** A previously well newborn develops poor feeding, vomiting, lethargy, and marked hyperammonemia with respiratory alkalosis but no major anion-gap acidosis. Which metabolic category is most likely?

- A.** Galactosemia, which usually presents with liver dysfunction and E coli sepsis after milk exposure.
- B.** A urea-cycle disorder causing impaired ammonia detoxification.
- C.** An organic acidemia, which more typically produces a prominent high-anion-gap metabolic acidosis.
- D.** A fatty-acid oxidation defect, which classically causes hypoketotic hypoglycemia during fasting.

**108.** A clinically stable preterm infant reaches the chronological age for routine immunization while still hospitalized. Which principle generally applies to routine vaccines?

- A.** Postpone routine immunizations until 40 weeks postmenstrual age because immune responses in very preterm infants are insufficient before term.
- B.** Schedule immunizations by corrected age through 2 years to account for prematurity.
- C.** Avoid inactivated vaccines during NICU hospitalization because they can cause vaccine-strain infection in preterm infants.
- D.** Use chronological age for routine immunization rather than delaying solely because of prematurity.

**109.** A newborn develops persistent nasal discharge, a diffuse maculopapular rash involving the palms and soles, hepatosplenomegaly, and abnormal long-bone radiographs. Which infection is most likely?

- A.** Neonatal HSV limited to skin-eye-mouth disease without hepatosplenomegaly or bone changes.
- B.** Early-onset group B streptococcal sepsis, which does not cause snuffles and long-bone lesions.
- C.** Congenital CMV with isolated periventricular calcifications and no systemic findings.
- D.** Congenital syphilis with snuffles, palm-and-sole rash, hepatosplenomegaly, and long-bone disease.

**110.** A newborn has copious oral secretions, coughing and cyanosis with the first feeding, and an orogastric tube coils in the upper mediastinum. What is the most likely anomaly?

- A.** Hirschsprung disease, which affects the distal colon and does not cause excessive oral secretions at birth.
- B.** Duodenal atresia, which causes bilious vomiting and a double-bubble pattern rather than inability to pass an orogastric tube.
- C.** Esophageal atresia with a distal tracheoesophageal fistula causing proximal tube coiling and airway symptoms.
- D.** Pyloric stenosis, which develops later and allows normal passage of an orogastric tube into the stomach.

**111.** A septic newborn develops diffuse oozing, thrombocytopenia, prolonged PT and aPTT, low fibrinogen, and elevated D-dimer. Which process is most likely?

- A.** Disseminated intravascular coagulation with systemic consumption of platelets and coagulation factors.
- B.** Isolated hemophilia A causing factor VIII deficiency without thrombocytopenia or low fibrinogen.
- C.** Vitamin K deficiency alone causing prolonged clotting times without elevated fibrin degradation products.
- D.** Neonatal alloimmune thrombocytopenia causing profound platelet loss with otherwise normal coagulation studies.

**112.** A macrosomic infant has shoulder dystocia. After birth the right arm is adducted and internally rotated with the elbow extended, while hand grasp is preserved. Which nerve injury is most likely?

- A.** Upper brachial plexus injury involving C5-C6 roots, producing an Erb palsy pattern.
- B.** Radial nerve injury alone, which causes wrist drop without the characteristic shoulder posture.
- C.** Facial nerve palsy, which affects facial movement rather than the upper extremity.
- D.** Lower brachial plexus injury involving C8-T1, which primarily weakens the hand and may cause Horner syndrome.

**113.** Investigators identify infants with NEC and a comparison group without NEC, then look backward at prior feeding exposures. What study design is this?

- A.** An ecological study because individual infant exposure data are being compared.
- B.** A prospective randomized controlled trial because exposure is assessed after enrollment.
- C.** A case-control study that begins with outcome status and looks backward for prior exposures.
- D.** A cross-sectional prevalence study because outcomes and exposures were selected simultaneously at birth.

**114.** A term newborn exposed to a selective serotonin reuptake inhibitor throughout late pregnancy has tremulousness, increased tone, intermittent tachypnea, and feeding difficulty beginning several hours after birth. The infant is hemodynamically stable. What is the best explanation?

- A.** A transient poor neonatal adaptation syndrome related to late-gestation SSRI exposure.
- B.** A congenital serotonergic receptor deletion causing permanent neonatal hypertonia and tachypnea.
- C.** An acute bacterial sepsis syndrome that is specifically caused by maternal SSRI therapy.
- D.** A neonatal opioid withdrawal syndrome caused by cross-reactivity at mu-opioid receptors.

**115.** Mask ventilation is ineffective in a newborn despite repositioning, a good seal, airway opening, and corrective maneuvers. Intubation cannot be achieved promptly by the available provider. What is the most useful rescue airway strategy?

- A.** Begin chest compressions while withholding ventilation until a more experienced intubator arrives.
- B.** Place a supraglottic airway if the infant's size and gestation are appropriate for the device.
- C.** Perform an emergency surgical tracheostomy as the routine next airway step in the delivery room.
- D.** Continue the same ineffective face-mask technique without changing the airway interface.

**116.** A newborn with hypoplastic left heart syndrome is stable on prostaglandin. Which physiologic feature explains why excessive supplemental oxygen can be harmful before stage I palliation?

- A.** Oxygen causes immediate left-ventricular hypertrophy that obstructs the systemic outflow tract.
- B.** Oxygen administration rapidly closes the foramen ovale and removes atrial-level left-to-right mixing within minutes.
- C.** Pulmonary vasodilation can steal flow from the systemic circulation in a parallel circulation dependent on balanced vascular resistances.
- D.** Oxygen directly constricts the pulmonary arteries and sharply reduces pulmonary blood flow.

**117.** A 25-week infant has recurrent apnea with bradycardia and desaturation after infection, anemia, glucose disturbance, and airway obstruction have been excluded. Which medication is first-line for apnea of prematurity?

- A.** Morphine sulfate to reduce respiratory variability and prevent recurrent central pauses.
- B.** Propranolol to blunt sympathetic surges associated with desaturation events.
- C.** Acetazolamide to induce metabolic acidosis as the preferred chronic respiratory stimulant.
- D.** Caffeine citrate to stimulate respiratory drive and reduce clinically significant apnea.

**118.** A newborn is large for gestational age with macroglossia, an omphalocele, ear creases, and recurrent hypoglycemia. Which syndrome is most likely?

- A.** Beckwith-Wiedemann syndrome with fetal overgrowth and hyperinsulinism risk.
- B.** Turner syndrome with lymphedema and coarctation rather than macroglossia and omphalocele.
- C.** Trisomy 18 with growth restriction and clenched hands rather than macrosomia.
- D.** Prader-Willi syndrome with severe neonatal hypotonia and poor suck but not somatic overgrowth.

**119.** A 30-week infant is receiving fortified human milk and has stable growth. Which micronutrient commonly requires supplementation because preterm infants have limited stores and rapid growth?

- A.** Iron to reduce the risk of iron-deficiency anemia after early neonatal stores are depleted.
- B.** Folate avoidance because supplementation suppresses neonatal erythropoiesis.
- C.** Give pharmacologic adult-dose vitamin A to prevent chronic lung disease in a growing preterm infant.
- D.** Copper restriction because preterm infants routinely accumulate toxic copper from human milk.

**120.** A very preterm infant receiving prolonged furosemide develops increased renal medullary echogenicity on ultrasound. Which complication is most likely?

- A.** Uric-acid nephropathy caused by furosemide-induced tumor lysis in the absence of malignancy.
- B.** Nephrocalcinosis related to hypercalciuria from loop-diuretic exposure and prematurity.
- C.** Posterior urethral valves caused by postnatal loop-diuretic exposure.
- D.** Renal cortical necrosis caused specifically by increased urinary calcium excretion.

**121.** A newborn develops poor feeding, abnormal tone, intermittent apnea, and urine with a sweet maple-syrup odor. Which disorder is most likely?

- A.** Maple syrup urine disease from deficient branched-chain alpha-ketoacid dehydrogenase activity.
- B.** Medium-chain acyl-CoA dehydrogenase deficiency, which causes hypoketotic hypoglycemia during fasting.
- C.** Classic galactosemia, which causes liver dysfunction and sepsis after galactose exposure.
- D.** Phenylketonuria, which is usually asymptomatic in the immediate newborn period and lacks the characteristic odor.

**122.** A preterm infant is born with diffuse granulomatous skin lesions and rapidly progressive sepsis after the mother had a febrile gastrointestinal illness. Which organism is most likely?

- A.** *Clostridium botulinum* causing descending paralysis after ingestion of spores.
- B.** *Listeria monocytogenes* causing granulomatosis infantiseptica.
- C.** Coagulase-negative *Staphylococcus* causing a catheter-associated late-onset bloodstream infection.
- D.** *Bordetella pertussis* causing neonatal apnea without congenital granulomatous lesions.

**123.** A newborn has bowel loops protruding through a right-sided paraumbilical abdominal wall defect without a covering membrane. What is the diagnosis?

- A.** Gastroschisis with uncovered bowel protruding through a right paraumbilical abdominal-wall defect.
- B.** Omphalocele, in which abdominal contents herniate through the umbilical ring within a protective sac.
- C.** Umbilical hernia, which is skin-covered and usually contains a small reducible portion of bowel.
- D.** Bladder exstrophy, which exposes bladder mucosa rather than free bowel loops.

**124.** A male newborn with a family history of hemophilia develops an intracranial hemorrhage after a difficult delivery. Platelet count and PT are normal, but aPTT is prolonged. Which diagnosis is most likely?

- A.** Neonatal alloimmune thrombocytopenia, which causes low platelet count with normal clotting-factor tests.
- B.** Disseminated intravascular coagulation, which usually prolongs both PT and aPTT and consumes platelets.
- C.** Hemophilia A or B causing deficiency of an intrinsic-pathway clotting factor.
- D.** Vitamin K deficiency, which typically prolongs PT early because factor VII falls rapidly.

**125.** After forceps-assisted delivery, a newborn cannot fully close the left eye and the left corner of the mouth does not move with crying, but forehead movement is also reduced. What is the most likely diagnosis?

- A.** Congenital myotonic dystrophy, which causes generalized weakness rather than isolated unilateral facial palsy.
- B.** A unilateral upper motor neuron lesion, which typically preserves forehead movement because of bilateral cortical innervation.
- C.** Peripheral facial nerve palsy from birth trauma affecting the entire ipsilateral face.
- D.** Brachial plexus palsy, which affects the upper limb rather than facial movement.

**126.** During positive-pressure ventilation, the newborn's heart rate is not rising and there is minimal chest movement. Which action should precede escalation to chest compressions?

- A.** Begin compressions because visible chest movement is not necessary for effective neonatal ventilation.
- B.** Correct ventilation technique and confirm effective lung inflation before judging ventilation a failure.
- C.** Give epinephrine through the endotracheal tube before addressing mask seal or airway position.
- D.** Lower inspiratory pressure further because any visible chest rise indicates excessive tidal volume.

**127.** A newborn has severe respiratory distress, cyanosis, pulmonary edema on chest radiograph, and echocardiography shows pulmonary veins draining to a vertical vein with obstruction. What is the definitive management?

- A.** Indomethacin to close the ductus, which is the primary source of pulmonary venous hypertension.
- B.** Balloon pulmonary valvuloplasty because valvar pulmonary stenosis causes the venous obstruction.
- C.** Urgent surgical repair of obstructed total anomalous pulmonary venous return.
- D.** Prolonged diuretic therapy alone because obstructed pulmonary venous return usually resolves as pulmonary resistance falls.

**128.** A newborn with a prenatal diagnosis of left congenital diaphragmatic hernia is delivered with severe respiratory distress and a scaphoid abdomen. Which initial respiratory maneuver should be avoided?

- A.** Intubate for controlled ventilation when respiratory support is required immediately after birth.
- B.** Place an orogastric tube for continuous gastric decompression while stabilizing ventilation.
- C.** Use gentle ventilatory pressures to reduce additional lung injury in the hypoplastic lungs.
- D.** Avoid routine bag-mask ventilation and proceed to controlled airway support with gastric decompression.

**129.** A newborn has severe hypotonia and poor feeding. The hands and feet are small, and genetic testing later identifies loss of paternally expressed genes in 15q11-q13. Which diagnosis is this?

