COMPREHENSIVE MOCK EXAM 7

Supplemental to: Neonatology Board Review 2026-2027

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American Board of Pediatrics Neonatal-Perinatal Medicine Subspecialty Certifying Examination — single-best-answer A-D format, domain-weighted to the ABP initial-certification content outline

Board-Review Questions

1. A term infant born through meconium-stained fluid has coarse breath sounds, hypoxemia, and a chest radiograph showing patchy bilateral infiltrates with hyperinflation. Which mechanism contributes importantly to this disease?

A. Diffuse atelectasis from primary surfactant deficiency

B. Congenital deficiency of alpha-1 antitrypsin in alveolar macrophages

C. Airway obstruction with ball-valve air trapping and chemical pneumonitis

D. Isolated pulmonary venous hypertension without parenchymal injury

2. A preterm infant has recurrent apnea and bradycardia. During one event, the ECG shows a narrow-complex rhythm at 280/min with abrupt onset and offset. Which diagnosis is most likely?

A. Ventricular fibrillation

B. Supraventricular tachycardia

C. Complete atrioventricular block

D. Sinus tachycardia from anemia

3. A breastfed 32-week infant is approaching discharge after a prolonged NICU stay. Which nutrient commonly requires specific supplementation in preterm infants because stores are limited and growth needs are high?

A. Pharmacologic vitamin C supplementation

B. Vitamin D supplementation for bone mineralization

C. Enteral iron supplementation for prematurity

D. Sodium supplementation for renal salt losses

4. A very preterm infant has bilateral cystic lesions in the periventricular white matter on follow-up cranial ultrasound. Which long-term neurologic outcome is most strongly associated with this injury?

A. Isolated peripheral facial palsy

B. Progressive muscular dystrophy

C. Congenital sensorineural deafness without motor impairment

D. Spastic diplegic cerebral palsy

5. A newborn has conotruncal heart disease, hypocalcemia, and a history of polyhydramnios due to poor fetal swallowing. Which genetic disorder should be strongly considered?

A. Congenital nephrotic syndrome

B. Beckwith-Wiedemann syndrome

C. Achondroplasia

D. 22q11.2 deletion syndrome

6. A 29-week infant is delivered after severe maternal hypertension, fetal growth restriction, and absent end-diastolic flow in the umbilical artery. The infant is small for gestational age and initially stable. Which hematologic abnormality is most likely during the first day of life?

A. Neutropenia related to placental insufficiency

B. Persistent lymphocytosis caused by chronic hypoxemia

C. Neutrophilia from fetal inflammatory activation

D. Isolated eosinophilia from maternal hypertension

7. A term newborn develops severe sepsis, coagulopathy, hepatitis, and myocarditis during the first week of life. Maternal illness was a nonspecific febrile syndrome near delivery. Which viral pathogen is a recognized cause of this fulminant neonatal presentation?

A. Perinatal human papillomavirus infection

B. Fulminant neonatal enterovirus infection

C. Neonatal rhinovirus respiratory infection

D. Congenital parvovirus B19 infection

8. A newborn has persistent hypoglycemia requiring a very high glucose infusion rate. During hypoglycemia, insulin is detectable, ketones are suppressed, and free fatty acids are low. Which diagnosis is most likely?

A. Adrenal insufficiency with marked ketone production

B. Isolated growth hormone excess

C. Congenital hyperinsulinism

D. Glycogen storage disease with appropriate ketosis

9. A study is designed to detect a clinically important reduction in severe intraventricular hemorrhage but enrolls far fewer infants than planned. The study finds no statistically significant difference. Which error becomes more likely?

A. Verification bias from missing a reference test

B. Publication bias within a single enrolled cohort

C. Type I error from repeated significance testing

D. Type II error from inadequate statistical power

10. A newborn with polyuria has hypernatremia and very dilute urine despite dehydration. Serum glucose is normal. Which diagnosis should be considered?

A. Diabetes insipidus with impaired urinary concentration

B. Congenital chloride diarrhea with concentrated urine

C. Syndrome of inappropriate antidiuretic hormone secretion

D. Acute oliguric kidney injury from perinatal asphyxia

11. A newborn who did not receive vitamin K prophylaxis develops gastrointestinal bleeding on day 5. Prothrombin time is markedly prolonged. Which diagnosis is most likely?

