COMPREHENSIVE MOCK EXAM 10

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

1. A 34-week infant is delivered after the mother received indomethacin for several days because of threatened preterm labor. Echocardiography shortly after birth shows right-ventricular pressure overload without structural heart disease. Which fetal drug effect is most relevant?

A. Cyclooxygenase inhibition increases fetal pulmonary blood flow and causes left-ventricular volume overload.

B. Indomethacin opens the ductus arteriosus and produces a large fetal left-to-right shunt before birth.

C. Maternal indomethacin primarily causes fetal systemic vasodilation without affecting the ductal circulation.

D. Prostaglandin inhibition can constrict the fetal ductus arteriosus and increase right-ventricular afterload.

2. A 28-week infant is born in a cold delivery room. After stabilization the axillary temperature is 35.2 C and the blood glucose is falling. Which delivery-room strategy most directly prevents this complication in very preterm infants?

A. Use a warm environment with a plastic wrap or bag, cap, and other measures that limit evaporative heat loss.

B. Use cold sterile towels during resuscitation to reduce cerebral metabolism before neurologic assessment.

C. Dry the infant extensively before placing the infant uncovered beneath room-temperature ambient air.

D. Delay all thermal interventions until after vascular access is secured and the first blood gas is obtained.

3. A 2-day-old infant develops shock as the ductus closes. Femoral pulses are weak, the right-arm blood pressure is substantially higher than the leg pressure, and the liver is enlarged. Which lesion is most likely?

A. Mild pulmonary valve stenosis causing excess lower-body perfusion after normal ductal closure.

B. Critical coarctation of the aorta causing ductal-dependent systemic blood flow to the lower body.

C. Small muscular ventricular septal defect causing abrupt systemic hypoperfusion when pulmonary resistance falls.

D. A large secundum atrial septal defect causing isolated right-sided volume loading without systemic obstruction.

4. A 26-week infant has diffuse ground-glass opacities, air bronchograms, low lung volumes, and increasing oxygen need shortly after birth. Which pathophysiologic abnormality is central to this disease?

A. Excess surfactant lowers surface tension too far and causes pathologic alveolar overdistention at low pressure.

B. Surfactant deficiency raises alveolar surface tension and promotes end-expiratory alveolar collapse.

C. Primary pulmonary venous obstruction causes diffuse edema while leaving alveolar stability unaffected.

D. Delayed clearance of fetal lung fluid is the dominant mechanism in very preterm surfactant-deficient lung disease.

5. A neonate with duodenal atresia also has a complete atrioventricular canal and a transient neonatal megakaryoblastic proliferation. Which chromosomal diagnosis best unifies these findings?

A. Trisomy 21 (Down syndrome), a common association with complete atrioventricular septal defect.

B. Noonan syndrome, which can cause pulmonary stenosis.

C. Trisomy 18, which more often includes clenched hands with overlapping fingers and severe growth restriction.

D. Turner syndrome, which typically presents in a phenotypic female with lymphedema and left-sided heart disease.

6. Enteral feeding is about to begin for a 780-g infant whose parent has established a growing supply of expressed colostrum. Which source should form the foundation of feeding when sufficient volume is available?

A. Preterm formula should replace maternal milk during early feed advancement to provide more predictable nutrient delivery.

B. Parenteral nutrition should be the exclusive nutrient source until the infant reaches term-equivalent age even when the gut is usable.

C. Unfortified sterile water should replace milk during the first weeks because enteral nutrients increase intestinal oxygen demand.

D. Mother's own milk should be prioritized, with fortification as needed to meet the preterm infant's nutrient requirements.

7. A 38-week infant has serum creatinine 1.1 mg/dL immediately after delivery and 0.8 mg/dL at 24 hours, with normal urine output. The mother's creatinine at delivery was 1.0 mg/dL. Which interpretation is most appropriate?

A. The value suggests intrinsic neonatal renal dysfunction because the newborn creatinine should already be below the maternal concentration.

B. Early neonatal creatinine partly reflects maternal creatinine and must be interpreted by trend and gestational age.

C. A normal urine output excludes any influence of maternal renal function on the infant's early serum creatinine.

D. Creatinine should fall below adult values immediately after cord clamping in most healthy newborn regardless of gestation.

8. A 9-day-old infant is recalled after an abnormal newborn screen. Confirmatory serum testing shows TSH 92 mIU/L with a low free thyroxine concentration, although feeding and examination are normal. Which action is most important?

A. No treatment is needed if the newborn is asymptomatic because congenital hypothyroidism resolves spontaneously in most cases.

B. Delay therapy until clinical signs such as macroglossia and developmental delay appear because early treatment can mask diagnosis.

C. Treat with propylthiouracil to suppress neonatal thyroid hormone synthesis while waiting for repeat screening.

D. Begin levothyroxine promptly after confirmatory testing because early treatment protects neurodevelopment.

9. Newborn screening shows absent T-cell receptor excision circles, and a repeat blood count shows an absolute lymphocyte count of 700/µL. At 2 weeks the infant also has persistent oral candidiasis. Which disorder requires urgent evaluation?

A. Chronic granulomatous disease causing defective phagocyte oxidative burst with recurrent bacterial and fungal infections.

B. Transient physiologic neutrophilia of the newborn, which is not a primary immunodeficiency.

C. Severe combined immunodeficiency with major impairment of T-cell function and often B- or NK-cell abnormalities.

D. Selective IgA deficiency causing impaired mucosal antibody protection with recurrent respiratory and gastrointestinal infections.

10. A term newborn becomes lethargic and tachypneic at 8 hours of age after prolonged rupture of membranes. Blood culture later grows a gram-positive coccus that is beta-hemolytic. Which pathogen is most likely?

A. Coagulase-negative Staphylococcus causing a late central-line-associated bloodstream infection in a prolonged NICU stay.

B. Candida albicans causing disseminated fungal disease after weeks of broad-spectrum antibiotics and invasive devices.

C. Group B Streptococcus causing early-onset neonatal sepsis acquired around labor and delivery.

D. Herpes simplex virus causing vesicular skin lesions and encephalitis.

11. A 27-week infant on day 12 develops abdominal distention, bloody stools, temperature instability, and apnea. Abdominal radiograph shows pneumatosis intestinalis and portal venous gas. Which diagnosis is most likely?

A. Duodenal atresia, which presents soon after birth with proximal obstruction and a double-bubble pattern.

B. Spontaneous intestinal perforation, which more often presents with isolated perforation and less diffuse inflammatory bowel disease.

C. Hirschsprung disease, which causes distal functional obstruction but not acute pneumatosis and systemic inflammatory deterioration.

D. Necrotizing enterocolitis with intestinal inflammation and intramural gas formation.

12. Cord blood from an infant born to a mother with clinically significant red-cell alloantibodies shows anemia, reticulocytosis, and a positive direct antiglobulin test; bilirubin is already rising at 4 hours. Which process explains the bilirubin production?

A. Immune-mediated hemolysis is increasing bilirubin production faster than the newborn can conjugate and excrete it.

B. Breast milk jaundice is increasing enterohepatic circulation and causing severe anemia with a positive antiglobulin test.

C. Physiologic jaundice is causing immune destruction of red cells and is expected to be prominent during the first 12 hours.

D. Biliary atresia is blocking conjugated bilirubin excretion and causing isolated direct hyperbilirubinemia within hours of birth.

13. A newborn has a sharply demarcated violaceous reticulated patch on one leg with visible telangiectatic vessels. The pattern does not disappear with warming. Which diagnosis is most likely?

A. Physiologic cutis marmorata, which is temperature dependent and typically resolves with warming.

B. Cutis marmorata telangiectatica congenita, a congenital capillary vascular malformation pattern.

C. Erythema toxicum, which consists of transient erythematous papules and pustules.

D. Staphylococcal scalded skin syndrome, which causes tender erythema and superficial desquamation.

14. A 6-week-old exclusively breastfed infant who received no vitamin K at birth presents with seizures and a large intracranial hemorrhage. PT is markedly prolonged and platelet count is normal. Which diagnosis is most likely?

A. Disseminated intravascular coagulation, which should usually include thrombocytopenia, low fibrinogen, and systemic illness.

B. Hemophilia A, which is inherited and would primarily prolong aPTT.

C. Late vitamin K deficiency bleeding due to inadequate vitamin K-dependent clotting factor activity.

D. Neonatal alloimmune thrombocytopenia, which causes severe thrombocytopenia.

15. A 38-week infant is 4 hours old after a placental abruption and has moderate encephalopathy with abnormal tone, weak suck, and electrographic seizures. Which therapy should be initiated promptly if standard eligibility criteria are met?

A. Therapeutic hypothermia started within 6 hours of birth and continued under a specialized cooling protocol.

B. Routine therapeutic hypothermia for most infant born before 34 weeks regardless of neurologic examination.

C. Delay all cooling until MRI confirms basal ganglia injury because imaging is required before therapy can begin.

D. Systemic hyperthermia to 39 C for 24 hours to increase cerebral blood flow after hypoxic-ischemic injury.

16. A child born at 30 weeks' gestation is 5 months old by calendar age and is being evaluated for early developmental milestones. Which age should be used for the initial milestone comparison?

A. Use the full 5-month chronological age because correction for prematurity is not useful once an infant leaves the NICU.

B. Use a corrected age of about 2.5 months by subtracting the 10 weeks of prematurity from chronological age.

C. Use 30 weeks as a fixed developmental age until the child reaches one year of chronological age.

D. Add the 10 weeks of prematurity to chronological age, making the developmental age older than the calendar age.

17. A 27-week infant weighing 930 g at birth is clinically stable at 31 weeks postmenstrual age. What eye-care principle applies?

A. The infant requires scheduled retinopathy of prematurity screening based on gestational age, birth weight, and postmenstrual age.

B. primarily term infants with congenital cataracts require ophthalmologic screening; very preterm infants are not at risk for retinal neovascularization.

C. ROP screening is unnecessary when oxygen saturation targets were followed because appropriate oxygen management eliminates the disease.

D. Postpone retinal examination until near term because clinically important proliferative ROP develops after vascularization is nearly complete.

18. A very preterm infant receives a water-soluble medication. Compared with an older child, why might the dose per kilogram needed to achieve a similar initial plasma concentration be relatively larger?

A. All neonatal medications remain exclusively intravascular because immature tissues do not permit drug distribution outside plasma.

B. Neonatal protein binding is so high that hydrophilic drug volume of distribution becomes smaller than plasma volume.

C. Preterm infants have a larger extracellular and total body water fraction, increasing the apparent volume of distribution for hydrophilic drugs.

D. Preterm infants have much less body water, concentrating hydrophilic drugs in a smaller volume after each dose.

19. Parents of an infant at the margin of viability receive counseling from the obstetric and neonatal teams. Prognostic estimates have substantial uncertainty and both intensive care and comfort-focused care are ethically supportable. Which approach is best?

A. Give primarily the physician's preferred option because discussing uncertainty undermines parental trust and creates unnecessary choice.

B. Use shared decision-making that explains uncertainty, elicits family values, and develops a plan that can be revisited as the infant's condition evolves.

C. Require intensive care in most case because parental values have no role when survival is biologically possible.

D. Require comfort care in most case because uncertainty makes any trial of neonatal intensive care ethically inappropriate.

20. In a trial, severe BPD occurs in 18% of treated infants and 24% of controls. What is the relative risk of severe BPD in the treatment group compared with controls?

A. The relative risk is 6 because percentages must be subtracted.

B. The relative risk is 0.75 because 0.18 divided by 0.24 equals 0.75.

C. The relative risk is 0.06 because 24% minus 18% equals a six-percentage-point absolute risk reduction.

D. The relative risk is 1.33 because 24% divided by 18% should be used when treatment is the numerator group.

21. A 29-week infant on CPAP has worsening work of breathing and rising oxygen requirement from surfactant-deficient RDS but remains spontaneously breathing. Which approach can deliver surfactant while minimizing exposure to prolonged invasive ventilation?

A. Give surfactant by nebulizing sterile water into the incubator so medication reaches the alveoli without airway instrumentation.

B. Stop CPAP for several hours after surfactant because distending pressure inactivates the administered phospholipid.

C. Use a less-invasive surfactant administration technique through a thin catheter while maintaining spontaneous breathing when feasible.

D. Increase CPAP support first and defer surfactant until the infant requires intubation for refractory respiratory failure.

22. Prenatal studies showed severe fetal growth restriction, a ventricular septal defect, and persistent clenched hands. After birth the second and fifth fingers overlap adjacent digits and the feet have prominent heels. Which chromosomal disorder best fits this pattern?

A. Trisomy 18 (Edwards syndrome), classically associated with overlapping clenched fingers and severe growth restriction.

B. Trisomy 21, which usually presents with hypotonia and characteristic facial findings.

C. 45,X Turner syndrome, which is associated with webbed neck and neonatal lymphedema.

D. 47,XXY Klinefelter syndrome, which rarely causes a recognizable dysmorphic neonatal phenotype of this severity.

23. A VLBW infant is ready for enteral feeding, but the mother cannot provide sufficient milk. Which alternative is generally preferred over preterm formula when screened donor human milk is available?

A. Soy formula is preferred because plant protein eliminates the need for mineral supplementation in extremely preterm infants.

B. Cow's-milk-based term formula is preferred because donor milk has a higher native protein concentration than preterm formula.

C. Amino-acid-free carbohydrate solution is preferred because intact nutrients should be avoided until 34 weeks postmenstrual age.

D. Pasteurized donor human milk is generally preferred, particularly to reduce necrotizing enterocolitis risk.

24. A preterm infant develops oliguria after severe sepsis. Serum creatinine rises progressively over 48 hours. Which finding best supports acute kidney injury rather than normal postnatal adaptation?

A. A sustained rise in creatinine together with reduced urine output and clinical evidence of impaired renal function.

B. Transient weight loss during physiologic diuresis with improving creatinine and stable perfusion.

C. A low blood urea nitrogen concentration in an infant receiving limited protein intake but otherwise normal renal function.

D. A single creatinine value equal to the maternal level immediately after birth with normal urine output.

25. A 10-day-old infant with vomiting and weight loss has sodium 122 mEq/L, potassium 7.0 mEq/L, hypoglycemia, and ambiguous genitalia. Which diagnosis is most likely?

A. Salt-wasting congenital adrenal hyperplasia from 21-hydroxylase deficiency.

B. Congenital hypothyroidism causing isolated low thyroid hormone without hyperkalemic salt wasting.

C. Central diabetes insipidus causing hypernatremia and dilute urine.

D. Hyperinsulinism causing hypoglycemia but not androgen excess, hyperkalemia, or severe sodium loss.

26. A newborn with 22q11.2 deletion has a very small thymus and markedly reduced T-cell numbers. Which embryologic defect explains the immune problem?

A. Overgrowth of the thymic medulla traps mature T cells and causes isolated thrombocytopenia without infection risk.

B. Failure of splenic red pulp development selectively eliminates neutrophils while leaving T-cell maturation intact.

C. Abnormal development of the third and fourth pharyngeal pouches causes thymic hypoplasia or aplasia.

D. Absence of fetal bone marrow prevents all B-cell production.

27. A 25-week infant develops early-onset sepsis with shock and meningitis. Blood culture grows a lactose-fermenting gram-negative rod. Which organism is a leading consideration?

A. Staphylococcus epidermidis, a gram-positive organism more typical of late-onset line-associated sepsis.

B. Escherichia coli, an important cause of early-onset sepsis and meningitis in very preterm infants.

C. Treponema pallidum, a spirochete causing congenital syphilis.

D. Bordetella pertussis, which causes paroxysmal cough later in infancy.

28. At laparotomy for pneumoperitoneum in a 23-week infant, the surgeon finds a single small ileal perforation with otherwise viable bowel and no diffuse inflammatory or necrotic changes. Which diagnosis best fits?

A. Classic necrotizing enterocolitis, which requires diffuse pneumatosis.

B. Spontaneous intestinal perforation, a focal perforation that can occur in extremely preterm infants early in life.

C. Meconium ileus, which causes obstruction from inspissated meconium and usually presents before feeds with a characteristic distal bowel pattern.

D. Hirschsprung-associated enterocolitis, which generally occurs in a term infant with distal aganglionosis.

29. A jaundiced newborn has low serum albumin, acidosis, and sepsis. Why do these findings increase concern about bilirubin neurotoxicity at a given total serum bilirubin concentration?

A. They increase albumin binding affinity, making bilirubin less available to cross cell membranes during critical illness.

B. They convert unconjugated bilirubin into conjugated bilirubin, which crosses the blood-brain barrier more readily.

C. They prevent bilirubin from entering the bloodstream, so a measured total level underestimates hepatic excretion primarily.

D. They can increase the fraction of unbound bilirubin capable of crossing into brain tissue.

30. A newborn has a solitary well-demarcated absence of skin on the vertex of the scalp with a thin translucent membrane over the defect. Which diagnosis is most likely?

A. Transient neonatal pustular melanosis, which causes superficial pustules and hyperpigmented macules.

B. Epidermolysis bullosa, which causes trauma-induced blistering at multiple sites.

C. Sebaceous nevus, which is a congenital plaque of abnormal skin.

D. Aplasia cutis congenita, a focal congenital absence of skin that most often involves the scalp.

31. A well-appearing firstborn term infant has diffuse petechiae and a platelet count of 8,000/μL. The mother has a normal platelet count. Which mechanism best explains neonatal alloimmune thrombocytopenia?

A. Maternal autoantibodies against the mother's own platelets cross the placenta primarily when the mother has severe thrombocytopenia.

B. Maternal platelet autoantibodies causing immune thrombocytopenia in the newborn despite a currently normal maternal platelet count.

C. Maternal antibodies formed against a paternally derived fetal platelet antigen cross the placenta and destroy fetal platelets.

D. Placental insufficiency causes platelet sequestration in the fetal spleen without involvement of maternal IgG antibodies.

32. A 33-week infant has severe encephalopathy after perinatal asphyxia. Which statement best reflects current evidence regarding therapeutic hypothermia?

A. Use encephalopathy severity as the main criterion and cool at 33 weeks if biochemical evidence of asphyxia is present.

B. Routine cooling is not recommended at this gestational age because established benefit applies to more mature infants and potential harm is a concern.

C. Therapeutic hypothermia has proven equal benefit in all gestational ages down to 24 weeks and should be routine.

D. Cooling is contraindicated primarily in infants above 42 weeks because mature brains are more vulnerable to hypothermia.

33. A former 25-week infant has no obvious developmental delay on the day of NICU discharge despite prior severe BPD and a major intraventricular hemorrhage. Which postdischarge strategy is most appropriate because important impairments may emerge later?

A. Delay referral to early intervention until a permanent diagnosis of cerebral palsy or intellectual disability has been established.

B. Prioritize pulmonary and growth follow-up after discharge, adding developmental services if clear delays become evident later.

C. Use routine primary-care surveillance initially and reserve formal developmental testing for persistent concerns after age three years.

D. Arrange structured high-risk follow-up with developmental surveillance, early intervention, hearing and vision follow-up, and specialty care as indicated.

34. At 35 weeks postmenstrual age, retinal examination in a former 25-week infant meets criteria for treatment-requiring ROP. Which response is most appropriate?

A. Observe until term-corrected age because ROP requiring treatment usually regresses before retinal vascularization is complete.

B. Stop all oxygen immediately even if the infant becomes hypoxemic because oxygen withdrawal is the definitive treatment for advanced ROP.

C. Arrange prompt treatment by an experienced ophthalmologist because progression can threaten vision if therapy is delayed.

D. Begin systemic corticosteroids as sole therapy because retinal ablation or anti-VEGF treatment has no role in ROP.

35. A gentamicin trough remains higher than intended 24 hours after a dose in a 25-week infant despite no new drug exposure. Which developmental physiologic feature most contributes to this prolonged elimination?