- A.** Angelman syndrome from loss of maternally expressed UBE3A function, usually recognized later by developmental phenotype.
- B.** Prader-Willi syndrome with neonatal hypotonia and feeding difficulty from loss of paternal 15q11-q13 expression.
- C.** Spinal muscular atrophy from biallelic SMN1 loss with anterior-horn-cell disease.
- D.** Congenital myotonic dystrophy from a maternally transmitted DMPK repeat expansion.

**130.** A VLBW infant is receiving exclusive human milk and has reached full enteral feeds. Which additional nutrient is commonly supplemented to support bone health and calcium metabolism?

- A.** Vitamin K as the sole ongoing strategy to prevent metabolic bone disease of prematurity.
- B.** Thiamine in pharmacologic doses because breast milk contains no bioavailable thiamine.
- C.** Vitamin D in an age-appropriate neonatal dose to support calcium and phosphorus homeostasis.
- D.** Vitamin C in megadose therapy to replace calcium fortification of human milk.

**131.** A newborn with severe birth asphyxia develops oliguria and then, several days later, a brisk polyuric phase with electrolyte losses. Which renal course does this pattern suggest?

- A.** Isolated prerenal azotemia because intact tubules cause prolonged salt-wasting polyuria during recovery.
- B.** Recovery from intrinsic acute kidney injury with a diuretic phase before tubular concentrating function normalizes.
- C.** Progressive bilateral renal agenesis because urine output typically increases before complete renal failure.
- D.** Syndrome of inappropriate antidiuretic hormone secretion because SIADH produces sustained polyuria.

**132.** A newborn becomes ill after a brief fast with hypoglycemia, low or absent ketones, and elevated liver enzymes. Which metabolic process should be suspected?

- A.** A fatty-acid oxidation defect that prevents generation of ketones during fasting.
- B.** A urea-cycle defect, which primarily causes hyperammonemia without hypoketotic hypoglycemia.
- C.** Congenital hypothyroidism, which causes persistent low thyroid hormone rather than fasting crises.
- D.** 21-hydroxylase deficiency, which causes salt wasting and hyperkalemia rather than absent ketones.

**133.** A 700-g infant has received several antibiotic courses and remains dependent on a central line. New apnea, thrombocytopenia, and worsening glucose control occur, and blood cultures continue to grow a yeast. Which organism group is the leading concern?

- A.** *Toxoplasma gondii* acquired from the central catheter during parenteral nutrition.
- B.** Respiratory syncytial virus causing isolated candidemia-like bloodstream findings.
- C.** *Candida* species causing invasive late-onset fungal infection.
- D.** Group B *Streptococcus* as the predominant pathogen after the first week in an ELBW infant with a long-term central line.

**134.** A newborn has a midline abdominal wall defect at the umbilical ring. Liver and bowel are contained within a translucent sac. Which diagnosis is most likely?

- A.** Necrotizing enterocolitis, which is an acquired intestinal inflammatory disease after birth.
- B.** Gastroschisis, which is typically right paraumbilical and lacks a covering sac.
- C.** Omphalocele, which is strongly associated with other congenital and chromosomal abnormalities.
- D.** Patent urachus, which causes urinary drainage from the umbilicus rather than herniated liver and bowel.

**135.** A jaundiced newborn has anemia, reticulocytosis, and episodic hemolysis after oxidant exposure. Peripheral smear shows bite cells. Which red-cell disorder is most likely?

- A.** Glucose-6-phosphate dehydrogenase deficiency causing oxidant-triggered neonatal hemolysis.
- B.** Hereditary spherocytosis, which produces spherocytes and increased mean corpuscular hemoglobin concentration rather than oxidant-induced bite cells.
- C.** Iron deficiency, which produces microcytosis rather than acute neonatal hemolysis.
- D.** Anemia of prematurity, which is hyporegenerative and develops later in very preterm infants.

**136.** A lethargic term infant with persistent hypoglycemia later has MRI injury predominantly in the posterior cerebral cortex and occipital regions. Which complication should be anticipated?

- A.** Spastic diplegia from classic periventricular leukomalacia in a term infant.
- B.** Congenital cataracts caused directly by low serum glucose during the first day.
- C.** Isolated hearing loss because neonatal hypoglycemia selectively injures the auditory nerve.
- D.** Visual processing impairment related to hypoglycemic occipital brain injury.

**137.** A cohort study finds an association between prolonged ventilation and neurodevelopmental impairment. Infants who required prolonged ventilation were also much more premature. What epidemiologic problem is illustrated?

- A.** Spectrum bias from studying a diagnostic test in a restricted range of disease severity.
- B.** Randomization, because gestational age has been equally distributed by chance between exposure groups.
- C.** Confounding by gestational age because it is associated with both exposure and outcome.
- D.** Blinding, because clinicians cannot see the infants' gestational ages during follow-up.

**138.** A newborn is delivered after chronic maternal fentanyl use and begins to show high-pitched crying, tremors, loose stools, and poor sleep at 36 hours of age. Which diagnosis best integrates these findings?

- A.** Transient tachypnea of the newborn caused by delayed clearance of fetal lung fluid.
- B.** Neonatal opioid withdrawal syndrome after chronic in-utero opioid exposure.
- C.** Neonatal serotonin syndrome from fetal opioid metabolism to serotonergic compounds.
- D.** Early-onset hypocalcemia causing an isolated neuromuscular syndrome without autonomic signs.

**139.** A newborn has vertebral segmentation defects, an imperforate anus, a ventricular septal defect, a tracheoesophageal fistula, renal anomalies, and a radial-ray defect. Which pattern is being described?

- A.** VACTERL association involving vertebral, anorectal, cardiac, tracheoesophageal, renal, and limb anomalies.
- B.** Pierre Robin sequence, which consists primarily of micrognathia, glossoptosis, and airway obstruction.
- C.** Beckwith-Wiedemann syndrome, which causes overgrowth, macroglossia, and abdominal wall defects.
- D.** CHARGE syndrome, which centers on coloboma, choanal atresia, cranial-nerve dysfunction, and ear anomalies.

**140.** A 29-week infant has been advancing feeds without abdominal signs. Gastric residual volumes vary from feed to feed, but the abdomen is soft and stooling is normal. What is the best feeding principle?

- A.** Advance feeds solely according to residual volume without considering abdominal examination or clinical status.
- B.** Stop feeds whenever any measurable residual is present because residual volume reliably predicts NEC.
- C.** Discard measured gastric residuals and replace the volume with sterile water to prevent electrolyte imbalance.
- D.** Do not use isolated gastric residual volume as the sole reason to stop otherwise well-tolerated feeds.

**141.** A newborn with central nervous system injury has hyponatremia, low serum osmolality, inappropriately concentrated urine, and no evidence of volume depletion. What is the most likely diagnosis?

- A.** Diabetes insipidus causing dilute urine, hypernatremia, and free-water loss.
- B.** Renal salt wasting causing volume depletion with high urinary sodium and compensatory ADH secretion.
- C.** Hypernatremic dehydration caused by insufficient intake and maximally concentrated serum sodium.
- D.** Syndrome of inappropriate antidiuretic hormone secretion.

**142.** After lactose-containing feeds are introduced, a newborn develops liver dysfunction, poor feeding, and gram-negative sepsis; urine contains reducing substances despite a negative glucose dipstick. Which inborn error should be suspected?

- A.** Maple syrup urine disease, which causes encephalopathy and branched-chain amino-acid accumulation without liver failure.
- B.** Biotinidase deficiency, which more often presents later with seizures, dermatitis, and alopecia.
- C.** Classic galactosemia from galactose-1-phosphate uridylyltransferase deficiency.
- D.** Phenylketonuria, which does not typically cause acute liver failure or E coli sepsis in the neonatal period.

**143.** A 3-week-old previously healthy infant develops fever, irritability, and meningitis. Blood and CSF grow group B Streptococcus. Which epidemiologic category is this?

- A.** A vaccine-preventable infection caused by failure of the infant's routine GBS immunization.
- B.** Late-onset group B streptococcal disease, which can present with bacteremia or meningitis after the first week.
- C.** Early-onset group B streptococcal disease from intra-amniotic exposure despite presentation at 3 weeks.
- D.** Congenital infection acquired transplacentally months before birth with calcifications.

**144.** A newborn screen identifies sickle cell disease in an asymptomatic term infant. Why are severe vaso-occlusive symptoms usually absent at birth?

- A.** Fetal hemoglobin remains the predominant hemoglobin and inhibits sickling during the early months.
- B.** The neonatal spleen has not yet formed, so sickled erythrocytes cannot be trapped in the circulation.
- C.** Maternal hemoglobin S crosses the placenta and temporarily replaces the infant's abnormal hemoglobin.
- D.** Sickle hemoglobin is not produced until after the first birthday because beta-globin genes are inactive in infancy.

**145.** A term infant with acute profound asphyxia has severe encephalopathy. MRI later shows injury to the basal ganglia, thalami, and perirolandic cortex. Which insult pattern is most consistent with this distribution?

- A.** Congenital CMV, which characteristically causes periventricular calcifications rather than this acute diffusion pattern.
- B.** An acute profound hypoxic-ischemic event with injury to high-metabolic-demand deep gray structures.
- C.** Periventricular leukomalacia of extreme prematurity, which primarily affects immature periventricular white matter.
- D.** A mild chronic placental insufficiency pattern limited to distal watershed white matter without deep-gray injury.

**146.** A trial reports a relative risk of 0.82 with a 95% confidence interval of 0.61 to 1.10 for mortality. Which interpretation is most appropriate?

- A.** The data are compatible with benefit or no benefit, and the confidence interval includes the null value of 1.0.
- B.** The result proves equivalence because the confidence interval crosses 1.0.
- C.** The treatment definitely lowers mortality because the point estimate is below 1.0.
- D.** The treatment definitely increases mortality because the upper confidence limit is above 1.0.

**147.** A mother develops intrapartum fever, uterine tenderness, and foul-smelling amniotic fluid after prolonged rupture of membranes. Which neonatal consequence is most directly increased by this maternal condition?

- A.** Late-onset coagulase-negative staphylococcal sepsis acquired from a central venous catheter.
- B.** Congenital cytomegalovirus infection acquired primarily through transplacental viral spread.
- C.** Neonatal tetanus caused by impaired transfer of maternal antitoxin across the placenta.
- D.** Early-onset neonatal sepsis from ascending intra-amniotic infection.

**148.** A 6-week-old former term infant has poor weight gain, tachypnea with feeds, hepatomegaly, and a holosystolic murmur at the lower left sternal border. Why did symptoms appear after several initially well weeks?

- A.** Falling pulmonary vascular resistance increased the left-to-right shunt across a large ventricular septal defect.
- B.** The ductus reopened spontaneously and redirected systemic venous blood into the left ventricle.
- C.** Pulmonary vascular resistance rose after birth, increasing pulmonary overcirculation through the defect.
- D.** Progressive closure of the foramen ovale created a new right-to-left shunt across the ventricular septum.

**149.** A term infant with pneumonia has labile hypoxemia, a loud second heart sound, and echocardiographic evidence of right-to-left shunting without structural heart disease. What is the most likely diagnosis?

- A.** Bronchopulmonary dysplasia caused by prolonged ventilator exposure in a term infant.
- B.** Isolated transient tachypnea of the newborn without elevation of pulmonary vascular resistance.
- C.** A large ventricular septal defect causing immediate severe left-to-right shunting on the first day.
- D.** Persistent pulmonary hypertension of the newborn with elevated pulmonary vascular resistance and right-to-left shunting.

**150.** Within 48 hours of protein feeding, a neonate becomes encephalopathic. Laboratory testing shows a marked anion-gap acidosis with ketonuria, elevated ammonia, and neutropenia rather than a primary respiratory disorder. Which class of metabolic disease is most likely?

- A.** A pure urea-cycle disorder, which usually causes respiratory alkalosis and hyperammonemia without marked ketosis.
- B.** Congenital hypothyroidism, which does not produce acute severe anion-gap acidosis.
- C.** Isolated growth hormone deficiency, which causes hypoglycemia without organic acid accumulation.
- D.** An organic acidemia such as propionic or methylmalonic acidemia.

**151.** A 28-week infant with a peripherally inserted central catheter develops temperature instability and apnea on day 24. Blood cultures repeatedly grow coagulase-negative *Staphylococcus*. Why is this organism important in the NICU?

- A.** It produces a characteristic neonatal exanthem that distinguishes contamination from infection.
- B.** It is uniformly a blood-culture contaminant and should not be treated even when multiple cultures are positive.
- C.** It is a common cause of late-onset catheter-associated bloodstream infection in very preterm infants.
- D.** It is the dominant transplacental cause of congenital microcephaly and intracranial calcifications.