A. Fetal and neonatal alloimmune thrombocytopenia

B. Vitamin K deficiency bleeding

C. Congenital factor XII excess

D. Disseminated intravascular coagulation from severe sepsis

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

A. Administer a rapid normal saline bolus before ventilation

B. Begin effective positive-pressure ventilation promptly

C. Start chest compressions with a three-to-one ratio

D. Give intravenous epinephrine through an umbilical line

13. A 40-week infant with persistent pulmonary hypertension is mechanically ventilated. Which blood-gas target is generally preferable to deliberate profound alkalosis?

A. Near-normal pH and carbon dioxide with adequate oxygen delivery

B. Sustained severe hypocapnia with pH above 7.60

C. Higher oxygen tension to maximize pulmonary vasodilation

D. Persistent metabolic alkalosis induced with bicarbonate infusions

14. A preterm infant with necrotizing enterocolitis develops sudden worsening abdominal distention and free intraperitoneal air. What does this finding indicate?

A. Isolated lactose intolerance without bowel injury

B. Improving bowel motility after decompression

C. Intestinal perforation requiring urgent surgical evaluation

D. Normal intraluminal gas in a premature intestine

15. A very preterm infant has low serum IgG compared with a term newborn. Which physiologic fact best explains this difference?

A. IgG cannot cross the placenta at any gestational age

B. Most transplacental IgG transfer occurs late in gestation

C. Preferential placental transfer of maternal IgM

D. Preterm infants destroy maternal IgG immediately after delivery

16. Which neonatal factor is most strongly associated with later motor impairment in an extremely preterm infant?

A. Severe intraventricular hemorrhage or cystic white-matter injury

B. Brief use of nasal cannula oxygen after feeding

C. Transient physiologic jaundice below treatment thresholds

D. Uncomplicated closure of the ductus arteriosus

17. A 25-week infant has retinal vascular changes detected on scheduled screening. Which neonatal exposure is a major modifiable risk factor for retinopathy of prematurity?

A. Deferred cord clamping in a stable infant

B. Routine vitamin K prophylaxis

C. Human-milk feeding

D. Excessive or highly variable oxygen exposure

18. A neonate receiving prostaglandin E1 for ductal-dependent congenital heart disease develops recurrent apnea. What is the most appropriate interpretation?

A. Apnea is a recognized adverse effect and airway support may be required

B. The medication should be replaced with indomethacin immediately

C. Apnea indicates new bacterial meningitis until proven otherwise

D. Apnea proves the ductus has closed despite the infusion

19. A 34-week infant tires during oral feeding, has desaturations, and loses milk from the mouth despite good hunger cues. Which developmental issue most likely explains the problem?

A. Immature protective airway reflexes during oral feeding

B. Inability to digest lactose before 40 weeks gestation

C. Immature coordination of sucking, swallowing, and breathing

D. Excessive esophageal peristalsis causing rapid gastric emptying

20. A 29-week infant on CPAP has increasing oxygen need and work of breathing. Radiography is consistent with respiratory distress syndrome. Which intervention most directly treats the underlying deficiency?

A. Give inhaled nitric oxide to replace endogenous surfactant

B. Begin loop diuretics to remove alveolar protein immediately

C. Increase inspired oxygen to 100% without changing ventilation

D. Administer exogenous surfactant using an appropriate airway technique

21. A neonate exposed to maternal magnesium sulfate is hypotonic and has weak respiratory effort immediately after birth. Which mechanism best explains the findings?

A. Neuromuscular depression from elevated neonatal magnesium levels

B. Central serotonin excess causing sustained muscle contraction

C. Increased acetylcholine release at the neuromuscular junction

D. Permanent anterior horn cell loss from magnesium toxicity

22. A newborn has cleft lip and palate, postaxial polydactyly, microphthalmia, and severe holoprosencephaly. Which diagnosis is most likely?