A. Renal clearance is higher in very preterm infants than in older children, so long intervals are used to prevent subtherapeutic peaks.

B. Neonates metabolize aminoglycosides rapidly in the liver, so extended intervals are needed to allow hepatic enzyme regeneration.

C. Aminoglycosides are eliminated mainly through the lungs, and premature infants exhale the drug more slowly during apnea.

D. Immature glomerular filtration slows aminoglycoside clearance, so more time is needed between doses to avoid accumulation.

36. A mother with limited English proficiency must consent to surgery for her newborn. Her 14-year-old child offers to interpret. What is the best approach?

A. Use a qualified medical interpreter so the parent can receive accurate information and participate directly in consent.

B. Proceed without interpretation if the surgeon provides a written English consent form and speaks slowly.

C. Ask another patient's family member to interpret because any bilingual person provides the same confidentiality and accuracy as a trained interpreter.

D. Use the minor child as the primary interpreter because family members are generally more accurate with medical terminology.

37. Investigators compare prior antenatal antibiotic exposure in infants with NEC and matched infants without NEC. They report an odds ratio of 2.0. What does this study design primarily estimate?

A. A case-control study estimates the odds of prior exposure among cases compared with controls, producing an odds ratio.

B. A randomized trial estimates incidence directly because infants are assigned to exposure after NEC status is known.

C. A cross-sectional survey estimates primarily prevalence.

D. A prospective cohort begins by selecting infants based on NEC diagnosis and then reconstructs exposures retrospectively.

38. A newborn is growth restricted and has microcephaly and congenital heart disease. The mother has phenylketonuria and had persistently high phenylalanine concentrations throughout pregnancy. What best explains the infant's abnormalities?

A. Maternal phenylalanine causes primarily neonatal hypoglycemia.

B. The fetus must inherit two PAH variants before maternal phenylalanine can affect organ development.

C. Maternal phenylalanine is teratogenic to the fetus even when the fetus does not have phenylketonuria.

D. Fetal PAH genotype is the main determinant of risk because placental transport substantially limits fetal phenylalanine exposure.

39. A newborn remains apneic with a heart rate of 70/min after the initial steps. Positive-pressure ventilation is started, but the heart rate does not rise and there is no visible chest movement. What should the team do first?

A. Give intravenous epinephrine before correcting the mask seal because medication is the fastest way to raise heart rate.

B. Stop positive-pressure ventilation and observe for spontaneous recovery because chest movement is not required during mask ventilation.

C. Begin chest compressions immediately because any heart rate below 100/min requires compressions regardless of ventilation.

D. Perform ventilation corrective steps to establish visible chest movement before moving to chest compressions.

40. A profoundly cyanotic newborn with d-transposition of the great arteries remains hypoxemic despite prostaglandin infusion. Echocardiography shows a very restrictive interatrial communication. Which procedure most directly improves mixing?

A. Immediate closure of the ductus arteriosus to prevent recirculation of oxygenated blood into the pulmonary artery.

B. Pulmonary artery banding to reduce pulmonary blood flow before adequate mixing has been established.

C. Closure of the foramen ovale with a device to separate systemic and pulmonary venous return more substantially.

D. Balloon atrial septostomy to enlarge interatrial communication between the parallel circulations.

41. Prenatal imaging in a male fetus showed megacystis and bilateral hydroureteronephrosis. After birth, catheterization is difficult and voiding cystourethrography shows dilation of the posterior urethra. Which lesion is most likely?

A. Posterior urethral valves causing bladder outlet obstruction in a male infant.

B. Multicystic dysplastic kidney causing unilateral nonfunctioning renal tissue without lower urinary tract obstruction.

C. Ureteropelvic junction obstruction causing isolated upper-tract dilation without bladder outlet obstruction or weak stream.

D. Renal vein thrombosis causing hematuria and renal enlargement.

42. A large-for-gestational-age newborn has recurrent severe hypoglycemia despite an adequate glucose infusion. During hypoglycemia, ketones and free fatty acids are suppressed. Which mechanism is most likely?

A. Cortisol excess is blocking hepatic glucose production and causing marked ketotic hypoglycemia.

B. Central diabetes insipidus is causing urinary glucose loss and low ketone production despite normal insulin activity.

C. Growth hormone excess is increasing lipolysis and ketone production while lowering serum glucose.

D. Persistent hyperinsulinism is suppressing glycogenolysis, lipolysis, and ketone production.

43. A healthy term newborn has substantial serum IgG at birth despite having produced little immunoglobulin independently. What is the principal source?

A. Maternal IgM crosses the placenta freely and is the major immunoglobulin class in normal neonatal serum.

B. Fetal B cells synthesize adult concentrations of IgG by midgestation, making placental transfer unimportant.

C. Breast milk IgA enters the neonatal bloodstream intact and accounts for most circulating IgG measured at birth.

D. Maternal IgG crosses the placenta through active Fc-receptor-mediated transport, especially late in gestation.

44. A 9-day-old infant develops clusters of vesicles on an erythematous base and focal seizures. The mother had no known genital lesions at delivery. Which diagnosis requires urgent empiric treatment while testing is obtained?

A. Neonatal herpes simplex virus infection, including possible central nervous system disease.

B. Congenital candidiasis, which may cause rash.

C. Erythema toxicum neonatorum presenting with transient inflammatory papules and pustules during the early newborn period.

D. Transient neonatal pustular melanosis, which is present at birth and leaves hyperpigmented macules without encephalitis.

45. A term newborn has abdominal distention and fails to pass meconium. Contrast enema shows a microcolon with multiple filling defects in the terminal ileum. Which diagnosis should prompt evaluation for cystic fibrosis?

A. Meconium ileus caused by inspissated intestinal contents in the distal small bowel.

B. Duodenal atresia, which causes proximal obstruction and a double-bubble sign.

C. Pyloric stenosis, which presents weeks later with nonbilious projectile emesis and no neonatal microcolon.

D. Hirschsprung disease, which typically shows a distal narrow segment with proximal dilation.

46. A 3-week-old infant has conjugated hyperbilirubinemia and repeatedly acholic stools. The team is concerned about a disorder in which delay of definitive surgical drainage can worsen long-term outcome. Which diagnosis is the priority to exclude?

A. Physiologic jaundice, which is unconjugated and should not cause acholic stools or dark urine at five weeks.

B. Biliary atresia causing progressive extrahepatic bile duct obstruction and neonatal cholestasis.

C. Breast milk jaundice, which causes prolonged unconjugated hyperbilirubinemia in an otherwise well infant with pigmented stools.

D. Hemolytic disease, which raises unconjugated bilirubin and reticulocytes.

47. A newborn develops annular erythematous lesions on the face and scalp after light exposure. The mother is asymptomatic but has anti-Ro/SSA antibodies. Which diagnosis is most likely?

A. Incontinentia pigmenti, an X-linked disorder with staged skin lesions.

B. Erythema toxicum presenting as transient erythematous papules and pustules during the first postnatal days.

C. Cutaneous neonatal lupus from passively transferred maternal autoantibodies.

D. Congenital candidiasis, which can cause diffuse papules or pustules.

48. A pale term newborn has hemoglobin 6 g/dL and a high reticulocyte count after placental pathology suggests a massive fetomaternal hemorrhage. Which compensatory finding is most consistent with a chronic rather than hyperacute blood loss?

A. Marked polycythemia is expected because chronic fetal hemorrhage stimulates maternal red-cell transfer across the placenta.

B. Reticulocytosis and erythroblastosis indicate that fetal marrow had time to respond before delivery.

C. A normal hemoglobin concentration is required because chronic blood loss is fully compensated by fetal erythropoiesis.

D. Complete absence of reticulocytes strongly suggests chronic blood loss because erythropoiesis shuts down when anemia develops slowly.

49. A term infant has electrographic seizures after hypoxic-ischemic injury despite normal glucose, calcium, and sodium. Which medication remains a common first-line antiseizure therapy in the neonatal period?

A. Oral carbamazepine is generally first line for any neonatal seizure regardless of etiology and route availability.

B. Continuous midazolam infusion as initial therapy because rapid onset can suppress electrographic neonatal seizures.

C. Phenobarbital is commonly used as initial treatment for acute neonatal seizures.

D. Phenytoin is contraindicated in all neonates and has no role as additional therapy when seizures persist.

50. A former extremely preterm toddler walks on toes with scissoring, increased lower-extremity tone, and relatively preserved upper-extremity function. Which cerebral palsy pattern is most consistent?

A. Ataxic cerebral palsy, which is characterized primarily by impaired balance and coordination.

B. Spastic diplegia with greater motor involvement of the legs than the arms.

C. Spastic hemiplegia, which primarily affects one side of the body.

D. Dyskinetic cerebral palsy, which is dominated by involuntary movements and variable tone.

51. At 3 weeks of age, an infant with an untreated dense cataract in one eye has developed a strong fixation preference for the fellow eye. What is the principal reason treatment of the visual-axis obstruction is time-sensitive?

A. A unilateral cataract affects primarily cosmetic appearance because the fellow eye prevents any developmental loss in the affected visual pathway.

B. Removing the cataract after adolescence produces the same visual outcome because visual cortical development is not time sensitive.

C. Prolonged deprivation of patterned vision can cause irreversible amblyopia during a critical period of visual development.

D. Congenital cataracts reliably clear without intervention once fetal lens proteins are replaced during infancy.

52. A neonate with hyperbilirubinemia is receiving several highly protein-bound medications. Which pharmacologic interaction can increase bilirubin neurotoxicity risk?

A. Increasing albumin binding generally raises free bilirubin because bound bilirubin crosses the blood-brain barrier more readily.

B. Protein displacement matters primarily for conjugated bilirubin, which is the principal cause of kernicterus in newborns.

C. A drug that displaces bilirubin from albumin can raise the unbound bilirubin fraction even if total bilirubin changes little.

D. Highly protein-bound drugs reduce free bilirubin by occupying albumin sites and stabilizing bilirubin binding in plasma.

53. A critically ill newborn has progressive multiorgan failure despite maximal therapy. The parents and clinical team agree that continued invasive treatment no longer achieves the goals they value for the infant. Which ethical action is permissible?

A. Redirect care toward comfort and withdraw burdensome life-sustaining treatments when this is consistent with the infant's best interests and agreed goals.

B. Require court review for most decision to withdraw ventilation even when the parents and treating team agree on the care plan.

C. Continue most available treatment indefinitely because withdrawing an intervention is ethically different from rarely starting it.

D. Stop analgesia once life-sustaining treatment is withdrawn because symptom relief might unintentionally shorten life.

54. A diagnostic test has a sensitivity of 90% and specificity of 95%. Disease prevalence becomes much lower in the screened population. Which test characteristic will usually decrease most?

A. Specificity necessarily rises to 100% whenever prevalence becomes low, regardless of test performance.

B. Negative predictive value decreases sharply because rare disease makes a negative result less reassuring.

C. Positive predictive value decreases because a larger proportion of positive results will be false positives when disease is rare.

D. Sensitivity necessarily falls because sensitivity is mathematically determined by disease prevalence in the screened population.

55. A term infant has severe oligohydramnios sequence with limb-position abnormalities, pulmonary hypoplasia, and renal failure. The mother took an angiotensin-converting enzyme inhibitor throughout the second and third trimesters. Which mechanism is most likely?

A. The medication stimulated fetal aldosterone secretion and caused polyuria with excessive amniotic fluid loss.

B. The drug caused isolated placental vasodilation while preserving fetal renal blood flow and urine output.

C. Blockade of the fetal renin-angiotensin system impaired renal perfusion and fetal urine production.

D. ACE inhibition increased fetal nephron filtration and produced obstructive uropathy from bladder overdistention.

56. After effective ventilation through an advanced airway, a newborn's heart rate remains 45/min. Chest compressions are begun. Which compression-to-ventilation ratio is used in standard neonatal resuscitation?

A. Continuous compressions at 100/min with ventilations withheld until the heart rate exceeds 60/min.

B. Three compressions followed by one ventilation, coordinated in a 3:1 ratio.

C. Thirty compressions followed by two ventilations, matching the standard adult single-rescuer sequence.

D. Fifteen compressions followed by two ventilations, matching the usual two-rescuer pediatric sequence.

57. A neonate has a large inlet ventricular septal defect, a primum atrial septal defect, and a common atrioventricular valve. Which chromosomal condition is strongly associated with this cardiac lesion?

A. Trisomy 13, which classically produces isolated pulmonary valve stenosis without septal defects.

B. Trisomy 21, which is strongly associated with complete atrioventricular septal defect.

C. Klinefelter syndrome, which commonly presents in the newborn period with complete atrioventricular block.

D. Turner syndrome, which is most strongly linked to aortic coarctation and bicuspid aortic valve.

58. A term infant delivered by elective cesarean section without labor develops tachypnea within an hour of birth. The chest film shows prominent central vascular markings, fluid in the fissures, and mild hyperinflation. Which diagnosis is most likely?

A. Surfactant-deficient respiratory distress syndrome causing low lung volumes and diffuse air bronchograms in a term infant.

B. Congenital diaphragmatic hernia causing bowel-filled hemithorax and severe mediastinal displacement.

C. Pulmonary interstitial emphysema causing linear and cystic lucencies along bronchovascular bundles.

D. Transient tachypnea of the newborn from delayed clearance of fetal lung fluid.

59. A newborn has cleft lip and palate, postaxial polydactyly, microphthalmia, and holoprosencephaly. Which chromosomal abnormality is most likely?

A. Trisomy 13 (Patau syndrome), associated with holoprosencephaly, facial clefts, and postaxial polydactyly.

B. 22q11.2 deletion, which is associated with conotruncal heart disease and hypocalcemia.

C. Trisomy 18, which is more strongly associated with overlapping clenched fingers and rocker-bottom feet.

D. Turner syndrome, which causes lymphedema and coarctation.

60. A 26-week infant is receiving full volumes of unfortified human milk but has poor weight gain and low serum phosphorus with rising alkaline phosphatase. Which nutritional modification is most appropriate?

A. Fortify human milk to increase protein, calcium, phosphorus, and other nutrient delivery for preterm growth.

B. Stop enteral feeds and provide glucose-primarily intravenous fluid until alkaline phosphatase normalizes.

C. Switch to unfortified term breast milk exclusively because term milk contains more calcium and phosphorus than preterm fortifier.

D. Dilute the milk with water to lower mineral intake because phosphorus deficiency is caused by excessive fortification.

61. A growth-restricted newborn fails the hearing screen and has thrombocytopenia. Cranial ultrasonography shows echogenic foci concentrated around the lateral ventricles. Which congenital infection is the leading concern?

A. Congenital toxoplasmosis, which more classically causes diffuse intracranial calcifications with hydrocephalus and chorioretinitis.

B. Congenital rubella, which is strongly associated with cataracts, PDA, and sensorineural hearing loss.

C. Congenital cytomegalovirus infection, which classically can produce microcephaly and periventricular calcifications.

D. Congenital syphilis, which can cause hepatosplenomegaly but not the classic CMV pattern of periventricular calcification and microcephaly.

62. Contrast enema in a newborn with persistent distal obstruction shows a narrow rectum and distal colon with an abrupt transition to dilated proximal bowel. Which disease most likely causes this pattern?

A. Necrotizing enterocolitis, which usually affects preterm infants with systemic illness and pneumatosis.

B. Meconium ileus, which is associated with cystic fibrosis.

C. Pyloric stenosis, which causes nonbilious vomiting after several weeks.

D. Hirschsprung disease due to congenital absence of enteric ganglion cells in the distal bowel.

63. During intensive blue-light therapy, a newborn's unconjugated bilirubin falls even though hepatic bilirubin-conjugating capacity remains immature. Which photochemical effect accounts for the treatment benefit?

A. Light directly increases UDP-glucuronosyltransferase gene expression enough to fully mature hepatic conjugation within minutes.

B. Phototherapy converts conjugated bilirubin back to unconjugated bilirubin so it can diffuse into the intestinal lumen.

C. Blue-spectrum light converts bilirubin into more water-soluble photoisomers that can be excreted without hepatic conjugation.

D. Phototherapy induces red-cell destruction so bilirubin can be cleared through renal filtration of hemoglobin fragments.

64. A female newborn develops linear vesicles on the limbs following the lines of Blaschko. Over time the lesions are expected to become verrucous and then hyperpigmented. Which disorder is most likely?

A. Epidermolysis bullosa simplex, which causes friction-induced blisters without a staged Blaschko-line evolution.

B. Transient neonatal pustular melanosis, which resolves rapidly leaving superficial hyperpigmented macules without verrucous stages.

C. Staphylococcal scalded skin syndrome, an acquired toxin-mediated illness with diffuse tenderness and desquamation.

D. Incontinentia pigmenti, an X-linked dominant disorder caused by pathogenic IKBKG variants.

65. A newborn with RhD alloimmune hemolytic disease has severe anemia and rapidly rising bilirubin despite intensive phototherapy. Which therapy can simultaneously remove antibody-coated cells and bilirubin while correcting anemia?

A. Phototherapy alone, which directly increases hemoglobin concentration and removes circulating anti-D antibodies.

B. Simple iron supplementation, which corrects hemolysis immediately and removes circulating maternal antibody within minutes.

C. Platelet transfusion, which treats red-cell antibody disease by replacing bilirubin-binding proteins in the plasma.

D. Exchange transfusion when severe hemolysis and hyperbilirubinemia meet treatment criteria.

66. A newborn has recurrent episodes of generalized stiffness and apnea triggered by unexpected touch or sound. Between episodes the neurologic examination is notable for an exaggerated startle response, and EEG during attacks shows no ictal activity. Which diagnosis is most likely?

A. Hereditary hyperekplexia from impaired glycinergic inhibition, producing exaggerated startle and stimulus-induced stiffness.

B. Benign neonatal sleep myoclonus, in which jerks occur during sleep and stop with arousal rather than being triggered by startle.

C. Neonatal epilepsy, which would be supported by an ictal electrographic correlate during the stereotyped events.

D. Jitteriness from hypoglycemia, which is tremulous, stimulus sensitive, and suppressible rather than causing sustained generalized rigidity.

67. A school-aged child born at 25 weeks has normal gross motor function but struggles with attention, processing speed, executive function, and mathematics. What does this illustrate?

A. Normal early motor development is expected to provide normal later executive function because all prematurity-related brain effects present by age two.

B. Neurodevelopmental sequelae of extreme prematurity can emerge as higher-order cognitive and behavioral demands increase with age.

C. Unrecognized sensorineural hearing loss is the most likely explanation for later academic difficulty in former preterm children.

D. Prematurity-related brain injury primarily affects motor pathways, making isolated executive-function impairment less likely.

68. A NICU infant who required prolonged intensive care needs newborn hearing screening before discharge. Which technique is especially useful for detecting auditory neuropathy in this population?

A. Automated auditory brainstem response assesses neural conduction as well as peripheral auditory function.

B. Tympanometry alone is sufficient because normal middle-ear pressure excludes cochlear and neural hearing loss.

C. Otoacoustic emissions alone are preferred because they directly measure auditory nerve conduction through the brainstem.

D. Behavioral audiometry is the standard newborn test because a neonate can reliably signal hearing thresholds by hand raising.

69. A newborn receives repeated doses of a medication that undergoes hepatic glucuronidation and then develops unexpectedly high drug concentrations. Which developmental principle is most relevant?

A. Hepatic metabolism is absent until six months of age, so no neonatal medication can undergo any liver biotransformation.