**152.** A newborn with trisomy 21 has leukocytosis with circulating blasts that resemble acute megakaryoblastic leukemia but then decline spontaneously over several weeks. What is the most likely diagnosis?

- A.** Physiologic neonatal leukocytosis, which does not produce a clonal megakaryoblastic population.
- B.** Congenital neutropenia from maternal hypertension, which causes low neutrophil counts rather than blasts.
- C.** Transient abnormal myelopoiesis associated with Down syndrome.
- D.** Juvenile myelomonocytic leukemia, which does not typically resolve spontaneously in the newborn period.

**153.** A newborn with suspected encephalopathy has an amplitude-integrated EEG showing a severely depressed background with recurrent seizures. What is the main value of this information?

- A.** It establishes the precise metabolic cause of encephalopathy without laboratory testing or imaging.
- B.** It helps characterize background function and seizure burden but should be integrated with examination and conventional EEG when available.
- C.** It excludes seizures whenever no overt motor activity is seen at the bedside.
- D.** It replaces the neurologic examination because background patterns are unaffected by medication or sleep state.

**154.** A NICU team tests a new handoff checklist on one small unit, reviews defects after one week, modifies the checklist, and repeats the cycle. Which quality-improvement method is being used?

- A.** A case-control study in which staff are selected according to whether a handoff error occurred.
- B.** A Plan-Do-Study-Act cycle with iterative small tests of change.
- C.** A meta-analysis combining published studies without making local process changes.
- D.** A fixed parallel-group efficacy trial that prohibits modification of the intervention after enrollment.

**155.** A stable vigorous infant is born at 30 weeks after an uncomplicated preterm delivery. Which umbilical-cord approach is favored when no immediate resuscitative intervention prevents it?

- A.** Leave the cord unclamped for 10 minutes to maximize placental transfusion before any assessment.
- B.** Clamp the cord immediately because a 30-week infant is likely to need early vascular access.
- C.** Use intact-cord milking repeatedly in this 30-week infant as the preferred alternative to delayed cord clamping.
- D.** Defer cord clamping for about 60 seconds while maintaining thermal support.

**156.** A newborn with maternal anti-Ro/SSA antibodies has a persistent ventricular rate of 55/min with atrial activity dissociated from ventricular activity. Which diagnosis is most likely?

- A.** Sinus bradycardia caused by neonatal hypothermia with intact atrioventricular conduction.
- B.** Supraventricular tachycardia with 2:1 conduction producing an apparently slow ventricular rate.
- C.** Patent ductus arteriosus with reflex bradycardia from wide pulse pressure.
- D.** Congenital complete atrioventricular block due to antibody-mediated injury of the conduction system.

**157.** A 24-week infant with severe RDS develops bloody endotracheal secretions, worsening oxygenation, and diffuse new alveolar opacities. What complication should be suspected?

- A.** Congenital lobar emphysema, which usually causes focal hyperinflation rather than diffuse bloody secretions.
- B.** Isolated apnea of prematurity, which does not produce new diffuse alveolar opacities or blood in the airway.
- C.** Pulmonary hemorrhage, often associated with severe illness and hemodynamically significant shunting.
- D.** Transient tachypnea, which typically presents with mild distress and fissural fluid after term cesarean birth.

**158.** A newborn has a coloboma, choanal atresia, cranial-nerve dysfunction causing feeding difficulty, genital hypoplasia, and abnormal external ears. Which syndrome is most likely?

- A.** Trisomy 21, which commonly includes hypotonia and atrioventricular septal defect rather than choanal atresia.
- B.** VACTERL association, which is defined by vertebral, anorectal, cardiac, tracheoesophageal, renal, and limb anomalies.
- C.** CHARGE syndrome, frequently related to CHD7 pathogenic variants.
- D.** Noonan syndrome, which commonly includes pulmonic stenosis and webbed neck without coloboma.

**159.** An 850-g infant is receiving human milk. The team asks whether a commercial probiotic should be given routinely to prevent NEC. Which statement best reflects AAP caution?

- A.** Routine universal probiotic administration to all preterm infants is not endorsed because product quality, strain, and safety concerns remain.
- B.** Commercial probiotic products have drug-level regulation and equivalent strain-specific efficacy across VLBW populations.
- C.** Probiotics are required whenever donor human milk is used because pasteurization makes NEC otherwise unavoidable.
- D.** Probiotics replace the need for human milk because they provide equivalent protection against NEC.

**160.** Prenatal imaging shows bilateral renal agenesis and severe oligohydramnios. Which neonatal complication is most likely to determine immediate survival?

- A.** Severe pulmonary hypoplasia from prolonged oligohydramnios.
- B.** Neonatal polyuria caused by compensatory overfunction of absent renal tissue.
- C.** Isolated direct hyperbilirubinemia caused by loss of renal bilirubin excretion.
- D.** Hyperexpanded lungs from excessive fetal lung fluid retention despite absent kidneys.

**161.** A newborn has marked hypotonia, weak antigravity movement, absent reflexes, and preserved alertness with normal eye movements. Which localization is most likely?

- A.** A unilateral hemispheric stroke, which would be expected to produce asymmetric rather than symmetric weakness.
- B.** A diffuse cortical encephalopathy, which typically impairs alertness and may produce abnormal central reflex patterns.
- C.** An isolated cerebellar lesion, which does not usually cause neonatal areflexia and profound weakness.
- D.** A peripheral neuromuscular disorder affecting motor unit function.

**162.** Monochorionic diamniotic twins are delivered at 30 weeks after ultrasound showed oligohydramnios in one sac and polyhydramnios in the other. The larger twin is plethoric and hypertensive. Which complication is most likely in this recipient twin?

- A.** Severe anemia from chronic net loss of blood to the smaller donor twin.
- B.** Polycythemia and volume overload from chronic unbalanced placental transfusion.
- C.** Maternal platelet alloantibodies causing thrombocytopenia confined to the recipient twin.
- D.** Congenital nephrotic syndrome caused by placental vascular anastomoses.

**163.** A term infant develops a regular narrow-complex tachycardia at 260/min with poor feeding but preserved blood pressure. Vagal maneuvers fail and there is no evidence of pre-excitation during the episode. What is the next acute treatment?

- A.** Give indomethacin because a PDA is the most common cause of a regular narrow-complex tachycardia.
- B.** Give rapid intravenous adenosine while continuous rhythm monitoring is in place.
- C.** Start oral digoxin and wait several hours before attempting acute rhythm termination.
- D.** Give intravenous atropine to slow atrioventricular nodal conduction and terminate the tachycardia.

**164.** A ventilated extremely preterm infant develops multiple small cystic radiolucencies tracking along bronchovascular bundles. Which air-leak syndrome is most likely?

- A.** Transient tachypnea producing interstitial fluid rather than air-filled linear and cystic lucencies.
- B.** Pneumomediastinum limited to the central mediastinal space without intrapulmonary air dissection.
- C.** Congenital pulmonary airway malformation caused by prenatal cyst formation rather than ventilation injury.
- D.** Pulmonary interstitial emphysema from air dissecting into the pulmonary interstitium.

**165.** A term infant has rhizomelic shortening of the limbs, frontal bossing, midface hypoplasia, and a relatively long trunk. Which molecular mechanism is most likely?

- A.** A COL1A1 loss-of-function variant causing brittle bones and recurrent fractures at birth.
- B.** An FBN1 variant causing long limbs and neonatal connective-tissue laxity.
- C.** A dystrophin deletion causing congenital muscle hypertrophy with normal skeletal proportions.
- D.** An activating FGFR3 variant causing achondroplasia.

**166.** A premature infant on prolonged parenteral nutrition develops progressive direct hyperbilirubinemia after enteral feeds were repeatedly interrupted. Which nutritional strategy may help reduce parenteral-nutrition-associated cholestasis?

- A.** Increase intravenous dextrose substantially because high glucose infusion prevents hepatic fat deposition.
- B.** Discontinue intravenous lipid indefinitely because lipid avoidance improves cholestasis without causing essential fatty-acid deficiency.
- C.** Advance enteral nutrition as tolerated and minimize unnecessary duration of parenteral nutrition.
- D.** Stop enteral feeds because luminal nutrients worsen bile flow in parenteral-nutrition-associated cholestasis.

**167.** A newborn is delivered to a mother who is HBsAg positive. Which immediate prophylaxis is indicated for the infant?

- A.** Use oral antiviral therapy alone and avoid passive or active immunization during the newborn period.
- B.** Give hepatitis B vaccine and hepatitis B immune globulin promptly after birth at separate sites.
- C.** Postpone hepatitis B vaccine until 2 months because maternal antibody substantially reduces neonatal vaccine response.
- D.** Give hepatitis B immune globulin without vaccine because passive antibody is sufficient after perinatal exposure.

**168.** A term infant has rhythmic jerking that cannot be stopped by holding the limb and is accompanied by eye deviation. How does this differ from benign neonatal jitteriness?

- A.** Seizures are not consistently stimulus-sensitive or suppressible and may have ocular or autonomic features.
- B.** Jitteriness is usually associated with eye deviation and persists unchanged when the limb is gently restrained.
- C.** Classify the rhythmic jerking as a benign sleep phenomenon because sleep-related events stop with arousal.
- D.** Treat jitteriness as a marker of cortical injury that generally requires antiseizure medication.

**169.** A patient with preterm prelabor rupture of membranes at 30 weeks has no fever, uterine tenderness, fetal compromise, or active labor. Which maternal strategy is used to prolong latency and reduce infectious morbidity while expectant management continues?

- A.** Administer an evidence-based latency antibiotic regimen while monitoring for infection and labor.
- B.** Perform serial digital cervical examinations to identify the earliest sign of labor progression.
- C.** Avoid antenatal corticosteroids because membrane rupture prevents fetal pulmonary benefit.
- D.** Give prolonged broad-spectrum antifungal therapy because *Candida* is the usual ascending pathogen.

**170.** A newborn has tricuspid valve displacement toward the right-ventricular apex, marked right atrial enlargement, and severe tricuspid regurgitation. Which congenital lesion is described?

- A.** Critical pulmonary stenosis with a normally inserted tricuspid valve.
- B.** Ebstein anomaly of the tricuspid valve with apical leaflet displacement and severe regurgitation.
- C.** Complete atrioventricular septal defect with a common atrioventricular junction.
- D.** Tetralogy of Fallot with anterior malalignment of the ventricular septum.

**171.** A former 25-week infant remains oxygen-dependent with chronic radiographic changes at 36 weeks postmenstrual age. Which diagnosis best describes this chronic respiratory morbidity of prematurity?

- A.** Transient tachypnea of the newborn, which should resolve within the first few postnatal days.
- B.** Primary ciliary dyskinesia, defined in preterm infants by ongoing oxygen need at 36 weeks postmenstrual age.
- C.** Bronchopulmonary dysplasia, assessed by respiratory support requirements around 36 weeks postmenstrual age.
- D.** Meconium aspiration syndrome, which requires post-term birth and meconium-stained amniotic fluid.

**172.** A newborn has multiple fractures of different ages, blue sclerae, and generalized osteopenia without evidence of trauma. Which diagnosis is most likely?

- A.** Hypophosphatasia caused solely by vitamin D deficiency after birth.
- B.** Congenital muscular dystrophy causing fractures from spasticity rather than brittle bone.
- C.** Achondroplasia due to FGFR3 activation with short limbs but normal bone fragility.
- D.** Osteogenesis imperfecta due to an abnormality of type I collagen.

**173.** A preterm infant gains weight rapidly but length and head circumference growth are poor and serum urea nitrogen is very low. Which nutritional issue should be considered?

- A.** Protein intake may be inadequate relative to total energy intake, limiting lean tissue growth.
- B.** Iron excess is the primary cause of poor length growth when weight gain is preserved.
- C.** Excess protein intake is the most likely cause because low urea nitrogen indicates nitrogen overload.
- D.** Too much calcium intake selectively suppresses linear and head growth despite adequate protein.

**174.** A preterm infant with necrotizing enterocolitis becomes more unstable despite bowel rest and broad-spectrum antibacterial therapy. Blood cultures remain positive for a yeast. What additional management principle is important?