A. Trisomy 18

B. Turner syndrome

C. Trisomy 13

D. Williams syndrome

23. A mother with severe fetal anemia from red-cell alloimmunization undergoes serial intrauterine transfusions. The neonate is born with a relatively low reticulocyte count despite anemia. Which mechanism best explains this finding?

A. Enhanced marrow erythroid hyperplasia from donor red cells

B. Suppression of fetal erythropoiesis after repeated transfusion

C. Acute hemolysis caused by neonatal vitamin K deficiency

D. Increased erythropoietin secretion from persistent hypoxemia

24. A newborn has snuffles, hepatosplenomegaly, rash involving the palms and soles, and long-bone abnormalities. Which infection is most likely?

A. Congenital syphilis

B. Neonatal candidiasis

C. Congenital cytomegalovirus

D. Congenital toxoplasmosis

25. A very preterm infant receives gentamicin for suspected sepsis. Compared with a term infant, which pharmacokinetic feature most often requires a longer dosing interval?

A. Accelerated hepatic metabolism through fully mature enzymes

B. Complete gastrointestinal absorption of parenteral gentamicin

C. Smaller extracellular fluid volume causing rapid drug elimination

D. Reduced renal clearance because glomerular filtration is immature

26. A medication error reaches a newborn but causes no apparent harm. Which professional response is most appropriate?

A. Conceal the error because no injury is currently visible

B. Alter the medical record so the dose appears correct

C. Disclose the error and perform a safety-focused systems review

D. Blame the individual nurse without reviewing system factors

27. A 38-week infant is visibly jaundiced at 18 hours of life. What is the most appropriate next step?

A. Observe briefly because early transitional jaundice can occur

B. Start exchange transfusion without measuring bilirubin

C. Measure bilirubin promptly and evaluate for pathologic early jaundice

D. Schedule bilirubin reassessment with routine outpatient follow-up

28. A newborn has sharply demarcated areas of absent skin over the scalp without inflammation. Which diagnosis is most likely?

A. Miliaria crystallina

B. Erythema toxicum neonatorum

C. Harlequin ichthyosis

D. Aplasia cutis congenita

29. A newborn has cyanosis, a harsh systolic ejection murmur at the left upper sternal border, and a boot-shaped cardiac silhouette. Pulmonary blood flow decreases as the ductus closes. Which anatomic combination defines the underlying lesion?

A. Pulmonary venous obstruction, intact ventricular septum, and right atrial hypoplasia

B. Atrial septal defect, mitral stenosis, left ventricular hypertrophy, and aortic dilation

C. Ventricular septal defect, overriding aorta, pulmonary stenosis, and right ventricular hypertrophy

D. Complete atrioventricular canal, pulmonary hypertension, and left ventricular outflow obstruction

30. A 26-week infant develops cystic lucencies tracking along bronchovascular bundles after several days of positive-pressure ventilation. Which diagnosis is most likely?

A. Pulmonary sequestration supplied by a systemic artery

B. Congenital pulmonary airway malformation presenting de novo

C. Pulmonary interstitial emphysema from alveolar air leak

D. Transient tachypnea from delayed fetal fluid absorption

31. A preterm infant has repeated stress-related hyperglycemia but no ketosis and no glucosuria at modest glucose concentrations. Which first management step is usually appropriate?

A. Reduce amino acid delivery while evaluating metabolic stress

B. Start lifelong insulin therapy without reviewing glucose delivery

C. Increase parenteral dextrose to suppress endogenous glucose production

D. Reassess glucose infusion rate and treat contributing illness or medications

32. A clinical trial reports a treatment effect with a 95% confidence interval that crosses the null value. What is the most appropriate statistical interpretation?

A. There is a 95% probability that the null hypothesis is true

B. The study establishes clinical equivalence between the treatments

C. The data are compatible with no difference at the conventional 0.05 level

D. The observed effect must be clinically important despite imprecision

33. A 24-week infant has progressive anemia at six weeks of life with low reticulocyte production and no evidence of hemolysis or bleeding. Which process best explains the anemia?

A. Autoimmune hemolysis from maternal anti-D antibodies developing after birth

B. Anemia of prematurity from low erythropoietin response and phlebotomy losses

C. Polycythemia caused by delayed cord clamping

D. Vitamin K deficiency causing isolated red-cell destruction

34. A term infant has persistent non-anion-gap metabolic acidosis, alkaline urine, and nephrocalcinosis. Which renal disorder is most likely?