B. Renal tubular secretion fully compensates for immature hepatic metabolism for most glucuronidated medication.

C. Immature hepatic conjugation pathways can reduce metabolic clearance of some drugs during the neonatal period.

D. Neonates have uniformly greater glucuronidation capacity than adults, so drug accumulation must have another explanation.

70. A NICU finds that infants from families living far from the hospital have substantially higher missed follow-up rates. Which quality and equity response is most appropriate?

A. Identify practical barriers with affected families and design supports such as transportation, telehealth, scheduling flexibility, or care coordination.

B. Label the families noncompliant and discharge them from follow-up because structural barriers are outside the health system's responsibility.

C. Use the same clinic schedule and transportation assumptions for most family because identical processes is expected to provide equitable access.

D. Stop collecting follow-up data by neighborhood because identifying disparities can itself create inequity even when services differ.

71. A randomized trial is designed with 80% power. What does power represent?

A. The probability of detecting a specified true effect when that effect actually exists, given the design assumptions.

B. The proportion of enrolled infants who must complete follow-up for randomization to remain valid.

C. The probability that any statistically significant result is definitely clinically important and free of bias.

D. The probability of rejecting the null hypothesis when the null hypothesis is actually true, which is the type I error rate.

72. A 37-week newborn has marked hypotonia, a weak cry, and shallow respirations immediately after delivery. The mother received repeated magnesium sulfate boluses during labor for severe preeclampsia. Which explanation is most likely?

A. Maternal magnesium is excluded by the placenta, making a medication effect physiologically implausible.

B. Magnesium causes neonatal catecholamine excess, leading primarily to hypertension and hyperreflexia.

C. Transplacental magnesium can transiently depress neonatal neuromuscular and respiratory function.

D. Magnesium exposure triggers neonatal acetylcholine excess and produces sustained spasticity after birth.

73. A newborn has severe bradycardia despite effective ventilation, coordinated compressions, and epinephrine. The history suggests acute fetal blood loss from a ruptured vasa previa, and the infant is pale with weak pulses. What additional intervention is appropriate?

A. Give a large dose of diuretic to reduce ventricular preload because neonatal bradycardia usually reflects volume overload.

B. Withhold intravascular fluid because volume expansion is contraindicated during any neonatal resuscitation.

C. Give sodium bicarbonate as the first treatment for presumed blood loss before restoring circulating volume.

D. Give volume expansion with isotonic crystalloid or blood when hypovolemia or blood loss is strongly suspected.

74. A neonate receiving prostaglandin E1 for a ductal-dependent heart lesion develops recurrent apnea shortly after the infusion is increased. Which interpretation is most appropriate?

A. Congenital central hypoventilation syndrome should be suspected as the principal cause of recurrent apnea during the infusion.

B. Apnea strongly suggests the ductus has closed and requires immediate discontinuation of prostaglandin before reassessment.

C. Prostaglandin causes primarily systemic hypertension.

D. Apnea is a recognized prostaglandin adverse effect, so airway support may be needed while ductal patency is maintained.

75. A post-term infant with meconium aspiration has patchy atelectasis alternating with hyperinflated regions. Which mechanism best explains the hyperinflation?

A. Complete obstruction of most affected airway increases distal ventilation and lowers the risk of air leak.

B. Meconium inhibits fetal lung-fluid absorption primarily, producing a radiographic pattern identical to uncomplicated TTN.

C. Partial small-airway obstruction can create a ball-valve effect with air trapping distal to meconium plugs.

D. Meconium uniformly dilates the bronchi and eliminates expiratory resistance throughout the entire lung.

76. A phenotypic female newborn has puffy hands and feet, a broad shield chest, low posterior hairline, and a coarctation of the aorta. Which karyotype is most likely?

A. 47,XX,+21 causing Down syndrome with atrioventricular septal defect and generalized hypotonia.

B. 45,X Turner syndrome, consistent with neonatal lymphedema and a left-sided obstructive cardiac lesion.

C. 47,XY,+18 causing Edwards syndrome with clenched hands and severe fetal growth restriction.

D. 46,XX with an FMR1 expansion causing fragile X syndrome and neonatal lymphedema.

77. A growth-restricted extremely preterm infant begins parenteral nutrition with substantial amino acid and energy delivery. Within two days the infant develops hypophosphatemia and hypokalemia. Which mechanism best explains the pattern?

A. An anabolic insulin response drives phosphorus and potassium into cells when nutrition is advanced after chronic fetal undernutrition.

B. High protein intake suppresses insulin substantially, forcing phosphate and potassium out of cells into the plasma.

C. Parenteral amino acids directly destroy renal tubules, causing obligatory phosphate wasting in most premature infant.

D. Respiratory alkalosis from early respiratory support is the principal cause of the potassium and phosphorus decline.

78. A newborn with severe oligohydramnios from prolonged bilateral renal dysfunction has profound respiratory failure despite relatively normal cardiac structure. Which prenatal consequence is most responsible?

A. Renal dysfunction increased fetal surfactant secretion, leading to abnormally small but highly compliant lungs.

B. Maternal creatinine crossed the placenta and directly destroyed type II pneumocytes in an otherwise normally expanded lung.

C. Excess amniotic fluid compressed the fetal thorax and prevented pulmonary vascular development.

D. Inadequate amniotic fluid impaired fetal lung expansion and caused pulmonary hypoplasia.

79. A 2-day-old infant of a diabetic mother develops jitteriness. Serum calcium is low and magnesium is normal. Which endocrine disturbance is a recognized contributor in this setting?

A. Parathyroid hormone excess causes renal calcium wasting and is the universal mechanism of early hypocalcemia in these infants.

B. Maternal insulin crosses the placenta and directly binds neonatal calcium, lowering the ionized fraction after birth.

C. Transient functional hypoparathyroidism can contribute to early neonatal hypocalcemia in infants of diabetic mothers.

D. Neonatal calcitonin deficiency causes calcium deposition in bone and is the primary cause of postnatal hypocalcemia.

80. A newborn has a positive pathogen-specific IgM test. Why can neonatal IgM be useful as evidence of congenital infection?

A. Breast-milk IgM is absorbed intact into the circulation and is the usual source of pathogen-specific serum IgM at birth.

B. IgM does not cross the placenta efficiently, so pathogen-specific IgM generally reflects fetal antibody production.

C. Maternal IgM crosses the placenta more efficiently than IgG, so any neonatal IgM simply mirrors maternal exposure.

D. IgM is produced primarily after six months of age, so a neonatal result must represent laboratory contamination.

81. A group O mother has a group A newborn who develops jaundice and mild anemia in the first day. Which feature makes ABO hemolytic disease possible even in a first pregnancy?

A. Naturally occurring maternal IgG anti-A or anti-B antibodies may already be present and can cross the placenta.

B. primarily maternal IgM anti-A antibodies cross the placenta, making first-pregnancy disease uniquely severe.

C. ABO hemolysis generally requires sensitization in a prior pregnancy, similar to the mechanism of RhD alloimmunization.

D. Fetal A and B antigen expression is too weak in a first pregnancy to permit clinically meaningful antibody-mediated hemolysis.

82. A 26-week infant has cranial ultrasound showing echogenicity that later evolves into cystic lesions in the periventricular white matter. Which injury is most likely?

A. Subdural hemorrhage from birth trauma, which occurs over the cerebral convexities.

B. Hypoxic injury limited to the basal ganglia and thalami, which is more typical of acute profound asphyxia in term infants.

C. Congenital hydrocephalus, which enlarges the ventricles.

D. Cystic periventricular leukomalacia, a severe preterm white-matter injury adjacent to the lateral ventricles.

83. A 20-month-old former VLBW infant has a concerning developmental screen but an unremarkable physical examination. What is the best next step?

A. Delay evaluation until kindergarten because standardized developmental assessment is not useful before school age.

B. Repeat the same screening test annually and take no action until the child loses previously acquired milestones.

C. Assume the screen is false positive because normal tone excludes language, cognitive, social, or fine-motor delay.

D. Perform or arrange formal developmental evaluation and initiate appropriate early-intervention referral without waiting for obvious neurologic signs.

84. A newborn develops cyanosis during feeding and quiet breathing but becomes pink while crying. A small catheter cannot be passed through either nostril. Which diagnosis is most likely?

A. Bilateral choanal atresia causing obstruction in a newborn who preferentially breathes through the nose.

B. Laryngomalacia with dynamic supraglottic collapse producing inspiratory stridor that worsens with feeding.

C. Tracheoesophageal fistula producing cough, choking, and recurrent aspiration during oral feeding.

D. Pierre Robin sequence with micrognathia and glossoptosis causing positional upper-airway obstruction.

85. A study uses a two-sided alpha of 0.05. What does the alpha level represent?

A. The probability of a type II error, failing to detect a true effect, which is determined primarily by the P value after analysis.

B. The probability that the null hypothesis is true whenever a study produces P greater than 0.05.

C. The prespecified probability of a type I error, rejecting the null hypothesis when it is actually true.

D. The fraction of participants expected to cross over between treatment groups during a randomized trial.

86. A fetus develops hydrops with cardiomegaly and severe anemia after the mother experiences a mild febrile illness and arthralgias during pregnancy. Which maternal infection most directly causes this pattern by suppressing fetal erythropoiesis?

A. Maternal varicella causes fetal hydrops mainly through chronic placental transfusion and hyperviscosity.

B. Maternal rubella primarily causes fetal anemia through immune destruction of mature red blood cells.

C. Congenital cytomegalovirus typically causes hydrops by inducing fetal insulin excess and polycythemia.

D. Parvovirus B19 infection can destroy erythroid precursors and produce profound fetal anemia.

87. A term infant born through meconium-stained fluid is limp and apneic. The airway is not visibly obstructed. What is the priority rather than routine tracheal suctioning?

A. Perform prolonged oropharyngeal suctioning until the fluid is substantially clear before assessing heart rate.

B. Delay positive-pressure ventilation for several minutes because meconium exposure makes ventilation dangerous.

C. Start the standard initial steps and provide effective ventilation promptly if the infant remains apneic or bradycardic.

D. Intubate most nonvigorous infant primarily to suction meconium before any attempt at ventilation.

88. A preterm infant has a large PDA with left atrial enlargement and worsening pulmonary edema. Serum creatinine is rising and urine output is poor. Which pharmacologic choice is less attractive because of its renal prostaglandin effects?

A. Acetaminophen is contraindicated primarily because it generally causes acute renal vasoconstriction in preterm infants.

B. Indomethacin may worsen renal perfusion and requires caution when significant renal dysfunction is present.

C. Caffeine must be avoided because it directly closes the ductus and precipitates systemic hypoperfusion.

D. Inhaled nitric oxide is the standard first-line drug for ductal closure and is limited mainly by renal toxicity.

89. A term newborn has recurrent hypoventilation most pronounced during sleep despite normal lung imaging, a normal neuromuscular examination, and no sedating drug exposure. Carbon dioxide rises substantially during sleep, and PHOX2B testing is planned. Which diagnosis is most likely?

A. Transient tachypnea of the newborn from delayed fetal lung-fluid clearance.

B. Congenital central hypoventilation syndrome associated with abnormal autonomic respiratory control.

C. Apnea of prematurity from immaturity of respiratory control in an extremely preterm infant.

D. Upper-airway obstruction from bilateral choanal atresia.

90. A neonate has conotruncal heart disease, hypocalcemic seizures, and recurrent infections with a small thymic shadow. Which diagnosis best unifies the findings?

A. Prader-Willi syndrome with severe hypotonia and poor feeding followed later by hyperphagia.

B. Williams syndrome with supravalvar aortic stenosis and infantile hypercalcemia but normal thymic development.

C. Beckwith-Wiedemann syndrome with macroglossia, organomegaly, and neonatal hyperinsulinemic hypoglycemia.

D. 22q11.2 deletion syndrome with thymic and parathyroid hypoplasia.

91. A 750-g infant cannot tolerate adequate enteral volumes during the first days of life. Which component should be included early in parenteral nutrition to limit cumulative protein deficit?

A. Provide amino acids early while monitoring metabolic tolerance and adjusting total nutrient delivery to the infant's condition.

B. Delay parenteral amino acids until enteral feeds advance because early protein may worsen metabolic acidosis in extremely preterm infants.

C. Provide primarily dextrose for the first two weeks because nitrogen intake worsens most episode of neonatal acidosis.

D. Delay protein until birth weight has doubled because early amino acids prevent normal postnatal adaptation.

92. A preterm infant has serum sodium of 124 mEq/L, low serum osmolality, concentrated urine, and no signs of sodium depletion after intracranial hemorrhage. Which mechanism is most likely?

A. Hyperaldosteronism causing sodium retention, hypertension, and hypernatremia.

B. Central diabetes insipidus causing free-water loss, dilute urine, and progressive hypernatremia.

C. Renal salt wasting with contracted extracellular volume and maximally dilute urine from suppressed antidiuretic hormone.

D. Inappropriate antidiuretic hormone activity causing water retention and dilutional hyponatremia.

93. A preterm infant becomes hypothermic after prolonged exposure during a procedure. Soon afterward the infant is hypoglycemic and develops metabolic acidosis. Which physiologic process links cold stress to these findings?

A. Neonates respond primarily with sustained skeletal-muscle shivering that generates heat without affecting glucose metabolism.

B. Hypothermia directly increases surfactant production and therefore causes acidosis primarily through excessive lung compliance.

C. Cold exposure suppresses brown-fat metabolism substantially, reducing both glucose and oxygen use below baseline levels.

D. Nonshivering thermogenesis increases oxygen and glucose consumption, and severe cold stress can outstrip metabolic supply.

94. A male infant has eczema, recurrent infections, and thrombocytopenia with unusually small platelets. Which immunodeficiency is most likely?

A. Chronic granulomatous disease, which causes defective oxidative burst.

B. Wiskott-Aldrich syndrome caused by an X-linked defect affecting cytoskeletal function in immune and platelet cells.

C. Severe combined immunodeficiency, which can cause infection.

D. DiGeorge syndrome, which may cause T-cell deficiency but is associated with thymic hypoplasia and hypocalcemia.

95. A preterm infant is delivered after the mother had a febrile influenza-like gastrointestinal illness. The newborn has early sepsis, pneumonia, meningitis, and scattered granulomatous skin lesions. Which vertically acquired pathogen is most likely?

A. Group B Streptococcus acquired during delivery.

B. Congenital cytomegalovirus infection.

C. Listeria monocytogenes acquired transplacentally or during birth.

D. Neonatal herpes simplex virus infection.

96. A 2-day-old infant has sudden bilious emesis, abdominal pain, and rapidly worsening perfusion. Upper gastrointestinal imaging shows a corkscrew configuration of proximal small bowel. Which diagnosis is the surgical emergency?

A. Gastroesophageal reflux with transient gastroduodenal reflux causing recurrent bilious-appearing emesis in early infancy.

B. Physiologic meconium retention, which can delay stool.

C. Midgut volvulus due to intestinal malrotation with twisting around the mesenteric pedicle.

D. Pyloric stenosis, which produces nonbilious vomiting and usually presents later without bowel ischemia.

97. A NICU graduate with meningitis passed the newborn hearing screen but later has delayed speech. Which follow-up principle is most appropriate?

A. Risk factors such as meningitis warrant ongoing hearing surveillance because delayed-onset or progressive hearing loss can occur.

B. A passed newborn screen permanently excludes hearing impairment, so no later audiologic evaluation is needed even with speech delay.

C. primarily infants who failed both ears on the newborn screen can develop sensorineural hearing loss after hospital discharge.

D. Speech delay after meningitis should be attributed to cognition alone because auditory injury rarely progresses after the neonatal period.

98. A neonate has micrognathia, glossoptosis, a U-shaped cleft palate, and positional upper-airway obstruction. Which diagnosis best describes the sequence?

A. Bilateral vocal cord paralysis causing persistent inspiratory stridor and impaired glottic airway opening.

B. Pierre Robin sequence resulting from mandibular hypoplasia with posterior tongue displacement and cleft palate.

C. CHARGE syndrome with choanal atresia, ear anomalies, and associated cranial nerve dysfunction.

D. Treacher Collins syndrome with mandibular hypoplasia and other characteristic craniofacial abnormalities.

99. NICUs rather than individual infants are randomized to use one of two infection-prevention bundles. What design issue is especially important in analysis?

A. Cluster randomization eliminates confounding substantially because most infant receives both infection-prevention bundles.

B. Randomizing hospitals converts the trial into a case-control study and prevents any causal inference about the intervention.

C. Outcomes within the same NICU may be correlated, so clustering must be accounted for in sample-size planning and analysis.

D. Individual outcomes are generally statistically independent once a hospital has been randomized, so no clustering adjustment is needed.

100. A 35-week infant is pale and hypotensive after an otherwise uncomplicated vaginal birth. The placenta appears normal. Maternal blood testing shows a large quantity of fetal hemoglobin-containing cells. Which diagnosis best explains the presentation?

A. A significant fetomaternal hemorrhage caused acute or subacute fetal blood loss before delivery.

B. Placental insufficiency produced neonatal polycythemia through chronic fetal hypoxemia and increased erythropoiesis.

C. Maternal thrombocytopenia caused isolated fetal platelet loss without a reduction in neonatal red-cell mass.

D. Delayed cord clamping caused excessive placental transfusion followed by acute intravascular hemolysis.

101. During resuscitation, the team wants the most rapid reliable continuous assessment of heart rate while positive-pressure ventilation is underway. Which method is most useful?

A. Measure noninvasive blood pressure repeatedly because cuff pressure changes provide the fastest heart-rate estimate.

B. Rely primarily on intermittent palpation of the umbilical pulse because electronic monitoring is slower and less accurate.

C. Use a chest radiograph after each ventilation cycle to estimate cardiac activity from mediastinal motion.

D. Apply electrocardiographic leads while continuing clinical and pulse-oximetry assessment.

102. A newborn with tetralogy of Fallot becomes acutely more cyanotic after agitation. The murmur becomes softer during the episode. Which physiologic change is driving the spell?

A. Sudden pulmonary vasodilation diverts nearly all right-ventricular output toward the lungs and away from the aorta.

B. A transient rise in systemic vascular resistance increases right-to-left shunting and is therefore the usual initiating event.

C. Acute closure of the VSD eliminates intracardiac mixing and forces all blood through the pulmonary artery.

D. Increased right-to-left shunting across the VSD because dynamic right-ventricular outflow obstruction has worsened.

103. A ventilated extremely preterm infant develops branching and cystic radiolucencies that follow the pulmonary interstitium after exposure to high airway pressures. Which diagnosis is most likely?

A. Transient tachypnea from delayed fetal lung-fluid clearance producing fluid-filled.

B. Pulmonary interstitial emphysema from air dissecting out of overdistended alveoli into the interstitium.

C. Pulmonary lymphangiectasia causing primarily homogeneous low lung volumes without cystic radiolucencies.

D. Chylothorax producing bilateral pleural fluid collections with compression of the lungs.

104. A newborn with neonatal hyperinsulinism has asymmetric overgrowth of one leg, macroglossia, and an umbilical wall defect. Which imprinting disorder should prompt later embryonal-tumor surveillance?

A. Smith-Lemli-Opitz syndrome caused by impaired cholesterol synthesis and typically associated with growth restriction.

B. Prader-Willi syndrome caused by absent paternal expression in 15q11-q13 and characterized by neonatal hypotonia.

C. Beckwith-Wiedemann syndrome caused by dysregulation of imprinted genes on chromosome 11p15.

D. Turner syndrome caused by X chromosome monosomy and characterized by short stature.

105. A preterm infant on prolonged parenteral nutrition receives almost no intravenous lipid because of concern about hyperglycemia. Several weeks later the infant has dermatitis and poor growth. Which deficiency should be considered?