- A.** Continue antibacterial therapy alone because Candida in neonatal blood cultures represents colonization.
- B.** Stop antimicrobial therapy because persistent yeast recovery is an expected inflammatory finding during necrotizing enterocolitis.
- C.** Use topical nystatin alone because systemic antifungal agents do not reach neonatal bloodstream infection.
- D.** Treat invasive candidiasis with systemic antifungal therapy and evaluate for disseminated disease and infected intravascular devices.

**175.** A preterm infant with a PDA has bounding pulses, a hyperdynamic precordium, and a widening pulse pressure. Which hemodynamic process best explains these findings?

- A.** Runoff from the systemic circulation into the pulmonary artery lowers diastolic pressure while increasing pulmonary flow.
- B.** Pulmonary venous obstruction causes isolated systolic hypertension without altered diastolic runoff.
- C.** A right-to-left ductal shunt increases systemic diastolic pressure by returning blood to the aorta.
- D.** Fixed obstruction of left-ventricular outflow raises diastolic pressure and reduces pulse amplitude.

**176.** A 23-week infant remains intubated on substantial ventilator support at 18 days of age and has a high predicted risk of severe bronchopulmonary dysplasia. Which corticosteroid strategy is consistent with AAP guidance?

- A.** Consider an individualized low-dose systemic dexamethasone course after discussing potential benefits and neurodevelopmental risks with the family.
- B.** Avoid systemic corticosteroids in this infant because respiratory benefit does not justify neurodevelopmental risk in ventilator-dependent prematurity.
- C.** Use a routine high-dose dexamethasone course because higher doses provide the best established long-term neurodevelopmental safety.
- D.** Start prolonged inhaled corticosteroids as standard therapy because they clearly reduce mortality compared with systemic treatment.

**177.** A newborn has profound symmetric weakness, absent deep tendon reflexes, tongue fasciculations, and preserved alertness. Newborn screening detects biallelic loss of SMN1. Which disorder is present?

- A.** Prader-Willi syndrome, which causes hypotonia but not tongue fasciculations or areflexia from motor-neuron loss.
- B.** Hypoxic-ischemic encephalopathy, which typically causes altered consciousness and central neurologic abnormalities.
- C.** Congenital hypothyroidism, which can cause hypotonia but not a pure lower-motor-neuron syndrome.
- D.** Spinal muscular atrophy from degeneration of anterior horn cells.

**178.** A 34-week infant has immature suck-swallow-breathe coordination and recurrent desaturation during bottle feeds but otherwise stable respiratory status. What is the best nutritional approach?

- A.** Withhold enteral feeding until coordinated oral feeding develops because gavage feeding delays suck-swallow-breathe maturation.
- B.** Use paced or gavage-assisted feeding while oral skills mature, guided by feeding readiness and safety.
- C.** Thicken bottle feeds empirically before assessing feeding mechanics because aspiration is the leading explanation for these desaturations.
- D.** Force full oral feeds to accelerate maturation because desaturation is expected and harmless.

**179.** A neonate with meningitis develops progressive ventriculomegaly and a rapidly enlarging head circumference. Which complication should be suspected?

- A.** Neonatal abstinence syndrome, which causes tremor and irritability but not progressive ventriculomegaly.
- B.** Postinfectious hydrocephalus from impaired cerebrospinal-fluid flow or resorption.
- C.** Benign enlargement of the subarachnoid spaces, which does not follow acute bacterial meningitis with rapid progression.
- D.** Microcephaly from premature suture fusion causing enlarged ventricles.

**180.** A term infant with structurally normal heart anatomy has labile severe hypoxemia after meconium aspiration. Echocardiography shows suprasystemic pulmonary artery pressure and right-to-left flow through the ductus arteriosus. Which physiology is causing the hypoxemia?

- A.** Right-to-left ductal shunting from persistent pulmonary hypertension of the newborn.
- B.** A large isolated atrial septal defect producing obligatory left-to-right flow at the atrial level.
- C.** A ventricular septal defect causing pulmonary overcirculation with equal preductal and postductal oxygenation.
- D.** An isolated patent foramen ovale with no elevation in pulmonary vascular resistance.

**181.** A 26-week infant is ventilated for severe RDS. The team wants to reduce ventilator-associated lung injury. Which general strategy is most appropriate?

- A.** Eliminate positive end-expiratory pressure because maintaining functional residual capacity promotes volutrauma.
- B.** Use the lowest effective tidal volumes and pressures while accepting modest permissive hypercapnia when clinically appropriate.
- C.** Maintain severe hypocapnia because low carbon dioxide reduces the risk of chronic lung disease and brain injury.
- D.** Use larger tidal volumes to normalize PaCO<sub>2</sub>, accepting higher peak pressures to avoid permissive hypercapnia.

**182.** Prenatal ultrasonography showed echogenic bowel. After birth, the infant develops distal intestinal obstruction, and imaging is consistent with thick meconium plugging the terminal ileum. Which inherited disorder should be evaluated?

- A.** Phenylketonuria from PAH deficiency, which is asymptomatic in the immediate newborn period.
- B.** Cystic fibrosis from pathogenic CFTR variants producing meconium ileus with inspissated intestinal contents.
- C.** Hirschsprung disease from distal aganglionosis, which more often shows a transition zone and explosive stool after examination.
- D.** Galactosemia from GALT deficiency, which causes liver dysfunction and sepsis rather than meconium ileus.

**183.** A VLBW infant receives fortified human milk but has poor growth despite adequate measured volume. What is the most useful next nutritional step?

- A.** Assume poor growth is genetic because nutrition does not influence head or linear growth in VLBW infants.
- B.** Increase free-water intake first because more fluid reliably improves lean body mass accretion.
- C.** Review actual energy and protein delivery, fortification technique, losses, and growth trends before simply increasing fluid volume.
- D.** Discontinue fortification because added protein is a common cause of extrauterine growth restriction.

**184.** A 37-week infant with encephalopathy is being transferred to a cooling center from a hospital without therapeutic hypothermia capability. What is the most important systems principle?

- A.** Recognize possible HIE early and arrange prompt transfer or initiation of controlled cooling so the therapeutic window is not missed.
- B.** Wait until 12 hours of age to reassess because cooling started earlier increases brain injury.
- C.** Use uncontrolled deep cooling during transport because exact temperature targets are not important.
- D.** Delay referral until seizures occur because infants without seizures do not benefit from neuroprotective evaluation.

**185.** A critically ill 24-week infant has a mean arterial pressure numerically below gestational age but warm extremities, brisk capillary refill, normal lactate, and adequate urine output. What is the best interpretation?

- A.** Adequate urine output is sufficient evidence of normal systemic perfusion despite the low blood pressure.
- B.** Any mean arterial pressure below gestational age proves shock and requires immediate vasopressors.
- C.** A low numeric pressure with normal perfusion proves adrenal insufficiency and requires hydrocortisone.
- D.** Blood-pressure number alone should not define cardiovascular insufficiency; assess systemic perfusion and physiology before treating.

**186.** A preterm infant has severe respiratory failure despite conventional ventilation, with difficulty achieving gas exchange without injurious peak pressures. Which ventilation mode may be considered as a rescue strategy?

- A.** High-frequency ventilation to provide gas exchange with very small tidal volumes when conventional ventilation is failing.
- B.** Negative-pressure ventilation through an iron lung as the standard NICU rescue modality.
- C.** Continuous oxygen by hood without positive pressure despite severe ventilatory failure.
- D.** Immediate discontinuation of mechanical ventilation to reduce barotrauma regardless of gas exchange.

**187.** A preterm infant with cholestasis has poor weight gain and steatorrhea. Which nutrient modification may improve absorption while the underlying liver disease is addressed?

- A.** Use a formula or supplement with a higher proportion of medium-chain triglycerides when clinically appropriate.
- B.** Provide predominantly long-chain triglycerides because their absorption is relatively independent of bile acids in neonatal cholestasis.
- C.** Replace calories with free water because carbohydrate and protein worsen fat malabsorption.
- D.** Remove dietary fat during cholestasis because enteral lipid impairs weight gain and increases steatorrhea.

**188.** A 28-week infant is clinically stable on day 2 and receives tiny volumes of human milk while most calories come from parenteral nutrition. What is the purpose of these trophic feeds?

- A.** Replace parenteral nutrition immediately because trophic feeds meet the complete caloric requirement.
- B.** Deliver the infant's full protein requirement because trophic volumes contain enough amino acids for growth.
- C.** Use trophic feeds primarily to suppress intestinal bacterial colonization by keeping milk volumes below a microbial-growth threshold.
- D.** Provide enteral stimulation that supports gut maturation while larger nutritional volumes are advanced cautiously.

**189.** A newborn fails critical congenital heart disease screening with persistent right-hand saturation of 88% and foot saturation of 87% on repeated measurements. What is the next step?

- A.** Perform prompt clinical evaluation for hypoxemic disease, including echocardiography when cardiac disease is suspected.
- B.** Discharge the infant because a small hand-foot difference makes critical congenital heart disease unlikely.
- C.** Repeat screening at the routine 2-month visit because early low saturations are expected after 24 hours.
- D.** Give prophylactic indomethacin before imaging because ductal patency is the usual cause of screening failure.

**190.** A 30-week infant with worsening RDS is intubated for surfactant. The infant rapidly improves after the dose. Which ventilator adjustment is often necessary soon afterward?

- A.** Reduce inspiratory pressure or oxygen promptly as compliance and oxygenation improve to avoid overdistention and hyperoxia.
- B.** Maintain the pre-surfactant oxygen concentration for several hours to prevent rebound hypoxemia.
- C.** Discontinue PEEP after surfactant because improved compliance makes end-expiratory pressure unnecessary.
- D.** Increase pressures automatically because surfactant transiently decreases lung compliance after administration.

**191.** A neonate has a single arterial trunk arising from the heart, a large ventricular septal defect, and increasing pulmonary overcirculation as pulmonary vascular resistance falls. What is the diagnosis?

- A.** Isolated ventricular septal defect with normal aortic and pulmonary origins.
- B.** Tetralogy of Fallot with an intact pulmonary valve and separate great arteries.
- C.** Persistent truncus arteriosus with a single arterial trunk and large ventricular septal defect.
- D.** Transposition of the great arteries with two distinct semilunar valves.

**192.** A newborn has progressive respiratory distress. Chest radiograph shows marked hyperinflation of the left upper lobe with compression of adjacent lung and mediastinal shift. What is the most likely diagnosis?

- A.** Respiratory distress syndrome producing diffuse low-volume granular lungs in a preterm infant.
- B.** Pulmonary interstitial emphysema limited to a ventilated extremely preterm lung.
- C.** Transient tachypnea producing bilateral perihilar streaking and fluid in the fissures.
- D.** Congenital lobar overinflation caused by a ball-valve airway abnormality.

**193.** A neonate with critical valvar pulmonary stenosis has severe cyanosis and a small right ventricle but a patent ductus. Which catheter-based therapy most directly addresses the primary obstruction?

- A.** Device closure of the ductus before relieving the right-ventricular outflow obstruction.
- B.** Coil embolization of the pulmonary arteries to reduce pulmonary blood flow.
- C.** Perform balloon atrial septostomy because enlarging atrial communication directly relieves valvar pulmonary outflow obstruction.
- D.** Balloon pulmonary valvuloplasty to relieve the critically stenotic pulmonary valve.

**194.** Prenatal ultrasound shows a multicystic lung lesion. After birth the infant has respiratory distress and imaging confirms a cystic intrapulmonary mass supplied by the pulmonary circulation. What diagnosis is most likely?

- A.** Congenital pulmonary airway malformation, a developmental multicystic lesion supplied by the pulmonary circulation.
- B.** Pulmonary interstitial emphysema acquired after prolonged positive-pressure ventilation.
- C.** Bronchopulmonary sequestration supplied by an anomalous systemic artery from the aorta.
- D.** Congenital diaphragmatic hernia with bowel loops occupying the hemithorax.

**195.** A fetus has severe oligohydramnios from bilateral renal agenesis. At birth the infant has profound respiratory insufficiency despite an open airway. What is the primary pulmonary consequence of the prolonged oligohydramnios?

- A.** Pulmonary hypoplasia with inadequate alveolar and vascular development.
- B.** Primary surfactant overproduction causing obstructive air trapping in otherwise normal lungs.
- C.** Meconium aspiration caused directly by the absence of fetal urine production.
- D.** Transient pulmonary edema caused by excess amniotic fluid entering the fetal airways.

**196.** An extremely preterm infant is receiving supplemental oxygen. Which complication is most closely related to excessive oxygen exposure during retinal vascular development?