A. Distal renal tubular acidosis

B. Nephrogenic diabetes insipidus

C. Proximal tubular glucose reabsorption excess

D. Acute poststreptococcal glomerulonephritis

35. A 26-week infant has just begun parenteral nutrition. Why are amino acids generally introduced early rather than withheld for several days?

A. To reduce catabolism and support positive nitrogen balance

B. To reduce dependence on enteral protein during early feeding advancement

C. To eliminate the risk of parenteral cholestasis

D. To stabilize glucose concentrations during early parenteral nutrition

36. A newborn with myelomeningocele develops progressive head enlargement, a bulging fontanelle, and downward deviation of the eyes. Which complication is most likely?

A. Congenital myotonic dystrophy

B. Hydrocephalus associated with Chiari II malformation

C. Isolated brachial plexus palsy

D. Benign enlargement of the subarachnoid spaces

37. A newborn has ambiguous genitalia, severe hypotonia, and dysmorphic features. The team is considering chromosomal and single-gene disorders. What is the most appropriate general first step in genetic evaluation?

A. Obtain a detailed family history and phenotype-directed genetic testing strategy

B. Assume the genital findings establish a specific karyotype

C. Begin with conventional karyotyping and defer sequencing until later

D. Order broad exome sequencing before completing the phenotype assessment

38. A vigorous 38-week infant is born after an uncomplicated vaginal delivery and does not need resuscitation. Which umbilical cord strategy is most consistent with current neonatal resuscitation guidance?

A. Reserve delayed clamping for infants below 35 weeks gestation

B. Defer cord clamping for at least about 60 seconds

C. Clamp the cord immediately to prevent neonatal polycythemia

D. Milk the intact cord routinely instead of delaying clamping

39. A 27-week infant develops grunting, retractions, and increasing oxygen requirement minutes after birth. Chest radiography shows low lung volumes with diffuse reticulogranular opacities and air bronchograms. Which process is primarily responsible?

A. Congenital absence of pulmonary lymphatic drainage

B. Pulmonary venous obstruction causing isolated interstitial edema

C. Excess surfactant causing air trapping and hyperinflation

D. Deficient surfactant causing alveolar collapse and low compliance

40. A preterm infant has direct hyperbilirubinemia, pale stools, and dark urine. Which feature makes physiologic jaundice unlikely?

A. Exclusive human-milk feeding with adequate growth

B. Conjugated hyperbilirubinemia with acholic stools

C. Indirect bilirubin below treatment thresholds

D. Jaundice appearing after the first day of life

41. A 2-day-old infant has shock and severe cyanosis. Echocardiography shows hypoplastic left ventricle, mitral atresia, and aortic atresia. Which circulation is necessary for systemic blood flow before surgical palliation?

A. Right ventricular output through a patent ductus arteriosus

B. Antegrade flow through a normally sized ascending aorta

C. Left ventricular output through a restrictive ventricular septal defect

D. Pulmonary venous return through the coronary sinus

42. A growth-restricted fetus has abnormal Doppler studies showing reversed end-diastolic flow in the umbilical artery. Which interpretation is most appropriate?

A. This indicates severe placental vascular resistance and fetal compromise

B. This is a normal late-gestation adaptation in small fetuses

C. This finding predicts excessive placental transfusion at delivery

D. This primarily reflects fetal cerebral vasoconstriction

43. A preterm infant has worsening oxygen requirement and increased tracheal secretions while mechanically ventilated. A tracheal aspirate grows bacteria, but the infant has no systemic instability and chest imaging is unchanged. What is the best interpretation?

A. The culture most likely reflects contamination during specimen collection

B. Continue antibiotics until the endotracheal tube is removed

C. Any positive tracheal culture proves bacterial pneumonia

D. Airway colonization must be distinguished from true ventilator-associated infection

44. A 25-week infant on parenteral nutrition has rising triglycerides after an increase in intravenous lipid infusion. What is the most appropriate response?