A. Copper overload because intravenous lipid is the main route for eliminating trace elements from the bloodstream.

B. Essential fatty acid deficiency from inadequate linoleic and alpha-linolenic acid delivery.

C. Essential amino acid deficiency caused specifically by lack of triglyceride infusion despite adequate parenteral protein.

D. Vitamin B12 excess caused by lipid restriction and accumulation of methylmalonic acid in adipose tissue.

106. A term infant after severe hypoxic brain injury develops polyuria, rising serum sodium, high serum osmolality, and very dilute urine. Which diagnosis is most likely?

A. SIADH causing water retention, low serum sodium, and concentrated urine despite low serum osmolality.

B. Congenital nephrotic syndrome causing heavy proteinuria and edema.

C. Central diabetes insipidus from impaired antidiuretic hormone secretion.

D. Posterior urethral valves causing obstructive oliguria and a distended bladder.

107. A newborn becomes lethargic and jaundiced after milk feeds begin. Blood culture grows gram-negative bacteria, and the newborn screen is abnormal for galactose metabolism. Which disease is most likely?

A. Congenital hypothyroidism presenting with prolonged jaundice, poor feeding, and temperature instability after newborn feeds begin.

B. Maple syrup urine disease, which causes branched-chain amino acid accumulation.

C. Medium-chain acyl-CoA dehydrogenase deficiency, which causes hypoketotic hypoglycemia during fasting.

D. Classic galactosemia from galactose-1-phosphate uridyltransferase deficiency causing lactose-triggered systemic illness.

108. An extremely preterm infant has lower serum IgG than a term infant of the same postnatal age. Which developmental fact best explains the difference?

A. Term infants synthesize adult amounts of IgG independently before delivery, whereas preterm infants make no immunoglobulin.

B. Most placental IgG transfer occurs late in gestation, so very preterm infants receive less passive maternal antibody.

C. Breastfeeding raises serum IgG by direct intestinal absorption, and term infants generally receive more breast milk than preterm infants.

D. Preterm infants destroy maternal IgG immediately after birth because fetal Fc receptors are absent from the placenta.

109. A newborn has snuffles, a diffuse maculopapular rash involving palms and soles, hepatosplenomegaly, and radiographic metaphyseal abnormalities. Which diagnosis is most likely?

A. Congenital rubella, which is more strongly associated with cataracts, hearing loss, and patent ductus arteriosus.

B. Neonatal HSV, which typically causes vesicular lesions and may cause encephalitis or disseminated hepatitis.

C. Congenital CMV, which can cause petechiae and microcephaly but not the classic snuffles and palm-sole rash pattern.

D. Early congenital syphilis caused by transplacental Treponema pallidum infection.

110. Prenatal imaging showed polyhydramnios and a persistently small stomach. After birth, a feeding tube stops in a blind upper esophageal pouch, while radiographs show air in the stomach and intestines. Which anatomic configuration is most likely?

A. Duodenal atresia, which allows an orogastric tube into the stomach and causes a double-bubble pattern.

B. Pure esophageal atresia without fistula, which would usually produce a gasless abdomen because no distal airway-bowel connection exists.

C. H-type tracheoesophageal fistula, in which the esophagus is continuous and the tube should pass to the stomach.

D. Esophageal atresia with a distal tracheoesophageal fistula, producing a proximal pouch and gas in the abdomen.

111. A preterm infant has a gradually falling hemoglobin at 5 weeks, low reticulocyte count, stable bilirubin, and no evidence of bleeding. Which physiologic factor contributes to anemia of prematurity?

A. Physiologic hemolysis is caused by maternal anti-D antibody in nearly most preterm infant regardless of blood type.

B. Iron overload from placental transfusion is the primary universal cause of low reticulocyte production in premature infants.

C. A relatively low endogenous erythropoietin response is inadequate for the degree of anemia in very preterm infants.

D. Excess erythropoietin secretion suppresses marrow red-cell production and causes reticulocytopenia during prematurity.

112. A 24-week infant has a grade IV intraventricular hemorrhage on cranial ultrasound. What does the parenchymal echodense lesion adjacent to the ventricle most likely represent?

A. Periventricular hemorrhagic infarction from impaired venous drainage rather than simple extension of blood into brain tissue.

B. Normal germinal matrix maturation that appears echogenic and has no relationship to hemorrhage or later injury.

C. A congenital cortical malformation caused by neuronal migration failure during the first trimester.

D. Perinatal arterial ischemic stroke involving a deep perforator territory adjacent to the lateral ventricle.

113. A time-to-extubation study ends before some infants have been extubated. How are those infants commonly handled in survival analysis?

A. They are assigned the median extubation time of the group to avoid any missing event times.

B. Classify infants without extubation as treatment failures at the study end and analyze the observed event times separately.

C. They are removed from the dataset largely so their observed ventilator time contributes no information.

D. They are censored at their last known follow-up time, contributing information up to that point without an observed event.

114. A pregnant patient with systemic lupus erythematosus has anti-Ro/SSA antibodies. Fetal echocardiography at 24 weeks shows persistent bradycardia with atrioventricular dissociation. What is the underlying mechanism?

A. Maternal beta-blocker exposure causing persistent fetal sinus bradycardia with reduced atrioventricular conduction.

B. Placental transfer of maternal T lymphocytes directly replaced fetal conduction tissue with scar tissue.

C. Maternal IgM antibodies crossed the placenta and transiently blocked the fetal sinoatrial node without tissue injury.

D. Maternal autoantibodies crossed the placenta and injured the fetal atrioventricular conduction system.

115. A 31-week infant is breathing spontaneously but has labored respirations and cyanosis with a heart rate of 150/min. Which support best fits the physiology before invasive ventilation is considered?

A. Provide continuous positive airway pressure with oxygen titrated to target saturations while the infant breathes spontaneously.

B. Begin chest compressions because respiratory distress alone indicates inadequate cardiac output despite a normal heart rate.

C. Intubate immediately for most preterm infant with any oxygen requirement, even when spontaneous breathing is effective.

D. Withhold distending pressure because CPAP is useful primarily after an infant has first received endotracheal ventilation.

116. A neonate with hypoplastic left heart syndrome is stable on prostaglandin. A clinician increases inspired oxygen substantially despite acceptable saturations, after which systemic perfusion worsens. What is the best physiologic explanation?

A. Oxygen converts the ductus into a fixed left-to-right shunt that improves systemic perfusion but raises coronary oxygen demand.

B. Higher oxygen tension closes the atrial septum and prevents any pulmonary venous blood from entering the systemic circulation.

C. Lower pulmonary vascular resistance can steal cardiac output toward the lungs and away from the systemic circulation.

D. Supplemental oxygen directly constricts the pulmonary arteries, causing an abrupt fall in pulmonary blood flow and left atrial pressure.

117. Lung histology from a former extremely preterm infant would be expected to show simplified alveoli and abnormal pulmonary vascular development after prolonged respiratory support. Which chronic neonatal lung disorder is being described?

A. Primary surfactant protein deficiency that is defined by spontaneous resolution before 36 weeks postmenstrual age.

B. Congenital lobar overinflation involving most lung lobe symmetrically after prolonged oxygen exposure.

C. Transient tachypnea of the newborn persisting for months because fetal lung fluid has rarely been absorbed.

D. Bronchopulmonary dysplasia related to disrupted lung development after extreme prematurity and respiratory injury.

118. A newborn has profound hypotonia and poor suck. Genetic testing shows loss of paternally expressed genes in chromosome region 15q11-q13. Which diagnosis is most likely?

A. Prader-Willi syndrome, which often presents in neonates with hypotonia and feeding difficulty.

B. Angelman syndrome, which results from loss of maternal expression in the same region and typically presents later with severe developmental impairment.

C. Beckwith-Wiedemann syndrome, an overgrowth disorder involving chromosome 11p15 imprinting.

D. Noonan syndrome, which is caused by RAS-MAPK pathway variants.

119. A 28-week infant is tolerating small-volume enteral feeds without abdominal distention, bloody stool, or cardiorespiratory instability. What is the main purpose of continuing minimal enteral nutrition while parenteral nutrition supplies most calories?

A. Small trophic feeds support gastrointestinal maturation and feeding tolerance while larger volumes are advanced cautiously.

B. Small enteral volumes sterilize the intestine and therefore eliminate the possibility of necrotizing enterocolitis.

C. Trophic feeds provide all required calories and protein even when primarily a few milliliters per kilogram are given each day.

D. Minimal feeds are used to intentionally keep the bowel inactive and reduce mucosal hormone release during prematurity.

120. A 25-week infant has serum potassium of 6.8 mEq/L on day 2 despite urine output exceeding 3 mL/kg/h and no evidence of hemolysis. Which phenomenon is most likely?

A. Central diabetes insipidus, which causes hypernatremia and dilute urine.

B. Nonoliguric hyperkalemia of extreme prematurity from immature cellular and renal potassium handling.

C. Physiologic hyperaldosteronism, which reduces renal potassium excretion and is universal in healthy term infants.

D. Obstructive renal failure from posterior urethral valves, which should cause marked oliguria and bladder distention.

121. A 5-day-old infant has poor feeding, alternating hypotonia and hypertonia, and a sweet odor to the urine. Plasma amino acids show markedly elevated leucine. Which disorder is most likely?

A. Urea-cycle disease, which causes hyperammonemia without the characteristic branched-chain amino acid accumulation and odor.

B. Galactosemia, which causes liver disease after milk exposure.

C. Phenylketonuria with phenylalanine accumulation causing neonatal neurologic dysfunction and an abnormal body odor.

D. Maple syrup urine disease from deficient branched-chain alpha-ketoacid dehydrogenase activity.

122. On day 2, a newborn develops rapidly progressive hyperpurulent conjunctivitis with marked eyelid edema after delivery to a mother with untreated gonorrhea. Which infection is most likely?

A. Chlamydia trachomatis conjunctivitis.

B. Neonatal herpes simplex keratoconjunctivitis.

C. Neisseria gonorrhoeae ophthalmia neonatorum.

D. Chemical conjunctivitis from ocular prophylaxis.

123. A newborn with trisomy 21 develops bilious vomiting within hours of birth. Abdominal radiograph shows a dilated stomach and proximal duodenum with no distal bowel gas. Which diagnosis is most likely?

A. Duodenal atresia causing complete proximal intestinal obstruction and the classic double-bubble appearance.

B. Pyloric stenosis, which is acquired after birth and causes nonbilious vomiting.

C. Necrotizing enterocolitis, which typically causes intestinal distention, pneumatosis, and systemic illness.

D. Hirschsprung disease, which usually causes distal colonic obstruction.

124. A critically ill newborn with septic shock develops simultaneous microvascular thrombosis and bleeding from line sites. Laboratory studies show falling platelets and fibrinogen with prolonged coagulation times and markedly increased fibrin-degradation products. Which process is occurring?

A. Isolated vitamin K deficiency, which should prolong coagulation tests.

B. Hemophilia B, which selectively reduces factor IX and usually leaves platelet count, PT, and fibrinogen normal.

C. Disseminated intravascular coagulation with systemic activation and consumption of platelets and coagulation factors.

D. Neonatal alloimmune thrombocytopenia, which causes severe platelet loss but should not consume fibrinogen or prolong both PT and aPTT.

125. Two hours after vacuum extraction, a newborn's head circumference has increased rapidly, hematocrit is falling, and a diffuse fluctuant scalp collection extends across the entire occiput without respecting sutures. Which lesion is the source of concern?

A. Subgaleal hemorrhage into the large potential space beneath the scalp aponeurosis.

B. Molding of the skull, which changes head shape.

C. Cephalohematoma, which is confined by periosteal attachments.

D. Caput succedaneum, which may cross sutures but is superficial edema.

126. A newborn required prolonged positive-pressure ventilation and chest compressions before circulation was restored. Which metabolic problem should be specifically monitored during the postresuscitation period because it can worsen neurologic injury?

A. Serum uric acid should be deliberately raised because hyperuricemia protects the brain after hypoxic-ischemic injury.

B. Postresuscitation glucose measurement is unnecessary unless the newborn is an infant of a diabetic mother.

C. Abnormal blood glucose, including hypoglycemia, should be detected and corrected during postresuscitation care.

D. Transient physiologic hyperbilirubinemia is the primarily metabolic abnormality that requires immediate neurologic monitoring after resuscitation.

127. A term infant is cyanotic and in severe respiratory distress. Echocardiography shows all pulmonary veins joining a confluence that drains below the diaphragm, with marked obstruction. Which consequence is expected?

A. Marked systemic hypertension because pulmonary venous obstruction increases left-ventricular preload and stroke volume.

B. Minimal respiratory symptoms because anomalous pulmonary venous return is well tolerated when the drainage pathway is obstructed.

C. Pulmonary venous hypertension with pulmonary edema and severe hypoxemia requiring urgent intervention.

D. Isolated bradycardia without pulmonary edema because venous obstruction affects primarily the cardiac conduction system.

128. A 28-week infant continues to have central apneic pauses with desaturation after secondary causes have been excluded. Which medication is used to improve respiratory drive and reduce these events?

A. Long-term systemic corticosteroids primarily to mature the central respiratory rhythm generator.

B. Caffeine citrate to stimulate respiratory drive and reduce clinically significant apnea.

C. Routine opioid infusion to suppress arousal and prevent desaturation-associated agitation.

D. Daily antihistamine therapy to reduce upper-airway secretions and increase respiratory effort.

129. A male infant with a normal chromosome count has fetal nuchal edema, widely spaced eyes, a broad chest, and dysplastic pulmonary valve stenosis. Which diagnosis is most likely?

A. Down syndrome, which is associated more strongly with atrioventricular septal defect than pulmonary valve stenosis.

B. Trisomy 13, which typically produces severe midline defects and polydactyly.

C. Noonan syndrome due to a disorder of the RAS-MAPK signaling pathway.

D. Turner syndrome, which requires an X chromosome abnormality and occurs in phenotypic females.

130. A stable preterm infant has several gastric residuals but a soft abdomen, normal stools, and no systemic illness. Which principle is most appropriate when assessing feeding intolerance?

A. Stop all feeds permanently after any residual because gastric contents are a specific diagnostic test for NEC.

B. Advance feeds because repeated residuals indicate slow gastric transit that may improve with larger, less frequent boluses.

C. Begin broad-spectrum antibiotics for seven days whenever residual milk is seen in a feeding tube of a preterm infant.

D. Interpret residuals together with the overall examination and clinical trajectory rather than treating residual volume alone as NEC.

131. A preterm infant receives prolonged high-dose furosemide and later has microscopic hematuria. Renal ultrasonography shows echogenic medullary deposits. Which complication is most likely?

A. Posterior urethral valves caused by medication-induced urethral membrane formation after birth.

B. Renal cortical necrosis caused by loop diuretics selectively thrombosing both renal arteries in a stable infant.

C. Autosomal recessive polycystic kidney disease caused directly by postnatal furosemide exposure.

D. Nephrocalcinosis related to increased urinary calcium losses and other prematurity-associated risk factors.

132. A 3-day-old male infant develops vomiting, lethargy, respiratory alkalosis, and rapidly rising plasma ammonia with relatively normal glucose. Which category of disorder should be suspected first?

A. A urea-cycle defect causing impaired nitrogen disposal and severe hyperammonemia.

B. Distal renal tubular acidosis, which causes hyperchloremic metabolic acidosis.

C. Galactosemia, which primarily produces hepatic dysfunction and sepsis risk after milk exposure.

D. Congenital hyperinsulinism, which produces hypoglycemia with suppressed ketones.

133. A pregnant woman develops varicella rash five days before delivery. Her newborn appears well at birth. Why is this timing concerning?

A. Varicella at this timing causes primarily congenital limb hypoplasia.

B. Maternal infection shortly before birth is expected to provide complete neonatal protection because maternal IgM crosses the placenta rapidly.

C. The infant can develop severe neonatal varicella because maternal infection occurred too close to delivery for adequate protective antibody transfer.

D. The infant is protected because maternal varicella-specific IgG rises rapidly enough in the days immediately before delivery.

134. Prenatal ultrasonography shows free-floating bowel adjacent to the umbilical cord insertion without a membrane-covered sac. At delivery the exposed loops emerge through a small right paraumbilical defect. Which lesion is present?

A. Bladder exstrophy, which exposes urinary bladder mucosa.

B. Gastroschisis, in which exposed bowel herniates through an abdominal wall defect without a membrane.

C. Omphalocele, which is a midline defect at the umbilical ring with herniated viscera covered by a sac.

D. Umbilical hernia, which is skin covered.

135. A male newborn develops prolonged bleeding after circumcision. Platelets and PT are normal, but aPTT is markedly prolonged. Maternal uncle had a similar disorder. Which diagnosis is most likely?

A. Neonatal alloimmune thrombocytopenia, which would cause a very low platelet count.

B. Hemophilia A from factor VIII deficiency inherited in an X-linked pattern.

C. Disseminated intravascular coagulation, which would usually affect platelets, fibrinogen, PT, and aPTT in a sick infant.

D. Vitamin K deficiency, which more often prolongs PT because factor VII has a short half-life and is vitamin K dependent.

136. Following a difficult shoulder delivery, a newborn has an asymmetric Moro response. Shoulder abduction and elbow flexion are weak, but wrist and finger grasp remain strong. Which brachial plexus pattern best explains the examination?

A. Lower trunk injury involving C8-T1 roots, which more strongly affects hand and finger function than shoulder abduction.

B. Isolated radial nerve injury, which causes wrist drop but not the classic shoulder and elbow posture with preserved grasp.

C. Upper trunk brachial plexus injury involving C5-C6 roots, producing the characteristic Erb palsy posture.

D. Spinal cord transection, which would produce broader bilateral neurologic deficits.

137. A therapy appears to reduce BPD in infants below 26 weeks but not in infants born at 28 to 30 weeks. The treatment-by-gestational-age interaction is statistically significant. What does this suggest?

A. The subgroup pattern is most likely residual confounding by gestational age despite random treatment assignment.

B. Gestational age may modify the treatment effect, so the magnitude of benefit differs across gestational-age strata.

C. Gestational age is necessarily a confounder because any subgroup difference is evidence that randomization failed.

D. The treatment has no overall effect because significant interaction means both subgroup effects must equal zero.

138. A woman uses cocaine shortly before developing painful vaginal bleeding and an emergency delivery at 33 weeks. The placenta shows retroplacental hemorrhage. Which maternal drug effect most likely contributed?

A. Catecholamine-mediated vasoconstriction increased the risk of placental abruption and fetal hypoxia.

B. The principal placental effect is nitric-oxide excess with profound maternal and fetal vasodilation.

C. Cocaine selectively suppresses fetal insulin secretion, causing placental hemorrhage through neonatal hypoglycemia.

D. Cocaine produces sustained uterine relaxation that prevents placental separation but causes post-term pregnancy.

139. A newborn has coloboma, congenital heart disease, choanal atresia, growth restriction, genital hypoplasia, and abnormal ears. Which syndrome is suggested by this combination?

A. VACTERL association, which requires a different pattern centered on vertebral, anal, cardiac, tracheoesophageal, renal, and limb anomalies.

B. CHARGE syndrome, often related to CHD7 variants and characterized by coloboma, choanal atresia, and ear anomalies.

C. Beckwith-Wiedemann syndrome, which is characterized by overgrowth and macroglossia.

D. Trisomy 18, which may cause multiple anomalies.

140. A former 25-week infant has persistently low serum phosphorus, high alkaline phosphatase, and poor linear growth despite adequate calories. Which condition is most likely?

A. Hyperparathyroid bone disease caused by universal neonatal calcium excess in infants fed fortified human milk.

B. Metabolic bone disease of prematurity related to inadequate mineral accretion and other risk factors.

C. Physiologic osteopetrosis of prematurity caused by excessive fetal calcium transfer during the third trimester.

D. Scurvy caused by routine vitamin C fortification in human milk, which blocks collagen formation in the neonatal skeleton.