- A.** Choanal atresia caused by oxygen-mediated failure of posterior nasal canalization.
- B.** Esophageal atresia caused by postnatal oxygen toxicity to the foregut.
- C.** Retinopathy of prematurity from disrupted normal retinal vascularization and later pathologic neovascularization.
- D.** Congenital cataract caused by direct oxidation of the fetal lens after birth.

**197.** A 25-week infant with severe RDS has no echocardiographic evidence of pulmonary hypertension but remains hypoxemic on mechanical ventilation. What is the role of inhaled nitric oxide in this situation?

- A.** Routine rescue use is not recommended for preterm respiratory failure without a specific pulmonary-hypertension indication.
- B.** It should replace mechanical ventilation because it provides adequate alveolar ventilation without pressure.
- C.** Use inhaled nitric oxide routinely in preterm RDS because pulmonary vasodilation has been shown to prevent bronchopulmonary dysplasia.
- D.** It is the preferred therapy for surfactant deficiency because it replaces missing phospholipids in the alveoli.

**198.** A preterm infant has recurrent desaturation spells. One episode is accompanied by new temperature instability, abdominal distention, and lethargy. How should this event be approached?

- A.** Evaluate for an acquired cause such as sepsis rather than assuming every spell is apnea of prematurity.
- B.** Increase caffeine without further evaluation because infection does not present with apnea in preterm infants.
- C.** Diagnose gastroesophageal reflux as the cause because abdominal distention excludes systemic infection.
- D.** Discontinue monitoring because apnea becomes benign once caffeine therapy has started.

**199.** A very preterm infant receiving noninvasive support has frequent apnea and rising carbon dioxide despite adequate caffeine therapy. Which finding most strongly supports escalation to invasive ventilation?

- A.** An oxygen saturation within the unit target range on stable CPAP settings.
- B.** Mild intermittent tachypnea with normal carbon dioxide and no increased work of breathing.
- C.** Persistent apnea with inadequate ventilation and gas exchange despite optimized noninvasive support.
- D.** A single brief self-resolving desaturation without bradycardia or change in blood gas.

**200.** A preterm infant has a persistent pleural effusion. Thoracentesis yields milky fluid with a high triglyceride concentration after enteral feeds have begun. What is the most likely diagnosis?

- A.** Meconium aspiration causing direct passage of intestinal triglyceride into the thorax.
- B.** Neonatal chylothorax from lymphatic leakage into the pleural space.
- C.** Pulmonary hemorrhage causing a lipid-rich pleural effusion after enteral feeding.
- D.** Transient tachypnea causing sterile chylous fluid to accumulate in the pleural space.

## Answers

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1. **D.** Antenatal corticosteroids given when preterm birth is likely reduce neonatal respiratory distress syndrome, intraventricular hemorrhage, and mortality by accelerating fetal maturation. Tocolytics may briefly delay delivery in selected patients but do not replace corticosteroids, and neither furosemide nor vitamin K produces the desired pulmonary maturation effect.
2. **B.** In neonatal resuscitation, apnea or ineffective breathing with a heart rate below 100/min is an indication for positive-pressure ventilation. Ventilation is the critical intervention because most neonatal bradycardia is respiratory; compressions and epinephrine are reserved for persistent severe bradycardia despite effective ventilation.
3. **A.** The 2025 AAP clinical report emphasizes that prophylactic treatment and routine early closure before 2 weeks have not been shown to improve major outcomes. Management should incorporate clinical and echocardiographic evidence of hemodynamic significance rather than treating ductal patency alone.
4. **A.** The classic preterm presentation and low-volume reticulogranular radiograph indicate respiratory distress syndrome from surfactant deficiency. Surfactant deficiency raises surface tension, decreases compliance, and promotes diffuse atelectasis.
5. **B.** The combination of characteristic facial features, hypotonia, and complete atrioventricular septal defect is strongly associated with trisomy 21. The other chromosomal disorders have distinct dysmorphic and cardiac patterns.
6. **C.** Current AAP guidance identifies mother's own milk as the optimal enteral nutrition for hospitalized VLBW infants because of nutritional and immunologic benefits. Because unfortified human milk may not meet the high protein and mineral needs of very preterm infants, fortification is commonly required.
7. **D.** At birth, neonatal serum creatinine reflects maternal creatinine because the two compartments equilibrate across the placenta. Over subsequent days the infant's value changes according to renal function, gestational age, and postnatal physiology.
8. **B.** Infants of diabetic mothers often have pancreatic beta-cell hyperplasia and high insulin concentrations at birth. After cord clamping, the exogenous maternal glucose source ends but insulin remains elevated, producing rapid glucose decline.
9. **B.** Maternal IgG is actively transported across the placenta, with the greatest transfer late in gestation. Very preterm infants therefore miss much of this third-trimester acquisition and begin life with lower IgG concentrations.

- 10. D.** Early-onset neonatal sepsis is usually acquired vertically and is classically associated with organisms such as group B Streptococcus and E coli, especially in preterm infants and in the setting of membrane rupture or intra-amniotic infection. Coagulase-negative staphylococci and Candida are more typical late nosocomial pathogens.
- 11. D.** Pneumatosis intestinalis in a preterm infant with feeding intolerance, abdominal distention, and bloody stool is characteristic of NEC. Management depends on severity and includes bowel rest, gastric decompression, antibiotics, supportive care, and surgical evaluation for perforation or necrotic bowel.
- 12. B.** AAP guidance emphasizes objective predischarge bilirubin assessment and follow-up intensity based on the bilirubin's relation to the age- and gestation-specific phototherapy threshold plus clinical risk factors. Visual assessment alone can miss clinically important hyperbilirubinemia.
- 13. B.** Erythema toxicum is a common benign eruption that appears during the first days after birth in otherwise well term infants. Lesions are transient erythematous papules or pustules and typically spare the palms and soles.
- 14. D.** Newborns have limited vitamin K stores, and prophylactic vitamin K substantially reduces early and classic vitamin K deficiency bleeding. Deficiency impairs factors II, VII, IX, and X and typically prolongs PT; hemophilia affects the intrinsic pathway instead.
- 15. A.** The 2026 AAP clinical report supports therapeutic hypothermia to about 33.5–34.5°C, started within 6 hours and continued for 72 hours, for eligible infants born at least 36 weeks with moderate-to-severe HIE. Safe treatment requires controlled temperature, neuromonitoring, neuroimaging, and follow-up.
- 16. B.** Corrected age accounts for the number of weeks an infant was born before 40 weeks and is commonly used during the first 2 years when interpreting early developmental milestones. It prevents expected immaturity from being mislabeled as developmental delay.
- 17. B.** ROP screening includes infants with birth weight at or below 1500 g or gestational age at or below 30 weeks, plus selected larger or more mature infants with an unstable clinical course. The 29-week, 1280-g infant meets both primary criteria.
- 18. C.** Very preterm infants have a large fraction of body weight as total and extracellular water. Hydrophilic drugs may therefore require a relatively larger loading dose per kilogram to achieve a target concentration even though maintenance dosing may be limited by immature clearance.
- 19. A.** At the limits of viability, ethically appropriate counseling is individualized and family-centered. Shared decision-making communicates uncertainty, discusses outcomes and burdens, elicits values, and identifies medically reasonable plans that can be revisited as the clinical situation evolves.
- 20. D.** Absolute risk reduction is the difference in event rates: 12% minus 8% equals 4 percentage points. Relative risk reduction would be 4/12, about 33%, which is a different measure.

- 21. C.** For many spontaneously breathing preterm infants, early CPAP supports functional residual capacity and can avoid or reduce mechanical ventilation. Intubation is reserved for infants who fail noninvasive support or require surfactant/ventilation for clinical reasons.
- 22. D.** Trisomy 18 classically presents with severe growth restriction, prominent occiput, micrognathia, clenched overlapping fingers, and rocker-bottom feet. These findings are distinct from the midline defects of trisomy 13 and overgrowth syndromes.
- 23. B.** When mother's own milk is insufficient, donor human milk is commonly preferred for very preterm infants because it is associated with lower NEC risk than formula. Donor milk still requires nutritional fortification to support adequate protein, mineral, and energy intake.
- 24. A.** In prerenal states, reduced renal perfusion activates sodium-retaining mechanisms while tubular function remains relatively preserved, producing low urinary sodium and fractional excretion values. Clinical perfusion and fluid history support the diagnosis.
- 25. A.** Congenital hypothyroidism is often clinically subtle at birth, which is why newborn screening is essential. Prompt levothyroxine treatment during the critical period of brain development improves neurodevelopmental outcome.
- 26. D.** Secretory IgA is abundant in human milk and functions mainly at mucosal surfaces rather than through systemic absorption. It can bind pathogens and toxins and support intestinal barrier defense.
- 27. A.** Neonatal HSV can present as skin-eye-mouth, CNS, or disseminated disease, and maternal history may be negative. Suspected CNS or disseminated infection is a medical emergency requiring prompt IV acyclovir while PCR and other studies are obtained.
- 28. A.** Spontaneous intestinal perforation usually occurs in extremely preterm infants early in life and can present as an isolated focal perforation with less systemic inflammation and without the pneumatosis typical of NEC. Surgical management is often required.
- 29. C.** G6PD deficiency reduces red-cell protection from oxidative stress and can cause abrupt hemolysis with severe unconjugated hyperbilirubinemia. Bite cells and oxidant exposure are important clues, and affected infants can develop bilirubin neurotoxicity quickly.
- 30. D.** Transient neonatal pustular melanosis is benign and may be present at birth. Fragile pustules evolve into hyperpigmented macules with a characteristic collarette of scale, distinguishing it from erythema toxicum and infection.
- 31. C.** A venous hematocrit at or above the usual polycythemia threshold can increase blood viscosity and cause neurologic, metabolic, gastrointestinal, and respiratory symptoms. Management depends on symptoms and severity rather than the hematocrit value alone.

- 32. C.** The germinal matrix is highly vascular and fragile in very preterm infants. Hemorrhage may remain subependymal or extend into the ventricle; severity and ventricular dilation influence prognosis and follow-up needs.
- 33. B.** ELBW and extremely preterm infants have increased risks of motor, cognitive, language, hearing, and visual impairments. Structured high-risk follow-up helps identify problems early and connects families with therapy and developmental services.
- 34. C.** Treatment-requiring ROP must be addressed promptly before tractional retinal detachment develops. Laser ablation and intravitreal anti-VEGF agents are established options in selected disease patterns, with especially careful long-term follow-up after anti-VEGF therapy because recurrence can be delayed.
- 35. B.** Aminoglycosides are eliminated primarily by the kidneys. Reduced and gestation-dependent glomerular filtration in newborns, especially preterm infants, prolongs elimination and often requires extended dosing intervals with concentration-guided monitoring.
- 36. D.** Qualified interpreters improve accuracy, confidentiality, and informed decision-making. Clinicians should speak directly to the parent, avoid using children or untrained relatives as the default interpreter, and use teach-back or other methods to confirm understanding.
- 37. D.** The absolute risk reduction is 0.20 minus 0.15, or 0.05. Number needed to treat is the reciprocal,  $1/0.05 = 20$ .
- 38. C.** Maternal magnesium sulfate before anticipated early preterm birth decreases the risk of cerebral palsy among survivors and is used for fetal neuroprotection in the appropriate gestational-age range. Digoxin, bicarbonate, and routine prolonged maternal oxygen are not neuroprotective strategies for this purpose.
- 39. C.** A heart rate below 60/min despite effective ventilation is the threshold for starting neonatal chest compressions. The standard coordinated ratio is 3 compressions to 1 ventilation, emphasizing ventilation because the usual mechanism of newborn cardiac compromise is asphyxia.
- 40. D.** Beyond 2 weeks, evidence is less definitive and treatment is individualized. Many clinicians attempt one or two pharmacologic courses, commonly with ibuprofen, while transcatheter or surgical closure may be considered for selected infants with persistent hemodynamically significant shunts.
- 41. B.** Excessive weight loss with hypernatremia in an otherwise healthy breastfed newborn should prompt urgent assessment of milk transfer and hydration. Treatment requires careful rehydration because overly rapid correction of chronic hypernatremia can cause neurologic injury.
- 42. B.** Classic 21-hydroxylase deficiency reduces cortisol and aldosterone synthesis while increasing adrenal androgen production. Salt-wasting infants develop hyponatremia, hyperkalemia, dehydration, and shock, and affected genetic females may have virilized external genitalia.