A. Increase the lipid infusion because hypertriglyceridemia indicates deficiency

B. Ignore the value because preterm infants cannot develop lipid intolerance

C. Reduce amino acids and increase lipid as the principal calorie source

D. Reduce or temporarily adjust lipid delivery while reassessing tolerance

45. A term infant is intubated for severe respiratory failure. End-tidal carbon dioxide remains absent, breath sounds are heard mainly over the stomach, and the heart rate is falling. What is the most likely problem?

A. Persistent pulmonary hypertension despite effective ventilation

B. Esophageal intubation requiring immediate airway correction

C. Spontaneous pneumomediastinum with correct tube placement

D. Right mainstem bronchial intubation with normal carbon dioxide tracing

46. A very preterm infant becomes cold during transport. Which physiologic feature makes this infant especially vulnerable to hypothermia?

A. Excess subcutaneous fat with high thermal conductivity

B. Large surface area, thin skin, and limited brown-fat stores

C. Mature vasomotor control that prevents heat conservation

D. Low evaporative water loss through a fully keratinized epidermis

47. A 39-week infant with a history of placental abruption has altered consciousness, hypotonia, weak suck, and recurrent seizures at 3 hours of life. Which diagnosis is most likely?

A. Benign neonatal sleep myoclonus

B. Isolated peripheral nerve injury

C. Hypoxic-ischemic encephalopathy

D. Physiologic transition after uncomplicated birth

48. A former 25-week infant is seen at 18 months chronologic age for developmental assessment. Which age should generally be used when interpreting milestones during early childhood?

A. Postmenstrual age rather than corrected age for milestone interpretation

B. Gestational age at birth without adding postnatal time

C. Corrected age based on the degree of prematurity

D. Chronologic age without adjustment for the degree of prematurity

49. A newborn has petechiae and severe thrombocytopenia. The mother has a normal platelet count, and a previous sibling had intracranial hemorrhage from neonatal thrombocytopenia. Which mechanism is most likely?

A. Maternal autoimmune thrombocytopenia affecting both mother and infant equally

B. Congenital absence of neonatal megakaryocytes from vitamin K deficiency

C. Platelet consumption related to placental transfusion at birth

D. Maternal alloantibodies against a paternally inherited fetal platelet antigen

50. A newborn has a midline neck mass that elevates with tongue protrusion and swallowing. Which embryologic remnant is responsible?

A. First branchial cleft remnant

B. Ectopic cervical thymic tissue

C. Thyroglossal duct remnant

D. Lymphatic malformation from jugular sacs

51. A 3-day-old infant develops poor feeding, tachypnea, weak femoral pulses, cool lower extremities, and metabolic acidosis. The right arm blood pressure is higher than the leg pressure. Which lesion is most likely?

A. Small muscular ventricular septal defect

B. Critical coarctation of the aorta with ductal closure

C. Large secundum atrial septal defect with left-to-right shunt

D. Mild pulmonary valve stenosis with preserved output

52. A neonate with multiple congenital anomalies has a normal conventional karyotype. The abnormalities suggest a submicroscopic deletion or duplication syndrome. Which test is most useful as a broad next diagnostic study?

A. Newborn metabolic screening alone

B. Chromosomal microarray analysis

C. Targeted newborn metabolic screening panel

D. Hemoglobin electrophoresis

53. A quality-improvement team tests a new central-line checklist in one NICU, reviews infection rates monthly, and repeatedly modifies the process. Which framework best describes this activity?

A. Blinded placebo-controlled drug development

B. One-time case-control sampling

C. Repeated cross-sectional prevalence measurements

D. Iterative plan-do-study-act cycles

54. A premature infant has edema, low serum sodium, and rapidly increasing weight after receiving large volumes of hypotonic fluid. Urine output is low. What is the best initial management principle?

A. Give additional free water because the serum sodium is low

B. Reassess fluid intake and restrict free water while evaluating renal function

C. Give hypertonic saline to correct the sodium before adjusting fluids

D. Treat the edema expectantly while maintaining the same fluid prescription

55. A term infant is plethoric, jittery, and tachypneic. Venous hematocrit is 72%. Which complication is most directly related to this finding?