141. A neonate has normal-anion-gap metabolic acidosis, hyperchloremia, urine pH persistently above 5.5, and nephrocalcinosis. Which renal tubular disorder is suggested?

A. Central diabetes insipidus, which causes water-balance abnormalities.

B. Nephrotic syndrome, which causes protein loss and edema without a primary defect in distal hydrogen ion secretion.

C. Distal renal tubular acidosis with impaired distal hydrogen ion secretion and inappropriately alkaline urine.

D. Proximal renal tubular acidosis with intact distal acidification and a persistently very low urine pH during systemic acidosis.

142. An infant becomes profoundly hypoglycemic during an intercurrent illness after a period of poor intake. Ketones are unexpectedly low. Which metabolic pathway defect should be considered?

A. Glycogen storage disease type I, which should generally produce abundant ketones during severe hypoglycemia.

B. A fatty-acid oxidation disorder causing hypoketotic hypoglycemia during fasting stress.

C. Classic galactosemia, which is triggered primarily by fasting and rarely by milk exposure.

D. Congenital adrenal hyperplasia, which causes isolated hyperketonemia without electrolyte disturbances or cortisol deficiency.

143. A 2-week-old infant develops fever, myocarditis, hepatitis, coagulopathy, and shock during a community outbreak of hand-foot-and-mouth disease. Which viral category should be considered?

A. Adenovirus rarely causes systemic neonatal disease and is limited to mild conjunctivitis in the first month of life.

B. Respiratory syncytial virus is the classic cause of neonatal hepatitis with profound coagulopathy and myocarditis.

C. Rotavirus commonly causes fulminant neonatal myocarditis while sparing the gastrointestinal tract.

D. Enterovirus infection can cause severe neonatal sepsis-like disease with myocarditis and hepatitis.

144. A newborn with early nonimmune hemolytic jaundice has a high mean corpuscular hemoglobin concentration, spherocytes, a negative direct antiglobulin test, and reduced eosin-5-maleimide binding on red cells. Which diagnosis is most likely?

A. G6PD deficiency, which is associated with oxidant-triggered bite cells and Heinz bodies.

B. Alpha thalassemia trait, which causes microcytosis.

C. ABO hemolytic disease, which can produce spherocytes but would more often have evidence of maternal antibody on testing.

D. Hereditary spherocytosis from a red-cell membrane cytoskeletal defect causing nonimmune hemolysis and spherocytes.

145. A newborn has intractable seizures beginning hours after birth. Standard antiseizure medications have limited effect. Seizures stop dramatically after pyridoxine is administered. Which disorder is suggested?

A. Benign familial neonatal seizures, which are defined by a predictable response primarily to glucose infusion.

B. Pyridoxine-dependent epilepsy due to a vitamin B6-responsive metabolic seizure disorder.

C. Hypocalcemic tetany, which is confirmed by vitamin B6 response even when serum ionized calcium is normal.

D. Neonatal abstinence syndrome, which causes tremor and irritability.

146. A receiver-operating-characteristic curve is constructed for a continuous sepsis biomarker. What does each point on the curve represent as the decision threshold changes?

A. A pair of sensitivity and false-positive rate values produced by a particular test threshold.

B. A comparison of treatment effect against sample size used to determine the number needed to treat.

C. A pair of disease prevalence and incidence values that remains fixed regardless of the biomarker threshold.

D. A direct plot of positive predictive value against negative predictive value that is independent of sensitivity and specificity.

147. A newborn is small for gestational age with microcephaly and a proportionately reduced length and weight. Early pregnancy was complicated by a major congenital infection. Which growth pattern is most consistent with an early global fetal insult?

A. Symmetric growth restriction affecting head circumference, length, and weight to a similar degree.

B. Selective macrosomia with increased fat mass and disproportionately large abdominal circumference.

C. Normal somatic growth with isolated postnatal loss of subcutaneous tissue from neonatal diuresis.

D. Asymmetric growth restriction with preserved head growth and isolated reduction in abdominal circumference.

148. A 3-week-old infant has tachypnea, sweating with feeds, hepatomegaly, and poor weight gain. Echocardiography shows a large unrestricted ventricular septal defect. Why did heart-failure symptoms become more prominent after the first week of life?

A. Pulmonary vascular resistance normally rises sharply after birth, driving more blood from the left ventricle into the lungs.

B. Closure of the foramen ovale converts the VSD into a right-to-left shunt that causes pulmonary venous congestion.

C. Falling pulmonary vascular resistance increased the magnitude of the left-to-right shunt and pulmonary overcirculation.

D. The neonatal myocardium becomes hypertrophic during the first weeks of life, increasing systemic vascular resistance enough to close the defect.

149. A delivery-room checklist is prepared for an infant with a large left congenital diaphragmatic hernia. Which proposed respiratory maneuver should be removed because it can inflate intrathoracic bowel and further compress the hypoplastic lung?

A. Assess preductal oxygenation and manage pulmonary hypertension while preparing for definitive care.

B. Place an orogastric tube for continuous decompression of the stomach and bowel.

C. Secure an endotracheal airway and use gentle ventilation tailored to avoid excessive pressures.

D. Avoid routine bag-mask ventilation because it can distend intrathoracic bowel and worsen lung compression.

150. A sick preterm infant has persistent hypotension, hyponatremia, hyperkalemia, and low cortisol during severe stress. Which endocrine problem should be considered if the findings do not respond to volume and routine support?

A. Hyperparathyroidism causes vasopressor-resistant shock by lowering cortisol synthesis and producing severe sodium loss.

B. Central diabetes insipidus typically causes hyponatremia and hyperkalemia while retaining free water during shock.

C. Adrenal insufficiency may contribute to catecholamine-resistant hypotension and electrolyte abnormalities.

D. Hyperthyroidism is the most likely cause because excess thyroid hormone directly produces hyperkalemic salt wasting in preterm infants.

151. A 23-week infant with a central venous catheter develops late-onset sepsis after three weeks in the NICU. Blood cultures repeatedly grow coagulase-negative staphylococci. What is the most likely source pattern?

A. A catheter-associated bloodstream infection from skin commensal organisms in a highly vulnerable preterm infant.

B. Transplacental congenital infection acquired during the first trimester and remaining latent primarily in the central line.

C. A classic early-onset infection acquired during labor that predictably remains culture-negative until the third week.

D. Foodborne infection from pasteurized donor milk because coagulase-negative staphylococci primarily invade through intestinal toxins.

152. A newborn with Down syndrome has leukocytosis and circulating megakaryoblasts but is otherwise stable. The blast count falls spontaneously over the next month. Which condition is most likely?

A. Congenital cytomegalovirus infection, which produces leukopenia.

B. Transient abnormal myelopoiesis associated with trisomy 21 and acquired GATA1 mutations.

C. Acute lymphoblastic leukemia, which characteristically resolves spontaneously in neonates with trisomy 21 without therapy.

D. Physiologic neonatal leukocytosis, which rarely includes a clonal population of megakaryoblasts or GATA1-associated disease.

153. A newborn is profoundly hypotonic but alert, with weak cry, tongue fasciculations, absent reflexes, and progressive respiratory weakness. Which diagnosis should be considered?

A. Spinal muscular atrophy due to loss of anterior horn cells, often from biallelic SMN1 variants.

B. Congenital myotonic dystrophy, which can cause hypotonia but is suggested by maternal myotonia.

C. Neonatal hyperthyroidism, which generally causes irritability, tachycardia, and hyperreflexia.

D. Central hypoxic-ischemic encephalopathy, which usually causes impaired alertness and may produce hyperreflexia.

154. A NICU tracks central-line infections monthly. After a new sterile-access bundle, the run chart shows eight consecutive points below the previous median. How should this pattern be interpreted?

A. The pattern strongly suggests causality beyond doubt because any eight-point shift can occur primarily after an effective intervention.

B. A randomized trial is required before a sustained run-chart shift can be interpreted as evidence of process change.

C. A sustained shift suggests special-cause change rather than ordinary random fluctuation and should prompt investigation of the new process.

D. The correct response is to recalculate the median after most data point, which prevents detection of sustained shifts over time.

155. A term infant is born to a mother who received no intrapartum antibiotic prophylaxis despite known group B streptococcal colonization. Which neonatal outcome is the prophylaxis designed primarily to reduce?

A. Early-onset group B streptococcal disease acquired around the time of labor and delivery.

B. Neonatal candidemia related to prolonged central venous catheter use in an intensive care unit.

C. Late-onset group B streptococcal meningitis acquired several weeks after birth from community exposure.

D. Congenital cytomegalovirus infection acquired transplacentally during the first trimester of pregnancy.

156. A neonate has a widely split first heart sound, severe tricuspid regurgitation, massive right atrial enlargement, and apical displacement of the septal tricuspid leaflet. Which diagnosis is most likely?

A. Tetralogy of Fallot with anterior malalignment of the ventricular septum and infundibular pulmonary stenosis.

B. Truncus arteriosus with a single semilunar valve supplying systemic, pulmonary, and coronary circulations.

C. Ebstein anomaly with atrialization of a portion of the right ventricle.

D. Complete atrioventricular septal defect with a common atrioventricular valve centered at the crux of the heart.

157. A term infant with severe parenchymal lung disease remains profoundly hypoxemic. Echocardiography shows a structurally normal heart with elevated pulmonary artery pressure and right-to-left shunting. Which pulmonary vasodilator can selectively reduce pulmonary vascular resistance in an appropriately ventilated infant?

A. Inhaled nitric oxide can selectively vasodilate ventilated pulmonary vascular beds in term or near-term PPHN.

B. Oral indomethacin is the preferred selective pulmonary vasodilator because prostaglandin inhibition lowers pulmonary resistance.

C. Intravenous phenylephrine directly relaxes pulmonary arterial smooth muscle while lowering systemic vascular resistance.

D. Nebulized epinephrine is the established first-line treatment for persistent pulmonary hypertension without airway disease.

158. A newborn has an anorectal malformation and tracheoesophageal fistula; imaging also shows vertebral, renal, cardiac, and radial-ray defects. No single-gene syndrome better explains the combination. Which diagnostic association is most appropriate?

A. Prader-Willi syndrome because hypotonia and feeding problems are expected with this congenital malformation pattern.

B. CHARGE syndrome because vertebral and radial anomalies are the defining features of CHD7-related disease.

C. Noonan syndrome because anorectal and tracheoesophageal anomalies are central RASopathy manifestations.

D. VACTERL association when multiple component anomalies occur without a unifying alternative syndrome.

159. An exclusively human-milk-fed preterm infant is approaching discharge. Which micronutrient deserves planned supplementation because human milk alone usually does not provide enough to meet the needs of a rapidly growing preterm infant?

A. Vitamin K should be avoided after birth because prophylactic vitamin K prevents later iron absorption.

B. Iodine should be given in pharmacologic doses to most preterm infant regardless of diet or thyroid status.

C. Iron supplementation is commonly required to support erythropoiesis and prevent iron deficiency.

D. Copper should routinely be withheld because human milk generally supplies toxic concentrations to preterm infants.

160. A neonate develops gross hematuria, thrombocytopenia, and a flank mass. Renal ultrasound shows an enlarged kidney with absent venous flow. Which diagnosis is most likely?

A. Posterior urethral valves, which cause bilateral obstruction and bladder distention.

B. Multicystic dysplastic kidney, which is congenital and typically appears as multiple noncommunicating cysts.

C. Central diabetes insipidus, which causes polyuria and hypernatremia without renal enlargement or thrombocytopenia.

D. Renal vein thrombosis, a neonatal vascular complication associated with dehydration and other prothrombotic states.

161. A neonate has exaggerated startle responses to sound and touch followed by generalized stiffness, but consciousness is preserved and EEG during events is normal. Which diagnosis is most likely?

A. Neonatal tetanus, which requires toxin exposure and typically causes persistent rigidity and spasms.

B. Hyperekplexia, a disorder of inhibitory glycinergic neurotransmission causing pathologic startle and stiffness.

C. Epileptic tonic seizures, which should consistently show ictal EEG changes during the stiffening episodes.

D. Benign neonatal jitteriness, which is stimulus sensitive but consists of tremulous movements.

162. A mother has anti-Kell alloantibodies and the fetus develops severe anemia. Compared with RhD alloimmunization, which additional mechanism is particularly important with Kell disease?

A. The Kell antigen is present primarily on maternal erythrocytes, so fetal anemia results from placental blood loss alone.

B. Antibodies can suppress fetal erythroid progenitor production in addition to destroying circulating red cells.

C. Kell disease causes fetal anemia by blocking iron transport across the placenta without immune red-cell effects.

D. Kell antibodies mainly increase fetal platelet destruction and rarely affect red-cell production or survival.

163. A stable neonate has a regular narrow-complex tachycardia at 250/min. Vagal maneuvers are unsuccessful, intravenous access is present, and the rhythm is consistent with reentrant supraventricular tachycardia. What is the next treatment?

A. Administer rapid intravenous adenosine while continuous rhythm monitoring is in place.

B. Start oral digoxin and wait several hours for conversion because acute therapy is unnecessary in sustained neonatal SVT.

C. Give atropine to slow conduction through the atrioventricular node and terminate the reentry circuit.

D. Deliver unsynchronized defibrillation at high energy despite stable perfusion and a regular narrow-complex rhythm.

164. Soon after a marked increase in pulmonary blood flow, an extremely preterm ventilated infant has abrupt respiratory failure, pink-bloody fluid filling the endotracheal tube, a falling hematocrit, and diffuse bilateral alveolar opacification. Which complication is most likely?

A. Pulmonary hemorrhage, often associated with extreme prematurity and increased pulmonary blood flow.

B. Transient tachypnea, which occurs at birth and is not characterized by sudden pulmonary bleeding in a ventilated preterm infant.

C. Uncomplicated apnea of prematurity, which produces bradycardia without blood in the airway or new alveolar infiltrates.

D. Congenital lobar emphysema, which produces focal hyperinflation.

165. Prenatal imaging documented recurrent long-bone and rib fractures without evidence of trauma. After birth the infant has blue-gray sclerae and generalized low bone density, and a parent reports multiple childhood fractures. Which disorder is most likely?

A. Hypophosphatasia caused by excess alkaline phosphatase activity with dense, overmineralized bones.

B. Thanatophoric dysplasia characterized by long limbs and a normally sized thorax without severe skeletal deformity.

C. Achondroplasia caused by activating FGFR3 variants and characterized by rhizomelic shortening without diffuse fractures.

D. Osteogenesis imperfecta caused by abnormal type I collagen synthesis or structure.

166. A preterm infant has hyponatremia, poor weight gain, high urine sodium losses, and otherwise adequate caloric intake. Which nutritional issue may be limiting growth?

A. Sodium restriction should be intensified because all preterm growth failure is caused by excessive extracellular sodium.

B. High urine sodium strongly suggests SIADH, in which additional sodium generally worsens total-body sodium depletion.

C. Sodium depletion can impair growth and may require supplementation when renal sodium losses are substantial.

D. Adequate calories and protein should overcome renal sodium losses, so sodium supplementation is unlikely to influence growth.

167. An ELBW infant with prolonged broad-spectrum antibiotic exposure and a central line develops thrombocytopenia, persistent hyperglycemia, and sepsis despite antibacterial therapy. Which pathogen should be strongly considered?

A. Parvovirus B19, which causes anemia by erythroid suppression.

B. Invasive Candida infection, which is favored by extreme prematurity, central access, and prolonged antibiotic exposure.

C. Group B Streptococcus acquired during delivery, which is the exclusive cause of late-onset sepsis in ELBW infants.

D. Treponema pallidum, which causes congenital syphilis.

168. A newborn with severe unconjugated hyperbilirubinemia develops poor feeding, hypotonia, and a high-pitched cry, followed by increasing tone and opisthotonus. Which neurologic syndrome is developing?

A. Spinal muscular atrophy, which causes lower-motor-neuron weakness without a high-pitched cry or bilirubin relationship.

B. Neonatal opioid withdrawal causing irritability, abnormal tone, and a high-pitched cry during the early neonatal period.

C. Acute bilirubin encephalopathy from unconjugated bilirubin neurotoxicity affecting vulnerable brain regions.

D. Benign neonatal jitteriness causing stimulus-sensitive tremulousness and increased tone in an otherwise stable infant.

169. A mother receives a complete course of antenatal corticosteroids at 27 weeks and remains pregnant. At 30 weeks she again has a high likelihood of delivery within the next week. Which principle guides consideration of a repeat course?

A. Weekly corticosteroid courses should continue until 37 weeks whenever preterm birth remains possible.

B. A single rescue course may be considered in selected patients still at high risk of preterm birth after an earlier course.

C. Antenatal corticosteroids have no benefit once any earlier course has been completed during the pregnancy.

D. Repeat corticosteroids are used primarily to prevent neonatal sepsis and do not influence pulmonary maturation.

170. A cyanotic newborn has pulmonary atresia with an intact ventricular septum and ductal-dependent pulmonary blood flow. Which initial medication is essential while anatomy and intervention are being defined?

A. A beta blocker to reduce systemic vascular resistance and thereby increase right-to-left ductal shunting.

B. Indomethacin to close the ductus and force antegrade flow across the atretic pulmonary valve.

C. High-dose furosemide to reduce ductal flow and increase right-ventricular filling pressure.

D. Prostaglandin E1 to maintain ductal patency and preserve pulmonary blood flow.

171. A mechanically ventilated preterm infant is switched to very rapid rates but shows worsening carbon dioxide retention and hyperinflation. What ventilator concept best explains the problem in a lung region with a long time constant?

A. Insufficient expiratory time causes incomplete lung emptying and progressive air trapping.

B. Carbon dioxide retention in this setting is caused primarily by excessive inspiratory time and rarely by shortened expiration.

C. Air trapping requires very low airway resistance, so increasing expiratory resistance would improve emptying.

D. A long time constant means the lung empties instantaneously, so higher rates generally reduce hyperinflation.

172. A neonate with disproportionate rhizomelic short stature is found to carry a heterozygous pathogenic FGFR3 variant associated with achondroplasia. Which molecular consequence best explains the growth abnormality?

A. Loss of type I collagen causing diffuse fractures and osteopenia.

B. A 22q11.2 deletion causing pharyngeal arch abnormalities and hypocalcemia.

C. Deficient dystrophin causing congenital long-bone shortening and craniofacial disproportion at birth.

D. An activating FGFR3 variant impairing endochondral bone growth in achondroplasia.

173. A 30-week infant is advancing oral feeds. During bottle feeding the infant has repeated coughing, desaturation, and bradycardia despite good baseline respiratory status. What should be considered before simply increasing nipple flow?

A. The pattern is diagnostic of necrotizing enterocolitis even without abdominal findings, stool changes, or systemic illness.

B. Faster nipple flow should be used because desaturation strongly suggests the infant is not receiving enough milk per swallow.

C. Oral feeding should continue through bradycardia to accelerate maturation of airway protective reflexes.

D. Immature suck-swallow-breathe coordination or aspiration should be assessed and feeding technique adjusted.

174. After neonatal bacterial meningitis with ventriculitis, serial cranial ultrasonography shows steadily expanding lateral ventricles and transependymal fluid flow. Which complication should be suspected?

A. Physiologic macrocephaly, which should not progress rapidly during an acute central nervous system infection.

B. Postinfectious hydrocephalus from impaired cerebrospinal fluid flow or absorption after meningitis.

C. Neonatal abstinence syndrome, which causes irritability and feeding difficulty.

D. Transient tachypnea, which causes pulmonary fluid retention.

175. A newborn has interrupted aortic arch, hypocalcemia, and a small or absent thymic shadow. Which genetic diagnosis should be strongly considered?