- 43. C.** SCID causes severe impairment of cellular immunity, often with secondary humoral dysfunction, and can present in infancy with persistent infections, thrush, diarrhea, and lymphopenia. Newborn screening using T-cell receptor excision circles allows presymptomatic detection in many regions.
- 44. C.** Symptomatic congenital CMV commonly causes growth restriction, petechiae, hepatosplenomegaly, microcephaly, periventricular calcifications, and hearing loss. The location of intracranial calcifications is a useful discriminator from toxoplasmosis.
- 45. B.** Acholic stools and conjugated hyperbilirubinemia are red flags for obstructive cholestasis, particularly biliary atresia. Early diagnosis matters because outcomes of surgical portoenterostomy are better when performed promptly.
- 46. D.** Conjugated hyperbilirubinemia is not physiologic. It warrants evaluation for causes such as biliary atresia, infection, metabolic disease, and parenteral-nutrition-associated cholestasis; phototherapy treats unconjugated bilirubin and is not definitive therapy for cholestasis.
- 47. B.** Staphylococcal scalded skin syndrome is caused by exfoliative toxins that split the superficial epidermis, producing tender erythema, bullae, and superficial desquamation with relative mucosal sparing. Systemic antistaphylococcal therapy and supportive care are required.
- 48. B.** Neonatal alloimmune thrombocytopenia can cause severe thrombocytopenia in an otherwise well infant even during a first affected pregnancy because the mother lacks a fetal platelet antigen inherited from the father. The mother typically has a normal platelet count.
- 49. C.** Periventricular leukomalacia is ischemic-inflammatory injury of immature white matter in preterm infants. Damage near descending corticospinal fibers serving the legs helps explain the later association with spastic diplegic cerebral palsy.
- 50. C.** Spastic diplegia disproportionately affects the lower extremities and is a classic cerebral palsy pattern in children born very preterm, particularly after periventricular white-matter injury. Persistent abnormal tone and motor patterns warrant early therapy and formal assessment.
- 51. D.** A dense congenital cataract blocks patterned visual input during a critical period, placing the infant at high risk for deprivation amblyopia. Timely ophthalmologic assessment and treatment are therefore essential to preserve visual potential.
- 52. A.** Newborns have limited capacity to glucuronidate and eliminate chloramphenicol, so accumulation can cause cardiovascular collapse and the classic gray discoloration. The syndrome illustrates how developmental pharmacokinetics can make pediatric dose extrapolation unsafe.
- 53. C.** Withholding and withdrawing life-sustaining treatment are ethically equivalent when based on the same goals and best-interest analysis. If burdensome treatment no longer offers proportionate benefit, transition to comfort-focused care can be appropriate, with aggressive symptom management.

- 54. C.** Sensitivity is the probability that a test is positive among individuals who truly have the disease. Highly sensitive tests have few false negatives, so a negative result can be useful for ruling out disease, although interpretation still depends on context and test performance.
- 55. B.** Asymmetric fetal growth restriction is typical of placental insufficiency, in which chronic nutrient and oxygen limitation leads to brain-sparing redistribution and relatively greater restriction of abdominal growth. Fetal hyperinsulinemia tends to cause macrosomia, while skeletal dysplasia and maternal thyroid disease do not produce the classic placental Doppler and brain-sparing pattern.
- 56. C.** Epinephrine is indicated when the heart rate remains below 60/min despite effective ventilation and coordinated chest compressions, with intravascular delivery preferred once access is established. Atropine, adenosine, and routine calcium are not standard treatments for asphyxial neonatal bradycardia in the delivery room.
- 57. D.** In transposition, survival depends on mixing between the parallel systemic and pulmonary circuits. Prostaglandin maintains ductal mixing, and a restrictive atrial communication with severe hypoxemia may require urgent balloon atrial septostomy; ductal closure would worsen the physiology.
- 58. A.** Exogenous surfactant replaces the deficient surface-active material responsible for low compliance and atelectasis in RDS. Oxygen and CPAP support gas exchange, but surfactant directly addresses the biochemical deficiency when disease severity warrants treatment.
- 59. C.** Trisomy 13 is strongly associated with holoprosencephaly and other midline defects, cleft lip or palate, microphthalmia, and postaxial polydactyly. The alternative syndromes do not fit this constellation.
- 60. B.** Human milk fortification is used in very preterm and VLBW infants because human milk alone may not meet their high protein and mineral requirements. Fortification supports growth and bone mineralization while preserving the benefits of human milk.
- 61. C.** The classic triad of congenital toxoplasmosis is hydrocephalus, intracranial calcifications that are often diffuse, and chorioretinitis. Maternal infection may be mild or unrecognized.
- 62. A.** Bilious emesis in a newborn is a surgical emergency until obstruction is excluded. Malrotation with volvulus can rapidly compromise the superior mesenteric blood supply, and the abnormal duodenal course on imaging supports urgent surgical intervention.
- 63. D.** Phototherapy alters bilirubin through photoisomerization and related photochemical reactions, creating products that can be eliminated without normal glucuronidation. It does not depend on immediate maturation of hepatic conjugating enzymes.
- 64. C.** Epidermolysis bullosa comprises inherited disorders in which defects in proteins linking epidermal or dermal layers cause blistering with minor trauma. A family history and recurrent friction-induced lesions support a structural skin disorder rather than infection.

- 65. A.** Maternal antiplatelet IgG can cross the placenta and reduce neonatal platelets in maternal immune thrombocytopenia. Unlike neonatal alloimmune thrombocytopenia, the mother herself is usually thrombocytopenic because her antibodies also target her own platelets.
- 66. C.** Phenobarbital remains a commonly recommended first-line antiseizure medication for most acute neonatal seizures, alongside urgent treatment of reversible causes such as hypoglycemia, hypocalcemia, infection, or stroke. Electrographic seizure burden can matter even when clinical movements are subtle.
- 67. C.** Neurodevelopment after very preterm birth is multidimensional. Some children without major early motor abnormalities later show language, attention, executive-function, learning, or behavioral difficulties, supporting longitudinal surveillance into childhood.
- 68. B.** Auditory brainstem response assesses neural conduction as well as hearing sensitivity and is preferred for many NICU infants because auditory neuropathy is more common in this population. Otoacoustic emissions primarily assess cochlear outer hair-cell function and can miss neural disorders.
- 69. A.** Ceftriaxone is highly protein bound and can displace bilirubin, raising concern for bilirubin neurotoxicity, and it can form dangerous precipitates with calcium-containing IV products. Alternative cephalosporins are often preferred in neonates when these risks are relevant.
- 70. D.** Equity work asks whether systems create different access or outcomes across groups and seeks modifiable structural barriers. Effective improvement uses stratified data, family input, and redesign of processes rather than attributing disparities to individual motivation alone.
- 71. B.** Specificity is the true-negative rate: the proportion of disease-free individuals who test negative. Positive predictive value is different and varies with disease prevalence.
- 72. D.** Maternal hyperglycemia drives fetal pancreatic beta-cell hyperplasia and hyperinsulinemia. After cord clamping, maternal glucose delivery stops abruptly while insulin remains high, creating early neonatal hypoglycemia; the alternative mechanisms do not fit the physiology of an infant of a diabetic mother.
- 73. C.** Routine tracheal suctioning of nonvigorous infants solely because of meconium-stained fluid is no longer recommended. Effective ventilation should not be delayed; airway suction is reserved for suspected obstruction that interferes with ventilation.
- 74. B.** Critical coarctation can present with shock when the ductus closes because lower-body systemic perfusion becomes duct-dependent. Prostaglandin E1 reopens or maintains the ductus and stabilizes systemic blood flow while echocardiography and surgical planning proceed.
- 75. A.** Cesarean delivery without labor is associated with delayed clearance of fetal lung fluid. The mild oxygen requirement and radiographic interstitial fluid with fissural fluid are typical of transient tachypnea, which usually improves over the first 1 to 3 days.

- 76. B.** Turner syndrome can be recognized in the newborn by congenital lymphedema, webbed neck, shield chest, and left-sided cardiac lesions such as coarctation. The phenotype and cardiac clues make monosomy X the best fit.
- 77. A.** Very preterm infants have high protein needs and limited nutrient stores, so parenteral amino acids and energy are started early when enteral intake is inadequate. Prolonged dextrose-only nutrition increases catabolism and does not support lean tissue accretion.
- 78. B.** Very preterm kidneys have limited tubular sodium reabsorptive capacity and can lose substantial sodium during growth. Persistent hyponatremia with urinary sodium loss may require individualized sodium supplementation after other causes are excluded.
- 79. B.** Newborns rely heavily on brown-adipose-tissue nonshivering thermogenesis. Cold stress increases metabolic rate, oxygen consumption, and glucose utilization and can worsen hypoxemia, acidosis, and hypoglycemia, especially in preterm infants.
- 80. A.** The 22q11.2 deletion can impair development of the thymus and parathyroids, producing T-cell lymphopenia and hypocalcemia along with conotruncal cardiac disease. Severity of immune dysfunction varies widely among affected infants.
- 81. A.** Preterm infants have an attenuated erythropoietin response to anemia, shorter red-cell survival, rapid growth, and often substantial blood-sampling losses. The result is a hyporegenerative anemia that reaches its nadir earlier and more severely than physiologic anemia of term infancy.
- 82. C.** Perinatal arterial ischemic stroke often presents with focal seizures in an otherwise relatively well term infant. MRI with diffusion-weighted imaging demonstrates the arterial-territory infarct and guides evaluation and prognosis.
- 83. D.** Some NICU risk factors are associated with delayed-onset or progressive hearing loss. A normal newborn screen is reassuring but does not eliminate the need for later audiologic follow-up when significant risk factors are present.
- 84. C.** Bilateral choanal atresia obstructs the posterior nasal passages. Because newborns preferentially breathe through the nose, cyanosis may worsen with feeding and improve with crying when the mouth opens.
- 85. D.** The noninferiority margin is prespecified and represents the greatest clinically acceptable decrement in efficacy. The confidence interval for the treatment difference is then compared with this margin; choosing the margin after seeing results would invalidate the inference.
- 86. B.** Anti-D IgG crosses the placenta and can produce fetal hemolytic anemia. Increased middle cerebral artery peak systolic velocity is a noninvasive marker of reduced blood viscosity and increased flow associated with significant fetal anemia, not polycythemia or isolated thrombocytopenia.

- 87. D.** For term and late-preterm newborns needing respiratory support, initial resuscitation generally begins with 21% oxygen, with titration based on preductal saturation targets and response. Routine use of 100% oxygen exposes the infant to unnecessary oxidative stress.
- 88. D.** With pulmonary atresia and inadequate antegrade pulmonary flow, pulmonary circulation may depend on the ductus arteriosus. Prostaglandin E1 is therefore a stabilizing therapy until catheter-based or surgical intervention is available.
- 89. B.** Meconium aspiration produces a combination of partial airway obstruction, air trapping, atelectasis, inflammation, and sometimes pulmonary hypertension. The radiograph is often patchy and hyperinflated rather than the low-volume diffuse pattern of surfactant-deficient RDS.
- 90. A.** The 22q11.2 deletion syndrome is associated with conotruncal cardiac defects, thymic hypoplasia with T-cell dysfunction, hypocalcemia from parathyroid hypoplasia, and characteristic craniofacial findings. The other variants produce different neonatal phenotypes.
- 91. C.** Essential fatty acids must be supplied because infants cannot synthesize them de novo. Prolonged lipid deprivation can cause dermatitis, growth failure, thrombocytopenia, and characteristic biochemical changes.
- 92. D.** Posterior urethral valves are a major cause of congenital lower urinary tract obstruction in male infants. Findings include poor stream, bladder distention and hypertrophy, hydroureteronephrosis, and potential renal dysplasia from prenatal obstruction.
- 93. D.** Severe hypomagnesemia can impair both secretion of parathyroid hormone and responsiveness to it, leading to persistent hypocalcemia. Calcium replacement alone may fail until magnesium is corrected.
- 94. B.** Neonatal lupus is caused by transplacental maternal anti-Ro/SSA or anti-La/SSB antibodies. The rash is transient as maternal antibodies clear, but congenital heart block reflects permanent injury to the fetal conduction system.
- 95. D.** Congenital rubella syndrome classically includes cataracts or other ocular disease, sensorineural hearing impairment, and congenital heart disease such as PDA or peripheral pulmonary stenosis. The pattern is highly characteristic.
- 96. C.** Hirschsprung disease causes functional obstruction because an aganglionic distal segment cannot relax normally. Delayed meconium, distention, and explosive stool after rectal examination are classic clues; rectal suction biopsy establishes the diagnosis.
- 97. D.** Early-intervention programs are designed to address functional risk and emerging delay, not only established disability. Early childhood is a period of rapid neurodevelopment and family adaptation, so timely support can be valuable even while the long-term diagnosis remains uncertain.