A. Autoimmune neutropenia from maternal antibodies

B. Severe hemophilia from excess red-cell mass

C. Hyperviscosity with impaired tissue perfusion

D. Vitamin K deficiency bleeding

56. A 33-week infant has stridor immediately after extubation but no significant oxygen requirement. The cry is weak, and symptoms worsen with agitation. Which diagnosis should be considered?

A. Post-intubation laryngeal edema or vocal-cord dysfunction

B. Transient tachypnea caused by residual fetal lung fluid

C. Persistent pulmonary hypertension without upper-airway disease

D. Primary surfactant deficiency causing distal airway collapse

57. A 900-g infant is tolerating full human-milk feeds but has poor weight gain and low serum phosphorus. Which nutritional change is most appropriate?

A. Dilute the milk with sterile water to reduce renal solute load

B. Add human-milk fortifier to increase protein and mineral delivery

C. Stop enteral feeds and use intravenous lipid alone

D. Use carbohydrate-rich enteral feeds until weight gain improves

58. A newborn has microcephaly, intracranial calcifications, hepatosplenomegaly, and thrombocytopenia. Maternal records show a primary cytomegalovirus infection during the first trimester. Which mechanism best explains why early gestational infection is associated with more severe fetal injury?

A. Disruption of organogenesis during periods of rapid fetal development

B. Progressive placental impermeability to virus later in pregnancy

C. Increasing maternal antibody transfer that amplifies viral replication

D. Decreased fetal inflammatory responses in the first trimester

59. A newborn has hydrocephalus, chorioretinitis, and diffuse intracranial calcifications. Which congenital infection is most likely?

A. Congenital toxoplasmosis

B. Cytomegalovirus infection

C. Neonatal enterovirus infection

D. Congenital syphilis

60. A 33-week infant develops hypotension after a large pneumothorax has been drained. Echocardiography shows good ventricular contractility and a small, underfilled left ventricle. Which intervention is most appropriate if examination also suggests low intravascular volume?

A. Increase afterload with indomethacin to raise systemic resistance

B. Administer furosemide to reduce presumed pulmonary edema

C. Begin prostaglandin E1 to increase ventricular preload

D. Give a cautious isotonic volume bolus while reassessing perfusion

61. A 28-week infant requires positive-pressure ventilation at birth. The first several inflations produce no chest movement despite a good mask seal and neutral head position. Which adjustment is reasonable to improve ventilation?

A. Increase peak inflation pressure gradually while reassessing chest movement

B. Lower peak inflation pressure to avoid any visible thoracic excursion

C. Begin compressions because preterm lungs are difficult to inflate

D. Use supplemental oxygen while delaying assisted ventilation briefly

62. A 24-week infant has required mechanical ventilation for several weeks. At 36 weeks postmenstrual age, the infant still needs supplemental oxygen and has radiographic areas of hyperinflation and atelectasis. Which diagnosis best fits?

A. Acute transient tachypnea caused by retained fetal lung fluid

B. Bronchopulmonary dysplasia associated with disrupted lung development

C. Isolated bacterial tracheitis without parenchymal lung disease

D. Persistent respiratory distress syndrome from unchanged fetal lungs

63. A newborn has excessive salivation, coughing, and cyanosis with the first feeding. An orogastric tube cannot be advanced into the stomach and coils in the upper chest. Which anomaly is most likely?

A. Hypertrophic pyloric stenosis

B. Hirschsprung disease

C. Duodenal atresia with distal obstruction

D. Esophageal atresia with a distal fistula

64. A term newborn has asymmetric arm movement after a difficult shoulder delivery. The affected arm is adducted and internally rotated with the elbow extended, but hand grasp is preserved. Which injury is most likely?

A. Lower brachial plexus injury involving predominantly C8-T1

B. Upper brachial plexus injury involving C5-C6

C. Bilateral phrenic nerve paralysis

D. Central spinal cord transection

65. A newborn has severe hypotonia and poor feeding. The pregnancy was notable for reduced fetal movement. Later testing shows loss of paternally expressed genes in chromosome region 15q11-q13. Which diagnosis is most likely?