A. A 15q11 deletion causing Prader-Willi syndrome with neonatal hypotonia and later hyperphagia.

B. A 22q11.2 deletion syndrome affecting pharyngeal arch development.

C. An FMR1 expansion causing fragile X syndrome with neonatal macroorchidism and aortic interruption.

D. A mitochondrial DNA deletion causing isolated cardiomyopathy without thymic or calcium abnormalities.

176. An infant on volume-targeted ventilation becomes more compliant after surfactant. Which advantage of volume targeting is most relevant as compliance changes?

A. The ventilator can reduce delivered pressure as needed to achieve the target tidal volume and limit volutrauma.

B. Targeted tidal volume is expected to provide normal gas exchange regardless of endotracheal leak or rapidly changing lung mechanics.

C. Volume targeting fixes inspiratory pressure permanently, so tidal volume rises as compliance improves after surfactant.

D. The mode prevents all spontaneous breathing and therefore eliminates any need to adjust ventilator support.

177. A newborn has very short limbs, a narrow bell-shaped thorax, macrocephaly, and severe respiratory failure from pulmonary hypoplasia. Radiographs show bowed femurs. Which lethal skeletal dysplasia is most likely?

A. Osteogenesis imperfecta type I, which typically causes mild childhood fractures.

B. Turner syndrome, which can cause short stature but not a lethal micromelic skeletal dysplasia at birth.

C. Achondroplasia, which usually has a larger thorax and is compatible with survival into adulthood.

D. Thanatophoric dysplasia, a severe FGFR3-related skeletal dysplasia with a narrow thorax and pulmonary hypoplasia.

178. A VLBW infant has adequate weight gain but disproportionately poor linear and head growth while receiving calorie-dense feeds with relatively low protein. Which adjustment is most appropriate?

A. Increase carbohydrate energy further because linear and head growth usually respond first to added calories even when protein density remains low.

B. Increase protein delivery within an appropriate energy-to-protein balance rather than adding calories alone.

C. Remove fortifier to reduce nitrogen intake because protein primarily increases fat mass in preterm infants.

D. Restrict all energy intake until length catches up because adequate weight gain strongly suggests the infant is overnourished.

179. A term infant with HIE has continuous EEG showing frequent electrographic seizures without visible clinical movements after antiseizure medication. What is the most important implication?

A. Electroclinical dissociation is mainly a problem in preterm infants, making EEG-detected events less convincing in a term infant.

B. Electrographic seizure monitoring is necessary because clinical observation alone can miss ongoing neonatal seizures.

C. The absence of visible movements strongly suggests the EEG events are artifacts and antiseizure treatment should be stopped immediately.

D. Clinical motor observation is sufficient for most neonatal seizure treatment decisions after antiseizure medication has been given.

180. A term infant with severe lung disease has preductal oxygen saturation of 94% and postductal saturation of 80%. Echocardiography shows right-to-left ductal shunting without a structural heart defect. What does the saturation difference indicate?

A. Pulmonary vascular resistance remains high enough to drive deoxygenated blood through the ductus into the descending aorta.

B. Aortic coarctation is causing oxygen extraction in the legs while pulmonary vascular resistance is normal.

C. d-Transposition should be the leading diagnosis when a preductal-postductal oxygen saturation gradient is present.

D. The left-to-right ductal shunt is delivering highly oxygenated blood preferentially to the lower extremities.

181. A preterm infant with RDS is receiving CPAP. Which physiologic benefit is the main reason CPAP can reduce work of breathing?

A. Continuous distending pressure helps maintain functional residual capacity and prevents repeated alveolar collapse.

B. CPAP removes all airway resistance by paralyzing the upper airway muscles during spontaneous breathing.

C. Continuous pressure suppresses surfactant production so alveoli become easier to recruit with lower surface tension.

D. CPAP primarily works by decreasing end-expiratory lung volume and promoting complete alveolar emptying.

182. A newborn is profoundly hypotonic with facial diplegia, weak respiratory effort, and clubfeet. The mother has grip myotonia and difficulty releasing her handshake. Which diagnosis is most likely?

A. Congenital hypothyroidism, which can cause hypotonia but not the characteristic maternal neuromuscular history.

B. Congenital myotonic dystrophy due to a maternally transmitted expanded DMPK repeat.

C. Spinal muscular atrophy caused by SMN1 loss, which is not associated with maternal myotonia from anticipation.

D. Prader-Willi syndrome, which causes hypotonia.

183. A human-milk-fed 27-week infant is clinically stable and gaining weight but receives no vitamin D source beyond breast milk. Which nutritional principle is appropriate?

A. Use ultraviolet phototherapy as the sole vitamin D source because enteral supplementation is ineffective in preterm infants.

B. Provide vitamin D supplementation according to preterm-infant recommendations because human milk alone is usually insufficient.

C. Defer vitamin D until substantial enteral volumes are established because early supplementation adds little to neonatal bone mineralization.

D. Avoid vitamin D during hospitalization because it prevents intestinal calcium absorption and increases rickets risk.

184. Serial ultrasonography after a large preterm intraventricular hemorrhage shows progressive increases in ventricular measurements before overt neurologic deterioration. Which posthemorrhagic process is developing?

A. Residual scalp edema from delivery trauma can explain a steadily increasing head circumference during the first several weeks.

B. Congenital microcephaly, which is defined by small.

C. Posthemorrhagic ventricular dilatation from impaired cerebrospinal fluid circulation after IVH.

D. Perinatal arterial stroke, which usually produces a focal infarct.

185. A 24-week infant has low measured blood pressure but warm extremities, normal capillary refill, adequate urine output, and normal lactate. Which approach is most appropriate?

A. Assume normal perfusion excludes all cardiovascular disease and discontinue hemodynamic monitoring largely.

B. Interpret blood pressure together with clinical perfusion rather than treating a numeric threshold in isolation.

C. Give repeated saline boluses until mean arterial pressure exceeds gestational age even if perfusion remains normal.

D. Start vasopressors primarily because systolic pressure is below 50 mm Hg, regardless of the infant's clinical state.

186. A term infant has persistent respiratory distress and a chest radiograph showing a hyperinflated left upper lobe compressing the remaining lung and shifting the mediastinum. Which diagnosis is most likely?

A. Hyaline membrane disease, which produces diffuse low-volume granular lungs in a premature infant.

B. Bilateral chylothorax, which causes pleural fluid and compressed lungs.

C. Congenital lobar overinflation caused by abnormal lobar airway support or obstruction.

D. Pulmonary hypoplasia from oligohydramnios, which causes bilateral small lungs.

187. A 28-week infant is receiving parenteral nutrition through a central line. Conjugated bilirubin gradually rises after several weeks with minimal enteral intake. Which intervention can reduce risk of intestinal-failure-associated or parenteral-nutrition-associated cholestasis?

A. Advance enteral nutrition when feasible and avoid unnecessary prolonged exclusive parenteral nutrition.

B. Stop all enteral milk because exposure of the intestine to nutrients increases bile stasis and conjugated bilirubin.

C. Increase continuous dextrose infusion markedly because higher carbohydrate delivery consistently reverses cholestasis.

D. Remove all amino acids permanently because protein is the sole cause of parenteral-nutrition-associated liver disease.

188. A preterm infant is receiving fortified human milk and has rapid weight gain from edema rather than improved length or head circumference. Which assessment best distinguishes true nutritional growth from fluid accumulation?

A. Track serial weight together with length, head circumference, fluid balance, and the clinical examination.

B. Measure serum glucose primarily because normal glucose strongly suggests that all weight gain represents new lean tissue.

C. Use weight alone because body water changes are negligible in preterm infants after the first day of life.

D. Weight velocity is the most reliable measure of tissue growth during hospitalization, so other anthropometric trends add limited information.

189. A clinically well newborn has repeat pulse-oximetry screening values of 89% in the right hand and 90% in a foot despite repositioning the probes and allowing adequate settling time. What should happen next?

A. Repeat screening at the two-week well visit because immediate evaluation is unnecessary in an asymptomatic infant.

B. Treat the screen as failed and promptly evaluate for congenital heart disease or another cause of hypoxemia.

C. Apply supplemental oxygen until the saturation rises above 95%, then consider the screening result normal without further assessment.

D. Discharge the infant because a preductal-postductal difference below 3 percentage points rules out important disease.

190. Prenatal ultrasonography shows a multicystic intrathoracic lung lesion supplied by the pulmonary circulation. After birth the infant has mild respiratory distress. Which congenital lesion is most consistent?

A. Bronchopulmonary sequestration, which is defined by normal airway connection and exclusive pulmonary arterial blood supply.

B. Congenital pulmonary airway malformation, a cystic developmental abnormality of the lung.

C. Congenital diaphragmatic hernia, which contains abdominal organs.

D. Pulmonary interstitial emphysema, which is an acquired air leak after positive-pressure ventilation.

191. A neonate has a single arterial trunk overriding a large ventricular septal defect. Over the next several days tachypnea and heart failure worsen. What is driving the change?

A. Spontaneous closure of the truncal valve causes severe left-ventricular outflow obstruction in nearly all affected neonates.

B. Rising pulmonary vascular resistance after birth diverts the entire common-trunk output to the lungs.

C. Declining pulmonary vascular resistance permits progressively greater pulmonary blood flow through the common arterial trunk.

D. Closure of the VSD eliminates systemic output and causes isolated pulmonary venous hypertension.

192. A neonate has an intrathoracic mass with no normal communication to the tracheobronchial tree. Doppler identifies an arterial feeder arising from the aorta. Which diagnosis is most likely?

A. Bronchopulmonary sequestration supplied by an anomalous systemic artery.

B. Transient tachypnea caused by retained fetal lung fluid without a discrete mass or anomalous arterial feeder.

C. Primary pulmonary hypertension caused by failure of postnatal vascular relaxation.

D. Congenital pulmonary airway malformation supplied exclusively by a normal branch pulmonary artery in most case.

193. A neonate has critical pulmonary valve stenosis with a severely elevated right-ventricular pressure and markedly reduced antegrade pulmonary blood flow. Which catheter intervention directly relieves the obstruction in suitable anatomy?

A. Balloon atrial septostomy to create mixing while leaving the fixed valvar obstruction unchanged.

B. Coil occlusion of the ductus arteriosus before the right ventricle can generate adequate pulmonary flow.

C. Transcatheter closure of a ventricular septal defect to reduce pressure within the right ventricle.

D. Balloon pulmonary valvuloplasty to open the critically stenotic pulmonary valve.

194. A newborn with respiratory distress has milky pleural fluid after enteral feeding begins. Pleural fluid contains abundant lymphocytes and elevated triglycerides. What is the diagnosis?

A. Pneumothorax, which contains pleural air.

B. Neonatal chylothorax from lymphatic fluid accumulating in the pleural space.

C. Empyema from bacterial pneumonia, which typically produces purulent neutrophil-rich pleural fluid.

D. Pulmonary hemorrhage, which fills the alveoli and airways with blood.

195. A term neonate has persistent hypoventilation and hypercapnia that are most pronounced during sleep. Lung imaging and cardiac evaluation are unrevealing. Which disorder should be considered when central ventilatory responses are impaired?

A. Apnea of prematurity, which is expected in a term infant with persistent hypercapnia and normal neurologic maturation.

B. Meconium aspiration syndrome, which should produce characteristic parenchymal lung abnormalities on imaging.

C. Congenital central hypoventilation syndrome associated with pathogenic PHOX2B variants.

D. Transient tachypnea, which causes tachypnea from retained lung fluid.

196. A newborn with pneumonia has stiff lungs and requires ventilation. Which change in lung mechanics is most likely?

A. Increased compliance means the lungs are stiff and require greater pressure for a small change in volume.

B. Reduced resistance alone explains stiff lungs even when pressure-volume behavior is substantially normal.

C. Reduced compliance means a larger pressure change is required to generate a given tidal volume.

D. High compliance is synonymous with low functional residual capacity and generally indicates diffuse atelectasis.

197. A very preterm infant is being extubated after improvement from RDS. Which factor most supports successful extubation?

A. Complete absence of spontaneous breaths while sedated because respiratory effort is unnecessary after extubation to CPAP.

B. Persistent severe apnea despite caffeine and frequent stimulation because extubation strengthens central respiratory drive.

C. Escalating oxygen requirement and worsening acidosis despite high ventilator pressures because noninvasive support is generally stronger.

D. Stable spontaneous respiratory drive with manageable oxygen need and adequate gas exchange on low ventilator support.

198. A 23-week infant has severe respiratory failure and marked ventilator asynchrony despite conventional support. The team considers high-frequency ventilation. Which general principle is correct?

A. The mode delivers very large tidal volumes at slow rates to wash out carbon dioxide from overdistended alveoli.

B. Once high-frequency ventilation stabilizes oxygenation, chest radiographs and blood gases can be minimized because delivered tidal volumes are small.

C. High-frequency ventilation eliminates the need to set mean airway pressure because oxygenation becomes independent of lung recruitment.

D. High-frequency ventilation uses very small tidal volumes at rapid frequencies while lung-volume strategy remains crucial.

199. A term infant with severe pulmonary hypertension is receiving inhaled nitric oxide. Methemoglobin levels rise substantially. Which mechanism explains this adverse effect?

A. Nitric oxide converts fetal hemoglobin into adult hemoglobin, reducing oxygen affinity by changing globin-chain synthesis.

B. Oxidation of hemoglobin iron reduces its ability to carry oxygen when methemoglobin accumulates.

C. Methemoglobin increases oxygen-carrying capacity because ferric iron binds oxygen more avidly than ferrous iron.

D. The adverse effect results from nitric oxide directly destroying platelets while leaving hemoglobin chemistry unchanged.

200. A newborn with severe meconium aspiration develops air leak and pulmonary hypertension. Which ventilation strategy principle is most appropriate?

A. Use the lowest pressures and tidal volumes that achieve adequate gas exchange while avoiding worsening air trapping.

B. Target profound hypocapnia with aggressive ventilation because low carbon dioxide is protective against neonatal brain injury.

C. Deliberately use very high tidal volumes to overcome meconium obstruction even if overdistention increases.

D. Eliminate expiratory time so trapped gas splints obstructed airways open throughout the respiratory cycle.

Answers

1. D. NSAIDs inhibit prostaglandin synthesis and can cause premature constriction of the fetal ductus arteriosus, particularly later in gestation. The resulting rise in right-sided pressure can produce tricuspid regurgitation or right-ventricular dysfunction. The drug does not open the ductus or selectively increase fetal pulmonary flow.

2. A. Very preterm infants lose heat rapidly because of thin skin, large surface area, and limited insulating fat. A warm room, radiant heat, polyethylene wrap or bag, and other thermal measures reduce hypothermia, which is associated with hypoglycemia, acidosis, and worse outcomes.

3. B. Critical coarctation can remain compensated while ductal flow supports the descending aorta. As the ductus constricts, lower-body perfusion falls, producing weak femoral pulses, acidosis, shock, and differential upper-versus-lower extremity pressures. Prostaglandin E1 is used to restore ductal patency while definitive therapy is arranged.

4. B. Respiratory distress syndrome in very preterm infants is driven by deficient surfactant, leading to high surface tension, atelectasis, decreased compliance, and impaired gas exchange. CPAP helps maintain alveolar recruitment, and exogenous surfactant addresses the underlying deficiency when indicated.

5. A. The combination of characteristic facial features, neonatal hypotonia, and complete atrioventricular septal defect strongly suggests Down syndrome. Cytogenetic confirmation is important because recurrence counseling differs among free trisomy, translocation, and mosaic forms.

6. D. Mother's own milk is the preferred enteral feeding for very low birth weight infants because it supports gastrointestinal, immune, and developmental outcomes. Because unfortified human milk may not meet the high protein and mineral needs of a growing preterm infant, appropriate fortification is commonly required.

7. B. At birth, neonatal serum creatinine reflects maternal levels because creatinine crosses the placenta. Subsequent trends depend on gestational age, renal maturation, perfusion, and illness, so an isolated early value does not by itself diagnose acute kidney injury.

8. D. Congenital hypothyroidism is often clinically subtle at birth, but untreated deficiency can cause irreversible neurodevelopmental impairment. Prompt confirmation and initiation of levothyroxine are therefore central to newborn-screening programs.

9. C. SCID can present in early infancy with persistent candidiasis, severe or unusual infections, diarrhea, failure to thrive, and lymphopenia. It is a medical emergency because definitive therapy and infection precautions should be instituted before life-threatening infections accumulate.

10. C. Group B Streptococcus remains an important cause of early-onset neonatal sepsis acquired around labor and delivery. Risk is influenced by maternal colonization, rupture of membranes, gestational age, intrapartum fever, and adequacy of intrapartum prophylaxis.

11. D. Pneumatosis intestinalis is a hallmark radiographic finding of NEC in the appropriate clinical setting. Portal venous gas further supports intestinal mucosal injury; management includes bowel rest, decompression, antibiotics, supportive care, and surgical consultation when indicated.

12. A. Jaundice in the first 24 hours is pathologic until proven otherwise. Hemolytic disease produces increased bilirubin load, anemia, and reticulocytosis; a positive direct antiglobulin test supports antibody-coated erythrocytes.

13. B. Cutis marmorata telangiectatica congenita presents at birth with a persistent reticulated vascular pattern that may include telangiectasia or skin atrophy. Unlike physiologic cutis marmorata, it does not simply disappear with warming.

14. C. Late vitamin K deficiency bleeding often occurs between several weeks and months of age, especially in exclusively breastfed infants who did not receive prophylaxis or who have unrecognized cholestasis. Intracranial bleeding is a feared presentation.

15. A. For eligible infants with moderate-to-severe hypoxic-ischemic encephalopathy, therapeutic hypothermia should begin as soon as possible within the established early window and continue for about 72 hours under protocolized monitoring. MRI is important for assessment but should not delay time-sensitive treatment.

16. B. Corrected age subtracts the number of weeks born before 40 weeks from chronological age. A baby born at 30 weeks was about 10 weeks early, so at 5 months chronologic age the corrected age is roughly 2.5 months; corrected age is useful for interpreting early milestones while the child's trajectory is followed.

17. A. Very preterm and very low birth weight infants are at risk for ROP and require timed retinal examinations according to screening criteria. Timing is based on gestational and postmenstrual age so that treatment-requiring disease can be identified before retinal detachment occurs.

18. C. Total body water and extracellular water are proportionally high in very preterm infants. Hydrophilic drugs therefore may have a larger volume of distribution per kilogram, affecting loading-dose requirements.

19. B. When more than one medically reasonable path exists, shared decision-making respects parents' role while clinicians provide recommendations, prognosis, and support. Decisions can be revisited as new clinical information changes the balance of benefits and burdens.

20. B. Relative risk is risk in the exposed or treated group divided by risk in the comparison group. Here 0.18/0.24 = 0.75, corresponding to a 25% relative risk reduction; the absolute risk reduction is 6 percentage points.

21. C. Less-invasive surfactant techniques can provide exogenous surfactant to selected spontaneously breathing preterm infants while avoiding or shortening mechanical ventilation. The infant still requires appropriate distending pressure and respiratory monitoring after administration.

22. A. Edwards syndrome commonly causes severe fetal growth restriction, prominent occiput, micrognathia, low-set ears, clenched hands with overlapping fingers, and rocker-bottom feet. Major cardiac and renal anomalies are frequent.

23. D. When mother's own milk is unavailable or insufficient, pasteurized donor human milk is commonly favored for VLBW infants because human milk feeding is associated with lower NEC risk. Donor milk often requires fortification to achieve adequate protein and mineral delivery for growth.