- 98. D.** Pierre Robin sequence begins with mandibular hypoplasia, leading to posterior displacement of the tongue and upper-airway obstruction; a U-shaped cleft palate is common. Airway and feeding management are immediate priorities.
- 99. C.** Intention-to-treat analysis preserves the original randomized groups and therefore protects against many biases introduced after randomization. Per-protocol analyses may be informative as secondary analyses, especially in noninferiority trials, but do not replace the randomized primary comparison.
- 100. C.** Maternal TSH-receptor stimulating antibodies can cross the placenta and cause fetal or neonatal thyrotoxicosis even when the mother's biochemical thyroid status is controlled. TSH itself does not cross the placenta in clinically important amounts, and the other proposed mechanisms do not explain antibody-mediated neonatal Graves disease.
- 101. D.** Preterm infants should receive carefully titrated oxygen rather than routine 100% oxygen. Current practice uses blended oxygen with pulse-oximetry guidance because both hypoxemia and excessive oxygen exposure can be harmful in this population.
- 102. C.** Hypercyanotic spells result from increased right-to-left shunting and reduced pulmonary blood flow. Increasing systemic vascular resistance with knee-chest positioning, calming the infant, and additional targeted therapy can reduce the shunt; systemic vasodilation would worsen it.
- 103. B.** Sudden cardiorespiratory collapse with unilateral absent breath sounds and supportive transillumination findings indicates tension pneumothorax. When the infant is unstable, immediate decompression is lifesaving and should not be delayed for radiographic confirmation.
- 104. B.** Noonan syndrome can resemble Turner syndrome phenotypically but occurs in either sex and is strongly associated with pulmonary valve stenosis and hypertrophic cardiomyopathy. It is usually caused by variants affecting the RAS-MAPK signaling pathway.
- 105. A.** Very preterm infants miss much of the third-trimester mineral accretion period and may develop metabolic bone disease if calcium and phosphorus delivery is inadequate. Low phosphorus and high alkaline phosphatase are common laboratory clues.
- 106. A.** The classic triad of renal enlargement, hematuria, and thrombocytopenia suggests renal vein thrombosis, especially in infants with dehydration, maternal diabetes, or other thrombotic risk factors. Doppler ultrasonography is typically used for evaluation.
- 107. B.** Very high ammonia with relatively little metabolic acidosis is a classic clue to a urea-cycle defect. Organic acidemias can also raise ammonia, but usually produce a substantial anion-gap acidosis and ketosis.

**108. D.** Most routine childhood immunizations are given according to chronological age in preterm infants, with specific exceptions and product considerations such as hepatitis B recommendations for very low birth weight infants. Prematurity alone is not a reason to postpone all vaccines to term-equivalent age.

**109. D.** Early congenital syphilis can cause snuffles, hepatosplenomegaly, rash involving palms and soles, anemia or thrombocytopenia, and osteochondritis or periostitis. Parenteral penicillin is the treatment of choice.

**110. C.** Failure to pass an orogastric tube with proximal coiling strongly suggests esophageal atresia. The common form has a distal tracheoesophageal fistula, allowing air into the stomach and increasing aspiration risk.

**111. A.** DIC is a consumptive coagulopathy triggered by severe systemic illness such as sepsis or asphyxia. Widespread coagulation activation consumes platelets and clotting factors and generates fibrin degradation products, producing both thrombosis and bleeding risk.

**112. A.** Erb palsy results from injury to upper brachial plexus roots, often during shoulder dystocia. The classic posture is shoulder adduction and internal rotation with elbow extension; preserved grasp suggests lower plexus function is intact.

**113. C.** Case-control studies begin with outcome status and then assess prior exposures. They are efficient for uncommon outcomes but are vulnerable to selection and recall or measurement bias and typically estimate odds ratios rather than incidence directly.

**114. A.** Late-pregnancy SSRI exposure can be followed by a self-limited poor neonatal adaptation syndrome with jitteriness, tone abnormalities, respiratory symptoms, and feeding difficulty. Sepsis should still be considered when clinically indicated, but SSRI exposure itself does not cause bacterial infection or opioid withdrawal.

**115. B.** A supraglottic airway is an effective alternative when face-mask ventilation is unsuccessful and endotracheal intubation is not feasible or fails in an appropriately sized newborn. Continuing ineffective ventilation or moving directly to compressions does not correct the primary problem, and surgical airway creation is not the routine neonatal rescue approach.

**116. C.** In hypoplastic left heart syndrome, systemic and pulmonary circulations are supplied in parallel from the right ventricle, and systemic output is duct-dependent. Marked reduction in pulmonary vascular resistance can divert excessive flow to the lungs and compromise systemic perfusion, so oxygen and ventilation are titrated to preserve balance.

**117. D.** Caffeine is the standard pharmacologic treatment for apnea of prematurity because it stimulates central respiratory drive and has a favorable safety profile. Secondary causes of apnea should be considered before attributing events solely to prematurity.

- 118. A.** Beckwith-Wiedemann syndrome is an overgrowth disorder characterized by macroglossia, abdominal wall defects, organomegaly, and neonatal hypoglycemia from hyperinsulinism. These features distinguish it from hypotonia and growth-restriction syndromes.
- 119. A.** Preterm infants have lower iron stores at birth, experience rapid growth, and may have substantial phlebotomy losses. Enteral iron supplementation is therefore commonly required once feeds are established, with the dose individualized to transfusion history and diet.
- 120. B.** Loop diuretics increase urinary calcium excretion and, together with prematurity and other mineral factors, can contribute to nephrocalcinosis. Ultrasound may show medullary echogenicity in affected infants.
- 121. A.** Maple syrup urine disease causes accumulation of branched-chain amino acids and ketoacids, producing neonatal encephalopathy and a characteristic sweet odor. Early recognition is crucial because rapid reduction of toxic metabolites can prevent further neurologic injury.
- 122. B.** Listeria can cross the placenta and cause severe fetal or neonatal infection. Granulomatosis infantiseptica, with disseminated granulomatous lesions, is a classic though uncommon manifestation of congenital listeriosis.
- 123. A.** Gastroschisis is a full-thickness abdominal wall defect, usually to the right of the umbilicus, with uncovered bowel exposed to amniotic fluid. Initial management includes protection of the bowel, thermoregulation, gastric decompression, fluids, and surgical planning.
- 124. C.** Hemophilia A and B are X-linked deficiencies of factors VIII and IX that prolong the aPTT while platelet count and PT remain normal. Affected male newborns can bleed after circumcision, procedures, or traumatic delivery.
- 125. C.** Peripheral facial nerve injury can occur from compression or instrumentation during delivery and affects the entire ipsilateral face. Central lesions often spare forehead function, making the pattern useful for localization.
- 126. B.** The most common reason for failure to improve during neonatal resuscitation is ineffective ventilation. Before compressions are started, the team should perform ventilation corrective steps and confirm chest movement and heart-rate response; compressions are indicated only after effective ventilation has been established and severe bradycardia persists.
- 127. C.** Obstructed total anomalous pulmonary venous return is a surgical emergency because pulmonary venous obstruction causes severe pulmonary edema, hypoxemia, and low output. Medical stabilization is temporary; definitive therapy is surgical redirection of pulmonary venous return to the left atrium.

- 128. D.** Bag-mask ventilation can insufflate bowel in the thorax and worsen lung compression in congenital diaphragmatic hernia. The infant is typically intubated, the stomach decompressed, and ventilation kept gentle while pulmonary hypertension and perfusion are stabilized before surgery.
- 129. B.** Prader-Willi syndrome results from loss of paternal expression in 15q11-q13 and often presents in the neonatal period with profound hypotonia and feeding difficulty. Hyperphagia and obesity emerge later, making the early phenotype very different from the later classic presentation.
- 130. C.** Vitamin D supports calcium and phosphorus homeostasis and is routinely provided in preterm nutrition plans. It complements, rather than replaces, adequate mineral and protein intake.
- 131. B.** Acute tubular injury may evolve from an oliguric phase into a diuretic recovery phase as filtration improves before tubular reabsorptive and concentrating capacity fully recovers. During this phase, careful replacement of fluid and electrolyte losses is required.
- 132. A.** Fatty-acid oxidation disorders often present during fasting or illness with hypoketotic hypoglycemia because the infant cannot effectively use fatty acids to generate energy and ketone bodies. Avoidance of fasting is a key preventive strategy after diagnosis.
- 133. C.** ELBW infants with central lines, parenteral nutrition, and prolonged antibiotic exposure are at increased risk for invasive candidiasis. Clinical findings can be nonspecific, and persistent thrombocytopenia or instability may be clues.
- 134. C.** Omphalocele is a midline defect through the umbilical ring with herniated viscera covered by a membrane. Because associated cardiac, chromosomal, and other anomalies are common, the infant requires careful evaluation beyond the abdominal wall lesion itself.
- 135. A.** G6PD-deficient red cells cannot generate adequate reduced glutathione during oxidative stress. Heinz bodies are removed by splenic macrophages, producing bite cells and episodes of hemolysis with potentially rapid bilirubin rise.
- 136. D.** Severe or prolonged neonatal hypoglycemia can injure the brain, with posterior and occipital regions frequently involved. Later visual dysfunction, epilepsy, and neurodevelopmental difficulties may occur depending on injury severity.
- 137. C.** Gestational age influences both the likelihood of prolonged ventilation and neurodevelopmental outcome, so it can confound the observed association. Design or analytic methods such as restriction, matching, stratification, or multivariable adjustment can address measured confounding.
- 138. B.** Neonatal opioid withdrawal commonly produces a combination of central nervous system irritability, gastrointestinal dysfunction, and autonomic symptoms after chronic prenatal opioid exposure. The timing and multisystem pattern are not explained by transient tachypnea, serotonin toxicity, or isolated hypocalcemia.

- 139. A.** VACTERL is a nonrandom association of vertebral, anorectal, cardiac, tracheoesophageal, renal, and limb abnormalities. The diagnosis is clinical and requires recognition of the multisystem pattern rather than a single defining gene in most cases.
- 140. D.** Gastric residuals are variable and have poor specificity for NEC or feeding intolerance when considered in isolation. Feeding decisions should integrate the infant's overall examination, abdominal findings, emesis, stool pattern, and systemic status.
- 141. D.** SIADH causes inappropriate water retention, hyponatremia, low serum osmolality, and urine that remains more concentrated than expected. Assessment of volume status is important because renal salt wasting can produce a superficially similar laboratory pattern with hypovolemia.
- 142. C.** Classic galactosemia can present rapidly after lactose exposure with hepatic dysfunction, jaundice, poor feeding, and a characteristic association with *E coli* sepsis. Galactose-containing feeds must be stopped while diagnosis and treatment proceed.
- 143. B.** Late-onset GBS disease typically occurs from about 7 days to 3 months and often presents with bacteremia or meningitis. Intrapartum prophylaxis reduces early-onset disease but does not eliminate late-onset disease.
- 144. A.** High fetal hemoglobin concentrations suppress polymerization of hemoglobin S, so most infants with sickle cell disease are clinically well in the newborn period. Symptoms emerge as fetal hemoglobin falls over the first months of life.
- 145. B.** Acute profound asphyxia can preferentially injure metabolically active deep gray nuclei and perirolandic cortex. More prolonged partial hypoxia is more likely to produce watershed cortical and subcortical injury.
- 146. A.** For a relative risk, 1.0 is the null value. Because the confidence interval includes 1.0, a conventional two-sided test would not show a statistically significant difference; this does not prove equivalence, and the interval still permits clinically important benefit or harm.
- 147. D.** Clinical intra-amniotic infection increases the newborn's risk of early-onset sepsis through ascending infection and intrapartum exposure. Catheter-associated sepsis is a later nosocomial problem, and congenital CMV and tetanus have different routes and epidemiology.
- 148. A.** A large VSD may be relatively quiet immediately after birth while pulmonary vascular resistance is high. As pulmonary resistance falls over the first weeks, left-to-right shunting and pulmonary overcirculation increase, producing heart failure symptoms and poor growth.
- 149. D.** Persistent pulmonary hypertension results from failure of the normal postnatal fall in pulmonary vascular resistance, causing right-to-left shunting through fetal channels and severe hypoxemia. Pneumonia and meconium aspiration are common associated triggers.

**150. D.** Organic acidemias cause accumulation of organic acids, leading to high-anion-gap acidosis and ketosis; secondary hyperammonemia and bone-marrow suppression can also occur. The acid-base pattern helps distinguish them from primary urea-cycle disorders.

**151. C.** Coagulase-negative staphylococci are common skin organisms and also major causes of late-onset bloodstream infection in preterm infants with indwelling vascular catheters. Clinical context and repeated cultures help distinguish true infection from contamination.

**152. C.** Transient abnormal myelopoiesis occurs almost exclusively in infants with trisomy 21 and is associated with GATA1-mutant megakaryoblasts. Many cases regress spontaneously, although severe neonatal complications can occur and survivors have increased later risk of myeloid leukemia of Down syndrome.

**153. B.** Amplitude-integrated EEG is useful for continuous trend monitoring of cerebral background and seizure activity in the NICU. It is less detailed than conventional EEG and should complement, not replace, clinical assessment and formal electroencephalography when diagnostic precision is needed.

**154. B.** Plan-Do-Study-Act cycles use rapid, iterative testing: plan a change, implement it on a small scale, study the results, and act on what was learned. This approach is designed for local process improvement and adaptation rather than definitive efficacy testing.

**155. D.** Current neonatal resuscitation guidance supports deferred cord clamping for at least about 60 seconds in many newborns who do not require immediate resuscitation, including preterm infants when feasible. Immediate clamping is not required solely because of prematurity, while prolonged delay or indiscriminate cord milking is not the routine standard.

**156. D.** Maternal anti-Ro/SSA and anti-La/SSB antibodies can injure the fetal conduction system and cause irreversible congenital complete heart block. Atrial and ventricular dissociation with profound persistent bradycardia is characteristic.

**157. C.** Pulmonary hemorrhage in a very preterm infant can present abruptly with blood from the airway, respiratory deterioration, and diffuse radiographic opacification. It is associated with severe lung disease and circulatory factors such as a large PDA.

**158. C.** CHARGE is an acronymic pattern that includes coloboma, heart defects, choanal atresia, growth and developmental issues, genital anomalies, and ear abnormalities. CHD7 variants are a common cause.

**159. A.** Although some trials and meta-analyses suggest probiotic benefit, the AAP has emphasized concerns about heterogeneous products, lack of pharmaceutical-grade regulation, potential contamination, and limited safety data in extremely vulnerable infants. Product- and population-specific judgment is required.

- 160. A.** Bilateral renal agenesis causes absent fetal urine production and profound oligohydramnios. The resulting pulmonary hypoplasia is often the immediate lethal problem, in addition to the absence of renal function.
- 161. D.** A weak but alert infant with areflexia suggests disease of the anterior horn cell, peripheral nerve, neuromuscular junction, or muscle. Central hypotonia more often accompanies abnormal consciousness, seizures, or preserved and sometimes brisk reflexes.
- 162. B.** Twin-twin transfusion syndrome results from unbalanced flow through placental vascular connections. The recipient may develop hypervolemia, polycythemia, hypertension, and cardiac strain, whereas the donor is more likely to be growth-restricted, hypovolemic, and anemic.
- 163. B.** For a hemodynamically stable infant with regular narrow-complex supraventricular tachycardia that persists after vagal maneuvers, rapid IV adenosine is first-line acute therapy. Unstable infants require synchronized cardioversion; atropine and ductal therapy do not terminate reentry SVT.
- 164. D.** Pulmonary interstitial emphysema occurs when alveolar air ruptures into the interstitial tissues, typically in ventilated very preterm infants. Radiographs show characteristic linear or cystic lucencies within the lungs.
- 165. D.** Achondroplasia results from gain-of-function variants in FGFR3 that inhibit endochondral bone growth. The neonatal phenotype is disproportionate rhizomelic short stature with macrocephaly and midface hypoplasia.
- 166. C.** Enteral feeding stimulates bile flow and reduces dependence on parenteral nutrition, while overfeeding carbohydrate and prolonged parenteral exposure can worsen liver injury. Management also includes avoiding excess energy delivery and considering lipid strategies when cholestasis is established.
- 167. B.** Infants born to HBsAg-positive mothers require both active immunization with hepatitis B vaccine and passive prophylaxis with hepatitis B immune globulin soon after birth. Follow-up serologic testing later confirms protection and excludes infection.
- 168. A.** Jitteriness is often stimulus-sensitive and can usually be stopped by gentle flexion or holding the limb; ocular deviation and autonomic changes are uncommon. True seizures are more stereotyped, less suppressible, and may include ocular or autonomic manifestations.
- 169. A.** For appropriately selected preterm prelabor rupture of membranes, latency antibiotics reduce infection and prolong pregnancy, while antenatal corticosteroids remain beneficial. Repeated digital examinations increase infection risk, and routine antifungal therapy is not the standard latency regimen.
- 170. B.** Ebstein anomaly is characterized by apical displacement of the septal and posterior tricuspid leaflets, atrialization of part of the right ventricle, and often substantial tricuspid regurgitation. The echocardiographic anatomy distinguishes it from septal and outflow-tract lesions.

- 171. C.** Bronchopulmonary dysplasia is the chronic lung disease of prematurity and is classified using oxygen and respiratory-support needs at a standardized postmenstrual age, commonly around 36 weeks. Persistent support after extreme prematurity is not compatible with transient tachypnea.
- 172. D.** Osteogenesis imperfecta is a heritable disorder of type I collagen that causes bone fragility, fractures, and often blue sclerae. The radiographic osteopenia and fractures beginning before or at birth support a severe collagen disorder rather than a postnatal nutritional deficiency.
- 173. A.** Weight gain alone can reflect fluid or fat accretion. Poor linear and head growth with low markers of protein intake should prompt review of protein and overall nutrient adequacy rather than assuming calories alone are sufficient.
- 174. D.** Candida recovered from blood in a sick preterm infant represents invasive infection until proven otherwise. Management includes systemic antifungal therapy, attention to central venous catheters, and evaluation for organ dissemination when appropriate.
- 175. A.** A hemodynamically important left-to-right PDA creates diastolic runoff from the aorta to the pulmonary artery and increases pulmonary blood flow. The result can be bounding pulses, a hyperdynamic precordium, and a widened pulse pressure.
- 176. A.** AAP guidance does not support routine corticosteroid use for all preterm infants, but low-dose systemic dexamethasone after the first week can be considered for selected infants who remain on significant respiratory support and have high BPD risk. High-dose dexamethasone is not recommended, and inhaled therapy has not shown a clear safety advantage.
- 177. D.** Spinal muscular atrophy is an anterior-horn-cell disorder caused by loss of SMN1 function. Weakness, areflexia, and tongue fasciculations with preserved sensorium point to a peripheral motor-neuron process rather than central encephalopathy.
- 178. B.** Late-preterm and preterm infants often need time to coordinate sucking, swallowing, and breathing. Safe feeding plans may combine gavage feeds, paced oral attempts, and therapy input rather than pushing volumes that provoke physiologic instability.
- 179. B.** Meningitis can impair CSF pathways through inflammation, debris, or scarring and can lead to hydrocephalus. Serial head circumference, cranial imaging, and neurosurgical assessment may be required when ventricular enlargement progresses.
- 180. A.** A substantial preductal-postductal saturation gradient suggests right-to-left shunting through the ductus, a classic manifestation of persistent pulmonary hypertension. The high pulmonary vascular resistance diverts deoxygenated pulmonary-artery blood into the descending aorta.
- 181. B.** Lung-protective ventilation minimizes volutrauma and barotrauma through appropriate tidal-volume targeting, sufficient PEEP, and avoidance of unnecessary hypocapnia. Modest permissive hypercapnia can be accepted in selected infants if pH and clinical status are appropriate.

- 182. B.** Meconium ileus is strongly associated with cystic fibrosis and can be the first clinical manifestation. A microcolon with inspissated distal ileal meconium supports obstructive disease from abnormal intestinal secretions.
- 183. C.** Growth failure in VLBW infants requires a systematic review of delivered nutrients, fortification, feeding interruptions, illness, and fluid status. Increasing volume alone may not correct inadequate protein or caloric density and can worsen fluid-related morbidity.
- 184. A.** Therapeutic hypothermia has a narrow evidence-based initiation window, so delivery centers need protocols for early recognition, stabilization, communication, and transfer. Cooling should be controlled to a defined target rather than improvised to excessively low temperatures.
- 185. D.** Very preterm infants can have low measured blood pressure without evidence of impaired systemic perfusion. Treatment decisions should integrate clinical perfusion, lactate, urine output, echocardiographic information when needed, and the underlying physiology rather than relying on a single numeric threshold.
- 186. A.** High-frequency ventilation is used in selected neonates with severe respiratory failure or problematic air leak when conventional ventilation cannot provide adequate gas exchange without injurious pressures. It is a rescue tool, not a substitute for needed ventilatory support.
- 187. A.** Medium-chain triglycerides are absorbed more directly and depend less on bile-salt-mediated micelle formation, so they can improve energy absorption in cholestatic infants. Essential long-chain fatty acids still must be supplied, so complete fat elimination is inappropriate.
- 188. D.** Trophic or minimal enteral feeds provide luminal stimulation to the immature gastrointestinal tract while full nutritional support is still provided by parenteral nutrition. Their goal is gut adaptation and feeding progression, not immediate fulfillment of total nutrient needs.
- 189. A.** Persistent low oxygen saturation constitutes a failed screen and requires evaluation for critical congenital heart disease and other causes of neonatal hypoxemia. A small preductal-postductal difference does not make the result normal, and treatment should be guided by diagnosis rather than empiric ductal closure.
- 190. A.** Exogenous surfactant can improve compliance and oxygenation quickly. Ventilator pressures and inspired oxygen should be reassessed frequently and reduced when possible to avoid volutrauma and excessive oxygen exposure.
- 191. C.** Persistent truncus arteriosus consists of a single great vessel supplying systemic, pulmonary, and coronary circulations, usually over a large VSD. As pulmonary vascular resistance falls, pulmonary overcirculation and heart failure can become prominent.

**192. D.** Congenital lobar overinflation classically produces focal hyperinflation, mass effect, and respiratory distress. The focal anatomic pattern distinguishes it from diffuse neonatal parenchymal diseases.

**193. D.** Critical valvar pulmonary stenosis is an anatomic obstruction at the pulmonary valve and is commonly treated with catheter balloon valvuloplasty when feasible. Ductal patency may support pulmonary blood flow temporarily, but closing the ductus before relieving the obstruction can be dangerous.

**194. A.** Congenital pulmonary airway malformation is a developmental cystic lesion of lung tissue that typically has pulmonary arterial supply. Sequestration is distinguished by systemic arterial supply, while diaphragmatic hernia contains abdominal viscera rather than a primary cystic lung mass.

**195. A.** Amniotic fluid volume and fetal breathing movements are important for lung growth. Severe prolonged oligohydramnios, as in bilateral renal agenesis, can cause marked pulmonary hypoplasia and lethal respiratory failure.

**196. C.** Excessive oxygen exposure contributes to the pathogenesis of retinopathy of prematurity by altering normal vascular growth in the immature retina. Oxygen is therefore titrated carefully in preterm infants to avoid both hypoxemia and hyperoxia.

**197. A.** Inhaled nitric oxide is an established pulmonary vasodilator for selected term or near-term infants with hypoxic respiratory failure and pulmonary hypertension. Routine use in preterm respiratory failure without a clear pulmonary-hypertension indication has not shown consistent outcome benefit.

**198. A.** Apnea is common in premature infants, but a change in pattern or associated systemic signs should prompt evaluation for secondary causes such as infection, anemia, metabolic disturbance, or airway disease. New lethargy and temperature or abdominal abnormalities make simple apnea of prematurity an unsafe assumption.

**199. C.** Escalation to invasive ventilation is appropriate when noninvasive support cannot maintain adequate ventilation, oxygenation, or respiratory drive. Recurrent clinically significant apnea with carbon-dioxide retention indicates failure of the current support strategy.

**200. B.** Chylothorax is a lymphatic pleural effusion that becomes milky after enteral fat intake and typically has elevated triglycerides with a lymphocyte-predominant cell count. It may be congenital or postoperative and can compromise ventilation and nutrition.