24. A. Neonatal AKI is identified by changes in serum creatinine and/or urine output interpreted in context. A progressive creatinine rise after the expected maternal contribution should be falling, especially with oliguria and systemic illness, supports renal injury.

25. A. Classic 21-hydroxylase deficiency causes deficient cortisol and often aldosterone with excess androgen production. Affected infants can develop life-threatening salt-wasting crisis with hyponatremia, hyperkalemia, dehydration, and hypoglycemia; virilization may be evident in 46,XX infants.

26. C. The thymus develops from the third pharyngeal pouch, and 22q11.2 deletion can impair this process. The degree of T-cell deficiency varies from mild reduction to rare complete thymic aplasia.

27. B. E coli is a major early-onset pathogen, particularly among very preterm and very low birth weight infants, and can cause severe bacteremia and meningitis. Empiric early-onset sepsis regimens are designed to cover both GBS and common gram-negative organisms.

28. B. Spontaneous intestinal perforation typically occurs in extremely preterm infants early in the course and may present as a focal perforation without the diffuse intestinal inflammation or pneumatosis of NEC. Clinical overlap exists, but timing and imaging can help distinguish them.

29. D. Unconjugated bilirubin circulates largely bound to albumin. Low albumin, acidosis, and competing substances can increase unbound bilirubin, while sepsis and clinical instability may increase vulnerability to neurotoxicity.

30. D. Aplasia cutis congenita is a focal congenital absence of skin, most often on the scalp. Management depends on defect size, depth, and associated skull or vascular involvement; large lesions may require imaging and specialized care.

31. C. Fetal and neonatal alloimmune thrombocytopenia is analogous to hemolytic disease but targets platelet antigens. Maternal alloantibodies can affect a first pregnancy and cause profound neonatal thrombocytopenia with risk of intracranial hemorrhage despite a normal maternal platelet count.

32. B. Evidence supporting therapeutic hypothermia is strongest for term and near-term infants meeting trial-based criteria. Current AAP guidance does not recommend therapeutic hypothermia for infants born below 35 weeks because benefit has not been established and harm is possible.

33. D. High-risk NICU graduates benefit from multidisciplinary follow-up that integrates growth, neurologic examination, developmental assessment, hearing, vision, feeding, and family support. Early intervention can begin based on risk or delay without waiting for a final diagnostic label.

34. C. Treatment-requiring ROP needs timely ophthalmologic therapy, commonly laser or intravitreal anti-VEGF depending on zone, stage, and clinical circumstances. Oxygen should be managed within appropriate neonatal targets rather than abruptly withdrawn.

35. D. Aminoglycosides are eliminated largely by the kidneys. Reduced and rapidly changing glomerular filtration in premature neonates prolongs elimination, so dosing intervals and therapeutic drug monitoring are individualized to gestational age, postnatal age, renal function, and drug concentrations.

36. A. Qualified interpreters improve accuracy, confidentiality, and informed participation. Family members, especially children, should not be relied on for high-stakes consent except in genuine emergencies when no safer alternative is available.

37. A. Case-control studies begin with outcome status and look backward at exposures. Because the number of cases and controls is set by design, incidence and direct risk ratios are usually not estimated; the odds ratio is the usual measure of association.

38. C. Poorly controlled maternal phenylketonuria creates a teratogenic intrauterine environment. High maternal phenylalanine crosses the placenta and can cause fetal growth restriction, microcephaly, congenital heart disease, and neurodevelopmental injury regardless of the fetus's own PAH genotype.

39. D. In neonatal resuscitation, effective ventilation is the critical intervention for most bradycardic newborns. If the chest is not moving, the team should correct ventilation technique and establish effective breaths before deciding that compressions are needed.

40. D. In d-TGA, systemic and pulmonary circulations run in parallel, so survival depends on mixing through the atrial septum, ductus, or ventricular septum. A restrictive atrial communication can cause severe hypoxemia despite prostaglandin, and balloon atrial septostomy can urgently improve mixing.

41. A. Posterior urethral valves are a major cause of congenital lower urinary tract obstruction in male infants. Severe obstruction can cause a thick-walled distended bladder, bilateral hydroureteronephrosis, renal dysplasia, oligohydramnios, and postnatal renal dysfunction.

42. D. Hyperinsulinism causes inappropriate glucose utilization and suppresses alternative fuel generation, so ketones and free fatty acids remain low during hypoglycemia. This biochemical pattern is an important clue when evaluating persistent neonatal hypoglycemia.

43. D. Maternal IgG is actively transported across the placenta, with much transfer occurring in the third trimester. This gives term infants passive systemic antibody protection; preterm infants have lower IgG concentrations because they miss part of this late gestational transfer.

44. A. Neonatal HSV may present with skin-eye-mouth disease, encephalitis, or disseminated infection, and maternal history can be absent. Vesicles plus seizures should prompt immediate acyclovir while surface, blood, and CSF testing is pursued as clinically appropriate.

45. A. Meconium ileus is strongly associated with cystic fibrosis and results from thick, tenacious meconium obstructing the distal ileum. A microcolon reflects an unused distal colon; uncomplicated cases may respond to therapeutic contrast enema.

46. B. Direct hyperbilirubinemia is never physiologic. Pale or acholic stools and dark urine suggest impaired bile flow, and biliary atresia requires prompt evaluation because earlier surgical drainage is associated with better outcomes.

47. C. Maternal anti-Ro/SSA or anti-La/SSB antibodies can cause neonatal lupus. Cutaneous lesions are often annular and photosensitive and generally resolve as maternal antibodies disappear, whereas congenital heart block may be permanent.

48. B. With subacute or chronic fetal blood loss, the marrow increases erythropoiesis, producing reticulocytosis and nucleated red cells. Hyperacute hemorrhage may cause shock before a strong marrow response has had time to develop.

49. C. Phenobarbital remains a common first-line antiseizure medication for acute neonatal seizures. Treatment also requires rapid correction of reversible causes and use of EEG monitoring because electrographic seizures may continue without obvious clinical movements.

50. B. Spastic diplegia is strongly associated with preterm white-matter injury and produces greater lower-extremity than upper-extremity involvement. Persistent toe walking, scissoring, and increased leg tone are characteristic findings.

51. C. A dense unilateral congenital cataract deprives the developing visual system of clear input and can produce profound amblyopia. Timely surgery when indicated and aggressive visual rehabilitation are essential for preserving useful vision.

52. C. Unconjugated bilirubin is largely albumin bound. Competition for binding sites can increase the free fraction available to cross into tissues, which is one reason certain highly protein-bound medications require caution in jaundiced neonates.

53. A. Withholding and withdrawing life-sustaining treatment are ethically equivalent when treatment no longer serves the patient's goals or best interests. Comfort care, including appropriate analgesia and family support, remains an active obligation.

54. C. Predictive values depend on prevalence. As disease becomes rarer, positive predictive value generally falls and negative predictive value rises, while sensitivity and specificity are intrinsic test-performance measures that are less directly affected by prevalence.

55. C. Second- and third-trimester exposure to ACE inhibitors or angiotensin receptor blockers can disrupt the fetal renin-angiotensin system. Reduced renal perfusion and urine production cause oligohydramnios, renal dysgenesis or failure, and secondary pulmonary hypoplasia.

56. B. Neonatal cardiac arrest is usually secondary to respiratory failure, so coordinated ventilation remains prominent during compressions. The standard neonatal sequence uses a 3:1 compression-to-ventilation ratio, yielding about 90 compressions and 30 breaths per minute.

57. B. Complete atrioventricular septal defect is strongly associated with Down syndrome. Turner syndrome is more closely linked to left-sided obstructive lesions, and the other listed chromosomal conditions do not have the same characteristic association.

58. D. TTN reflects delayed resorption of fetal lung fluid and is more common after cesarean delivery without labor. Typical findings include tachypnea, perihilar streaking, fluid in fissures, and mild hyperinflation, with improvement over the next one to three days.

59. A. Patau syndrome commonly includes severe central nervous system malformations such as holoprosencephaly, facial clefts, microphthalmia, polydactyly, and congenital heart disease. The constellation is highly characteristic of trisomy 13.

60. A. Preterm infants have high protein and mineral needs that unfortified human milk often cannot meet. Fortification increases nutrient density and helps support bone mineralization and lean growth while preserving many benefits of human milk.

61. C. Congenital CMV can cause fetal growth restriction, petechiae, hepatosplenomegaly, microcephaly, periventricular calcifications, sensorineural hearing loss, and other neurologic injury. Many infected infants are asymptomatic, so targeted or universal screening practices vary.

62. D. Hirschsprung disease results from failed migration of enteric neural crest cells, leaving an aganglionic distal segment that remains tonically contracted. Delayed meconium passage, abdominal distention, and explosive stool after rectal examination are classic clues.

63. C. Phototherapy changes bilirubin configuration and structure to form products that are more water soluble and can be excreted in bile and urine without normal conjugation. Effectiveness depends on irradiance, exposed surface area, and light wavelength.

64. D. Incontinentia pigmenti typically affects females and evolves through vesicular, verrucous, hyperpigmented, and sometimes atrophic stages along Blaschko lines. Dental, ocular, and neurologic abnormalities can accompany the skin findings.

65. D. Exchange transfusion replaces sensitized red cells, lowers bilirubin, removes circulating maternal antibody, and corrects severe anemia. It is reserved for infants meeting established criteria because the procedure carries important risks.

66. A. Hyperekplexia is a disorder of inhibitory glycinergic neurotransmission, often involving GLRA1 or related genes. Newborns can have marked startle, generalized stiffness, and dangerous apnea precipitated by tactile or auditory stimuli despite a nonictal EEG.

67. B. Former extremely preterm children can develop executive-function, attention, language, learning, and behavioral difficulties even when major motor disability is absent. Some challenges become apparent only when school demands increase.

68. A. Auditory brainstem response evaluates the auditory pathway through the brainstem and can detect neural abnormalities that may be missed by otoacoustic emissions alone. NICU infants have a higher risk of auditory neuropathy and need screening strategies suited to that risk.

69. C. Drug-metabolizing pathways mature at different rates after birth. Some conjugation pathways, including glucuronidation for certain substrates, are limited in neonates, which can prolong half-life and increase accumulation unless dosing is adjusted.

70. A. Equity work requires identifying modifiable barriers and partnering with families and communities to improve access. Equal treatment is not always equitable when transportation, language, work schedules, housing, or other structural factors create unequal ability to use care.

71. A. Statistical power is 1 minus the type II error probability and reflects the chance of detecting a prespecified true effect under the assumed sample size, variability, effect size, and alpha level. Power does not guarantee clinical importance or absence of bias.

72. C. Magnesium crosses the placenta, and elevated neonatal magnesium can cause hypotonia, diminished reflexes, poor feeding, and respiratory depression. The effect is usually transient as the infant clears the magnesium; it is not a syndrome of hyperreflexia or catecholamine excess.

73. D. Volume expansion is considered when a newborn fails to respond to standard resuscitation and there is evidence of hypovolemia or acute blood loss. Isotonic crystalloid or emergency blood can restore circulating volume; routine empiric volume loading without evidence of hypovolemia is not appropriate.

74. D. Prostaglandin E1 can cause apnea, hypotension, fever, and flushing. Because ductal patency may be lifesaving in critical congenital heart disease, the response is to support ventilation as needed rather than reflexively stop a necessary infusion.

75. C. Meconium can obstruct airways unevenly. Partial obstruction allows air entry during inspiration but limits expiration, producing air trapping and hyperinflation, while complete obstruction produces atelectasis. This heterogeneity contributes to ventilation-perfusion mismatch and air-leak risk.

76. B. Turner syndrome often presents with congenital lymphedema, webbed neck or broad chest, and left-sided cardiac lesions such as coarctation and bicuspid aortic valve. The classic karyotype is 45,X, although mosaic and structural X abnormalities also occur.

77. A. Growth-restricted infants can develop a refeeding-like syndrome when robust nutrition abruptly stimulates anabolism. Insulin-mediated cellular uptake of phosphorus and potassium can cause significant hypophosphatemia and hypokalemia, so early electrolyte monitoring is important.

78. D. Amniotic fluid is important for normal fetal lung expansion. Severe early oligohydramnios from bilateral renal agenesis or profound urinary tract obstruction can produce pulmonary hypoplasia, which may dominate the neonatal presentation.

79. C. Infants of diabetic mothers are at increased risk for early hypocalcemia, related in part to altered parathyroid regulation and associated hypomagnesemia in some cases. Symptomatic infants require evaluation and treatment based on ionized calcium and the clinical picture.

80. B. Unlike IgG, IgM is a large pentamer and does not significantly cross the placenta. Thus pathogen-specific IgM in the neonate can support congenital infection, although test sensitivity and specificity vary by pathogen and assay.

81. A. Some group O mothers have IgG anti-A and anti-B antibodies that can cross the placenta without prior pregnancy sensitization. ABO hemolytic disease is often milder than Rh disease but can cause early jaundice and anemia.

82. D. Periventricular leukomalacia affects immature white matter near the lateral ventricles and is associated with later motor impairment, particularly spastic diplegia. Modern preterm brain injury is often noncystic, but cystic PVL remains a recognizable severe pattern.

83. D. Developmental screening identifies children who need further evaluation; a normal general examination does not exclude developmental delay. Early assessment and intervention can address language, motor, cognitive, feeding, and social-emotional needs while diagnostic clarification continues.

84. A. Bilateral choanal atresia can cause cyclic cyanosis that improves with crying because the mouth opens and bypasses the nasal obstruction. Immediate airway stabilization is followed by diagnostic confirmation and surgical planning.

85. C. Alpha is the tolerated type I error rate under repeated testing when the null hypothesis is true. A P value is compared with alpha, but neither alpha nor the P value is the probability that the null hypothesis itself is true.

86. D. Parvovirus B19 infects erythroid progenitor cells and can cause transient aplastic anemia in the fetus. Severe anemia may lead to high-output cardiac failure and hydrops. Rubella, CMV, and varicella have different characteristic fetal effects.

87. C. Routine endotracheal suctioning for a nonvigorous infant with meconium-stained fluid is not recommended in the absence of airway obstruction. Resuscitation should focus on timely effective ventilation, with suctioning reserved for suspected obstruction that interferes with ventilation.

88. B. Cyclooxygenase inhibitors such as indomethacin can reduce renal blood flow and urine output, making significant renal dysfunction an important consideration when choosing PDA therapy. Management should integrate the infant's hemodynamics, age, contraindications, and current evidence rather than treat an echocardiographic finding in isolation.

89. B. Congenital central hypoventilation syndrome is characterized by inadequate autonomic ventilation, especially during sleep, without primary pulmonary or neuromuscular disease. Pathogenic PHOX2B variants are strongly associated and testing is part of diagnostic evaluation.

90. D. A 22q11.2 deletion can disrupt pharyngeal pouch development, producing conotruncal cardiac defects, thymic hypoplasia with T-cell dysfunction, and hypoparathyroidism with hypocalcemia. The neonatal presentation may vary widely in severity.

91. A. Extremely preterm infants have high protein needs and rapidly accrue negative nitrogen balance if amino acids are delayed. Early parenteral amino acid provision is standard when enteral intake is inadequate, with monitoring for acid-base, renal, and electrolyte tolerance.

92. D. SIADH produces water retention, low serum osmolality, and inappropriately concentrated urine. Neurologic injury and pulmonary disease can trigger ADH release; management depends on volume status, severity, and the underlying cause.

93. D. Newborns rely heavily on brown-fat nonshivering thermogenesis. Cold stress increases catecholamine-driven oxygen and glucose consumption; if demands exceed supply, hypoglycemia, hypoxemia, and metabolic acidosis can develop.

94. B. Wiskott-Aldrich syndrome classically combines eczema, recurrent infection or immune dysfunction, and thrombocytopenia with small platelets. It is X-linked and can present during infancy with bleeding or infection.

95. C. Maternal listeriosis can be mild or influenza-like yet lead to preterm birth and severe early neonatal sepsis, pneumonia, or meningitis. Granulomatosis infantiseptica, with disseminated granulomatous lesions, is a classic severe congenital manifestation of Listeria infection.

96. C. Malrotation narrows the mesenteric base and predisposes the midgut to twist, causing vascular compromise. Bilious emesis in a newborn must be treated as potentially obstructive, and volvulus requires urgent surgical evaluation to prevent intestinal necrosis.

97. A. Newborn hearing screening detects hearing status at that time but does not eliminate risk of later-onset or progressive loss. Infants with important risk factors, including meningitis and some ototoxic exposures, need appropriate audiologic follow-up.

98. B. Pierre Robin sequence begins with mandibular hypoplasia, which displaces the tongue posteriorly and can interfere with palatal closure, producing a U-shaped cleft. Airway and feeding management depend on severity and position.

99. C. Cluster-randomized trials assign groups such as hospitals or units rather than individuals. Patients within a cluster tend to be more similar, creating intracluster correlation that reduces effective sample size and must be considered in design and analysis.

100. A. Fetomaternal hemorrhage can produce neonatal pallor, shock, and anemia, and fetal cells in the maternal circulation support the diagnosis. Placental insufficiency more often increases fetal erythropoiesis, while maternal thrombocytopenia does not explain severe neonatal anemia.

101. D. ECG provides rapid and accurate heart-rate assessment during neonatal resuscitation and is especially useful when advanced resuscitation is required. Pulse oximetry remains important for oxygen saturation, but it may take longer to obtain a reliable signal immediately after birth.

102. D. Hypercyanotic spells result from increased right-to-left shunting as infundibular obstruction worsens and/or systemic vascular resistance falls. A softer murmur can reflect reduced flow across the obstructed outflow tract. Measures that raise systemic vascular resistance and reduce infundibular spasm improve pulmonary blood flow.

103. B. Pulmonary interstitial emphysema occurs when alveolar rupture allows air to dissect along perivascular and peribronchial interstitial tissues. It is associated with mechanical ventilation and immature lungs and may be focal or diffuse.

104. C. Beckwith-Wiedemann syndrome is an overgrowth disorder associated with macroglossia, abdominal wall defects, organomegaly, hemihyperplasia, and neonatal hypoglycemia. Affected children also require tumor surveillance because of increased embryonal tumor risk.

105. B. Intravenous lipid provides energy and essential fatty acids. Prolonged severe lipid restriction can cause essential fatty acid deficiency, with dermatitis, thrombocytopenia, growth failure, and biochemical abnormalities.

106. C. Central diabetes insipidus causes inadequate ADH secretion, leading to inability to concentrate urine, free-water loss, polyuria, and hypernatremia. Severe central nervous system injury is one potential cause in a neonate.

107. D. Classic galactosemia presents after exposure to lactose-containing feeds with poor feeding, vomiting, jaundice, liver dysfunction, and risk of E coli sepsis. Immediate removal of galactose and lactose is essential while confirmatory testing is obtained.

108. B. Placental transfer of maternal IgG accelerates during the third trimester. Extremely preterm infants are born before much of this transfer occurs, contributing to lower passive antibody levels and vulnerability to infection.

109. D. Congenital syphilis can cause rhinitis or snuffles, hepatosplenomegaly, jaundice, rash involving palms and soles, anemia, thrombocytopenia, and characteristic long-bone changes. Evaluation and penicillin therapy depend on maternal treatment history and neonatal findings.

110. D. The most common esophageal atresia pattern includes a blind proximal pouch and a distal fistula to the trachea. The tube coils proximally, while air entering the distal fistula produces gas in the stomach and bowel.

111. C. Anemia of prematurity reflects shortened red-cell survival, phlebotomy losses, rapid growth, and an inappropriately low erythropoietin response relative to anemia. Nutritional status and transfusion practices also influence severity.

112. A. The severe parenchymal lesion historically called grade IV IVH is better understood as periventricular hemorrhagic infarction, usually related to venous congestion and impaired drainage associated with a large germinal-matrix or intraventricular hemorrhage.

113. D. Censoring indicates that the exact event time is not observed beyond a known follow-up point. Methods such as Kaplan-Meier analysis can use each participant's available time at risk under assumptions about censoring.

114. D. Anti-Ro/SSA and anti-La/SSB IgG antibodies can cross the placenta and cause inflammatory injury and fibrosis of the fetal conduction system, producing congenital complete heart block. IgM does not cross the placenta effectively, and the finding is not explained solely by medication exposure.

115. A. A spontaneously breathing preterm infant with respiratory distress can often be supported with CPAP, which helps maintain functional residual capacity and may reduce invasive ventilation. Compressions are not indicated with a normal heart rate, and routine intubation is not required simply because the infant is preterm.

116. C. In parallel single-ventricle physiology, excessive pulmonary vasodilation can increase Qp at the expense of systemic blood flow. Management aims for balanced pulmonary and systemic circulations rather than the highest possible oxygen saturation.

117. D. BPD is a chronic lung disease of prematurity resulting from arrested alveolar and vascular development plus inflammatory and ventilator-related injury. Modern definitions use respiratory support requirements at specified postmenstrual ages to characterize severity.

118. A. Prader-Willi syndrome results from absent paternal gene expression in 15q11-q13. Neonates commonly have severe hypotonia and poor feeding; hyperphagia, obesity risk, and characteristic behavioral features become more prominent later.

119. A. Minimal enteral or trophic feeding exposes the gut to nutrients and may promote motility, hormonal signaling, and mucosal maturation while nutritional needs are still met largely parenterally. It is not nutritionally complete and does not eliminate NEC risk.

120. B. Extremely preterm infants can develop significant hyperkalemia during the first days despite adequate urine output. Immature Na/K-ATPase activity, cell potassium shifts, and limited renal handling contribute; pseudohyperkalemia from hemolysis must also be excluded.

121. D. Maple syrup urine disease impairs metabolism of leucine, isoleucine, and valine. Severe neonatal forms cause poor feeding, encephalopathy, abnormal tone, and the characteristic odor, with leucine being particularly neurotoxic.

122. C. Gonococcal ophthalmia neonatorum can present in the first several days with rapidly progressive hyperpurulent conjunctivitis and marked eyelid edema. It is an ophthalmic emergency because corneal ulceration and perforation can occur and systemic antimicrobial treatment is required.

123. A. Duodenal atresia presents soon after birth with bilious vomiting and the double-bubble sign. It is associated with trisomy 21 and other congenital anomalies and requires surgical correction after stabilization.

124. C. DIC is a secondary process in severe illness such as sepsis or asphyxia. Widespread coagulation activation consumes platelets and factors and generates fibrin degradation products, producing simultaneous thrombosis risk and bleeding.

125. A. The subgaleal space can hold a large fraction of a neonate's blood volume. A diffuse fluctuant swelling crossing sutures with pallor, tachycardia, falling hematocrit, or shock after vacuum delivery is a medical emergency.

126. C. Postresuscitation care includes close monitoring of temperature, respiratory and cardiovascular status, glucose, and neurologic function. Hypoglycemia can compound brain injury and should be recognized and treated rather than assumed to be limited to infants of diabetic mothers.

127. C. Obstructed total anomalous pulmonary venous return causes severe pulmonary venous congestion, edema, and hypoxemia and is a surgical emergency. Obstruction prevents adequate return of oxygenated pulmonary venous blood to the systemic circulation.

128. B. Caffeine is standard therapy for apnea of prematurity because it stimulates respiratory drive and improves diaphragmatic function. Before attributing apnea to prematurity, clinicians should evaluate for secondary causes such as infection, anemia, temperature instability, and metabolic abnormalities.

129. C. Noonan syndrome can resemble Turner syndrome phenotypically but occurs in all sexes and is usually caused by RAS-MAPK pathway variants. Pulmonary valve stenosis and hypertrophic cardiomyopathy are important associated cardiac findings.

130. D. Gastric residuals are common and nonspecific in preterm infants. Feeding decisions should integrate abdominal findings, stool characteristics, cardiorespiratory stability, emesis, laboratory data, and imaging when indicated rather than rely on residual volume alone.

131. D. Loop diuretics increase urinary calcium excretion, and preterm infants have multiple additional risk factors for nephrocalcinosis. Renal ultrasound may show medullary echogenicity; many cases improve over time, but renal follow-up may be needed.

132. A. Urea-cycle disorders can present after protein feeding begins with severe hyperammonemia, encephalopathy, vomiting, and respiratory alkalosis from central hyperventilation. Hyperammonemia is a metabolic emergency requiring immediate treatment while the specific diagnosis is established.

133. C. Maternal varicella around the time of delivery can expose the infant to virus before sufficient protective maternal antibody has been transferred. Severe neonatal disease can occur, so exposed infants require risk-based prophylaxis and close management.

134. B. Gastroschisis is usually right of the umbilicus and lacks a protective sac, leaving bowel exposed to amniotic fluid. Initial care includes protecting bowel, preventing heat and fluid loss, gastric decompression, and surgical management.

135. B. Hemophilia A is an X-linked factor VIII deficiency that produces an isolated prolonged aPTT with normal platelets and PT. Neonates can present with bleeding after procedures, cephalohematoma, or intracranial hemorrhage.

136. C. Erb palsy follows injury to the upper brachial plexus, usually C5-C6, and causes the classic waiter's-tip posture. Preserved grasp suggests lower plexus function is intact; most stretch injuries improve, but serial examination is important.

137. B. Effect modification or interaction occurs when the association between treatment and outcome differs across levels of another variable. This is conceptually distinct from confounding and may have important clinical implications if prespecified and supported by credible data.

138. A. Cocaine increases sympathetic activity and vasoconstriction, which can compromise uteroplacental blood flow and increase the risk of placental abruption. The acute obstetric event can lead to fetal hypoxia, prematurity, and neonatal complications.

139. B. CHARGE is an acronym describing a characteristic cluster of coloboma, heart defects, choanal atresia, growth or developmental problems, genital abnormalities, and ear anomalies. CHD7 variants are a common molecular cause.

140. B. Most fetal calcium and phosphorus accretion occurs late in gestation, so very preterm infants begin life with limited mineral stores. Inadequate calcium or phosphorus delivery, prolonged parenteral nutrition, immobility, and some medications contribute to metabolic bone disease.

141. C. Distal RTA results from impaired distal hydrogen ion secretion, producing a hyperchloremic metabolic acidosis with inappropriately high urine pH. Chronic acidosis and alkaline urine promote nephrocalcinosis and can impair growth.

142. B. Fatty-acid oxidation defects impair generation of energy and ketone bodies during fasting. Hypoketotic hypoglycemia during illness or prolonged fasting is a key diagnostic clue and can be accompanied by liver dysfunction, cardiomyopathy, or rhabdomyolysis depending on the disorder.

143. D. Enteroviruses can cause severe neonatal infection, particularly when acquired perinatally, with sepsis-like illness, myocarditis, hepatitis, coagulopathy, meningoencephalitis, and high mortality in severe cases.

144. D. Hereditary spherocytosis can present in neonates with jaundice and anemia. A family history of hemolysis or splenectomy and spherocytes with a negative direct antiglobulin test support a membrane disorder rather than immune hemolysis.

145. B. Pyridoxine-dependent epilepsy is an important treatable cause of refractory neonatal seizures. A dramatic response to pyridoxine supports the diagnosis, although treatment should occur with cardiorespiratory monitoring because apnea can occur during administration.

146. A. An ROC curve plots sensitivity against 1 minus specificity across possible thresholds. The area under the curve summarizes discrimination, but the clinically preferred threshold also depends on consequences of false positives and false negatives.

147. A. An early, global insult can reduce cell number throughout the fetus and produce symmetric fetal growth restriction. Later placental insufficiency more often produces asymmetric growth restriction with relative head sparing.

148. C. As pulmonary vascular resistance falls after birth, a large VSD permits increasing left-to-right flow, leading to pulmonary overcirculation and congestive heart failure. Symptoms therefore often emerge over the first several weeks rather than immediately at birth.

149. D. In congenital diaphragmatic hernia, bag-mask ventilation can inflate herniated bowel and further compromise the hypoplastic lung. Initial stabilization includes prompt intubation, gastric decompression, gentle ventilation, and careful management of pulmonary hypertension.

150. C. Adrenal insufficiency can contribute to refractory hypotension, particularly in critically ill preterm infants, and may be accompanied by electrolyte disturbances. Diagnosis is challenging and should be based on the clinical context and appropriate endocrine assessment.

151. A. Coagulase-negative staphylococci are common causes of late-onset bloodstream infection in very preterm infants with central lines. Diagnosis requires careful interpretation because these organisms are also common skin contaminants.

152. B. Transient abnormal myelopoiesis occurs in some neonates with trisomy 21 and is linked to GATA1 mutations. It often regresses spontaneously but can cause serious organ complications and is associated with later risk of myeloid leukemia of Down syndrome.

153. A. A bright or alert infant with profound weakness, areflexia, and tongue fasciculations suggests a lower motor neuron disorder such as spinal muscular atrophy. Newborn screening and rapid molecular diagnosis are crucial because disease-modifying therapy is time sensitive.

154. C. Run-chart rules use nonrandom patterns such as sustained shifts or trends to identify possible special-cause variation. A shift after an intervention supports the possibility of real process change, although teams should still examine context and other concurrent changes.

155. A. Appropriate intrapartum antibiotic prophylaxis substantially reduces early-onset GBS disease by lowering exposure during labor and delivery. It does not reliably prevent late-onset GBS disease or unrelated congenital and nosocomial infections.

156. C. Ebstein anomaly is characterized by apical displacement of the tricuspid valve leaflets, atrialization of the right ventricle, and often severe tricuspid regurgitation. The severity of neonatal cyanosis depends on right-ventricular function and pulmonary blood flow.

157. A. Inhaled nitric oxide reaches ventilated lung regions and can improve ventilation-perfusion matching by selectively lowering pulmonary vascular resistance. It is used in selected term or near-term infants with hypoxemic respiratory failure and PPHN after adequate lung recruitment and stabilization.

158. D. VACTERL association is considered when several of the characteristic vertebral, anorectal, cardiac, tracheoesophageal, renal, and limb anomalies coexist and another specific syndrome does not better explain the findings. Evaluation should search systematically for all component defects.

159. C. Preterm infants have lower iron stores and rapid postnatal growth, so enteral iron supplementation is commonly needed once clinically appropriate. Exact dosing depends on birth weight, diet, transfusion history, and local guidance.

160. D. Renal vein thrombosis classically presents with hematuria, thrombocytopenia, and an enlarged kidney, although the full triad is not always present. Risk factors include dehydration, sepsis, maternal diabetes, and intravascular catheters.

161. B. Hyperekplexia presents in neonates with an exaggerated startle reflex and episodes of generalized stiffness, sometimes with dangerous apnea. Normal ictal EEG and stimulus-triggered rigidity help distinguish it from epilepsy.

162. B. Anti-Kell antibodies can cause severe fetal anemia through both hemolysis and suppression of erythropoiesis because Kell antigens are expressed on erythroid precursors. This can produce substantial anemia even when reticulocytosis is less prominent than expected.

163. A. Adenosine transiently blocks AV nodal conduction and is first-line acute therapy for many regular narrow-complex reentrant SVTs when vagal maneuvers fail and the infant is stable. Synchronized cardioversion is used when significant hemodynamic instability is present.

164. A. Pulmonary hemorrhage presents with acute bloody airway secretions, respiratory deterioration, and diffuse infiltrates. Risk factors include extreme prematurity, severe RDS, and a hemodynamically significant PDA or sudden increase in pulmonary blood flow.

165. D. Osteogenesis imperfecta is a collagen disorder with bone fragility, fractures, osteopenia, and in some forms blue sclerae. Severity ranges from mild familial disease to lethal perinatal forms.

166. C. Immature kidneys can waste sodium, and total-body sodium depletion may contribute to poor growth even when serum sodium is only modestly low. Selected preterm infants benefit from assessment of sodium balance and individualized supplementation.

167. B. Extreme prematurity, central venous catheters, broad-spectrum antibiotics, and other intensive-care exposures increase the risk of invasive candidiasis. Persistent sepsis with thrombocytopenia or unexplained hyperglycemia should raise suspicion when bacterial cultures and therapy do not explain the course.

168. C. Acute bilirubin encephalopathy can evolve from lethargy and hypotonia to irritability, high-pitched cry, hypertonia, retrocollis, and opisthotonus. Prevention requires timely recognition and treatment of severe hyperbilirubinemia.

169. B. For selected patients who remain at high risk of preterm delivery after an earlier course, a single repeat or rescue course may be considered according to obstetric guidance. Repeated scheduled weekly courses are not routine, and the purpose remains fetal maturation rather than infection prevention.

170. D. When pulmonary blood flow depends on the ductus, prostaglandin E1 maintains patency until catheter-based or surgical intervention can establish a more durable source of pulmonary blood flow. Closing the ductus in this setting can be catastrophic.

171. A. The respiratory time constant equals resistance multiplied by compliance. Lung units with long time constants fill and empty slowly; if the next breath begins before expiration is complete, gas trapping and inadvertent PEEP can develop.

172. D. Achondroplasia is caused by activating FGFR3 variants that inhibit chondrocyte proliferation in the growth plate. The phenotype includes rhizomelic limb shortening, macrocephaly or frontal bossing, and midface hypoplasia.

173. D. Safe oral feeding depends on coordinated sucking, swallowing, breathing, and airway protection. Recurrent coughing, desaturation, or bradycardia during feeds suggests discoordination or aspiration risk and calls for feeding assessment rather than simply faster flow.

174. B. Meningitis can lead to ventriculitis, ependymal injury, and impaired CSF circulation or resorption, causing hydrocephalus. Serial head circumference and neuroimaging help identify this complication during and after severe neonatal CNS infection.

175. B. Conotruncal and aortic arch abnormalities, hypocalcemia from parathyroid hypoplasia, and thymic hypoplasia are classic features of 22q11.2 deletion syndrome. The combination should prompt genetic evaluation and assessment of immune and calcium status.

176. A. Volume-targeted ventilation adjusts pressure to achieve a selected tidal volume, allowing pressure to fall when compliance improves. This can reduce excessive tidal volumes and may lessen ventilator-induced lung injury compared with fixed pressure strategies.

177. D. Thanatophoric dysplasia is a severe FGFR3-related skeletal dysplasia characterized by profound limb shortening, narrow thorax, macrocephaly, and pulmonary hypoplasia. It is usually lethal in the perinatal period without extraordinary respiratory support.

178. B. Preterm growth quality depends on adequate protein as well as energy. Calories without sufficient protein can promote fat gain while limiting lean tissue, length, and head growth, so nutrient composition should be reviewed rather than simply increasing energy density.

179. B. Neonatal seizures frequently become electroclinical-dissociated, especially after treatment, so EEG or continuous EEG is important for detection and treatment assessment. Clinical events alone both miss true seizures and can misclassify nonseizure movements.

180. A. In persistent pulmonary hypertension, elevated pulmonary vascular resistance can cause right-to-left shunting through the ductus. Deoxygenated blood enters the descending aorta, creating lower postductal than preductal saturation when the right arm is supplied proximal to the ductal insertion.

181. A. CPAP provides distending pressure throughout the respiratory cycle, stabilizing the chest wall and alveoli, maintaining functional residual capacity, and improving oxygenation. It does not eliminate airway resistance or suppress surfactant production.

182. B. Congenital myotonic dystrophy is typically transmitted by an affected mother and may be severe because of repeat expansion. Neonates can have marked hypotonia, respiratory failure, facial weakness, and feeding difficulty; maternal myotonia is an important diagnostic clue.

183. B. Human milk contains limited vitamin D, and preterm infants require adequate vitamin D along with calcium and phosphorus to support bone health. Supplementation is individualized according to feeding regimen and local recommendations.

184. C. Large IVH can obstruct CSF pathways or impair resorption, leading to progressive posthemorrhagic ventricular dilatation. Serial ultrasound measurements and head circumference help determine whether neurosurgical drainage or shunting strategies are needed.

185. B. There is no single blood-pressure number that defines inadequate perfusion in every preterm infant. Management should integrate blood pressure trends with capillary refill, urine output, lactate, ventricular function, and the overall clinical context.

186. C. Congenital lobar overinflation causes hyperexpansion of one or more lobes, often from deficient bronchial cartilage or airway obstruction. Mass effect can compress adjacent lung and shift the mediastinum, producing respiratory distress.

187. A. Prolonged parenteral nutrition, lack of enteral stimulation, infection, and nutrient imbalances contribute to cholestasis. Advancing enteral feeds as tolerated and optimizing parenteral nutrition composition can reduce risk; evaluation must also exclude other causes of direct hyperbilirubinemia.

188. A. Weight can change rapidly with fluid status, so growth assessment should include anthropometric trends and clinical context. Length and head circumference help evaluate tissue growth, while edema or fluid shifts can make weight alone misleading.

189. B. Persistently low saturation fails pulse-oximetry screening even when the preductal-postductal difference is small. The infant requires prompt clinical evaluation for critical congenital heart disease and other important causes of hypoxemia such as pulmonary disease or infection.

190. B. Congenital pulmonary airway malformation is a developmental lesion composed of abnormal cystic lung tissue and typically receives pulmonary arterial supply. Sequestration instead lacks normal bronchial connection and usually receives systemic arterial supply.

191. C. In truncus arteriosus, pulmonary and systemic circulations share a common outflow. As pulmonary vascular resistance falls, pulmonary overcirculation increases and congestive heart failure may become prominent. Definitive management is surgical repair.

192. A. Bronchopulmonary sequestration consists of nonfunctioning lung tissue without normal airway communication and with systemic arterial blood supply, often from the aorta. The anomalous feeding vessel is a key diagnostic feature.

193. D. Balloon valvuloplasty is a primary catheter-based treatment for critical valvar pulmonary stenosis when anatomy is suitable. Prostaglandin may maintain ductal pulmonary flow during stabilization, but it does not relieve the fixed valvar obstruction itself.

194. B. Chylothorax results from leakage of lymphatic fluid into the pleural space and may be congenital or postoperative. After fat-containing feeds, the fluid becomes milky and typically has high triglycerides and a lymphocyte predominance.

195. C. Congenital central hypoventilation syndrome is characterized by inadequate autonomic ventilatory responses, especially during sleep, and is strongly associated with PHOX2B variants. Affected infants may require long-term ventilatory support and evaluation for associated autonomic disorders.

196. C. Compliance describes change in volume for a given change in pressure. Stiff, edematous, or atelectatic lungs have low compliance, so greater pressure is needed to generate a given tidal volume.

197. D. Successful extubation requires sufficient respiratory drive and lung function to maintain ventilation and oxygenation on noninvasive support. Severe apnea, worsening gas exchange, or absence of spontaneous breathing argues against readiness.

198. D. High-frequency modes use rapid oscillations or jets with small tidal volumes, but appropriate lung recruitment and mean airway pressure remain essential. Gas exchange and lung inflation still require close monitoring to avoid underinflation or overdistention.

199. B. Nitric oxide can oxidize hemoglobin iron from the ferrous to the ferric state, forming methemoglobin, which cannot bind oxygen normally. Methemoglobin levels are therefore monitored during inhaled nitric oxide therapy.

200. A. Meconium aspiration often creates heterogeneous obstruction and overdistention, increasing air-leak risk. Gentle ventilation that limits volutrauma and allows adequate expiration is preferred while supporting oxygenation and treating associated pulmonary hypertension.