A. Williams syndrome

B. Prader-Willi syndrome

C. DiGeorge syndrome

D. Angelman syndrome

66. A preterm infant has frequent small gastric residuals but a soft abdomen, normal stooling, stable vital signs, and no blood in the stool. What is the best interpretation?

A. Gastric residuals alone are an imperfect marker of serious feeding intolerance

B. Any residual proves necrotizing enterocolitis and requires surgery

C. Residuals show that human milk should be permanently discontinued

D. The finding establishes congenital intestinal obstruction

67. A newborn develops hypocalcemic seizures at 36 hours of life. The infant was born to a mother with diabetes and is otherwise stable. Which electrolyte abnormality should also be considered because it can make hypocalcemia difficult to correct?

A. Hypokalemia from congenital hyperaldosteronism

B. Clinically significant hypomagnesemia

C. Hyperphosphatemia related to reduced renal phosphate excretion

D. Hypernatremia from diabetes insipidus

68. In a neonatal trial, mortality is 20% with standard care and 15% with a new therapy. What is the absolute risk reduction?

A. 5 percentage points

B. 25 percentage points

C. 33 percentage points

D. 75 percentage points

69. A newborn has severe thrombocytopenia at birth, petechiae, and a normal-appearing mother with a normal platelet count. What is the most likely diagnosis?

A. Maternal immune thrombocytopenia with severe maternal thrombocytopenia

B. Fetal and neonatal alloimmune thrombocytopenia

C. Hereditary spherocytosis

D. Vitamin K deficiency bleeding

70. A newborn has unilateral multicystic dysplastic kidney and a normal contralateral kidney with normal blood pressure and renal function. Which management is usually appropriate?

A. Prophylactic chemotherapy to prevent malignant transformation

B. Immediate bilateral nephrectomy

C. Chronic dialysis from the first week of life

D. Ultrasound surveillance and monitoring of the functioning kidney

71. A 31-week infant on nasal CPAP develops recurrent apnea with oxygen desaturation and bradycardia, but evaluation shows no infection, anemia, or seizures. Which treatment is most appropriate?

A. Acetazolamide to induce metabolic alkalosis

B. Morphine to decrease respiratory variability

C. Caffeine citrate to stimulate respiratory drive

D. Propranolol to blunt sympathetic activation

72. A newborn has severe cyanosis. Echocardiography shows all pulmonary veins forming a confluence that drains below the diaphragm, with marked obstruction to pulmonary venous return. Which finding is most likely on chest radiography?

A. Isolated right upper-lobe hyperinflation

B. Diffuse pulmonary edema with a relatively small heart

C. Boot-shaped heart with decreased pulmonary markings

D. Marked cardiomegaly with oligemic lung fields

73. A 32-week infant is born to a mother with systemic lupus erythematosus and anti-Ro/SSA antibodies. The infant has a persistent ventricular rate of 55/min but is otherwise well perfused. Which diagnosis is most likely?

A. Junctional rhythm from fetal magnesium exposure

B. Sinus bradycardia caused by transient neonatal hypothyroidism

C. Long-QT syndrome caused by maternal beta-blocker therapy

D. Congenital complete atrioventricular block from maternal autoantibodies

74. A newborn has vesicular skin lesions, seizures, and elevated liver transaminases on day 7. Maternal history is negative for known herpes infection. What is the best management?

A. Use topical antiviral therapy while awaiting lesion PCR results

B. Wait for maternal serology before beginning antiviral therapy

C. Give intrapartum penicillin to the mother after neonatal symptoms begin

D. Start intravenous acyclovir promptly while obtaining HSV studies

75. An extremely preterm infant has a very low serum albumin concentration and is receiving a highly protein-bound medication. What pharmacologic consequence may occur?

A. Complete inability of the drug to cross any tissue membrane

B. Lower free drug concentration despite unchanged total concentration

C. A larger unbound drug fraction with greater pharmacologic effect

D. More rapid renal elimination because of a higher unbound fraction

76. A term infant with immune-mediated hemolysis has rapidly rising unconjugated bilirubin and develops poor feeding, hypotonia, and a high-pitched cry. What is the major concern?

A. Cholestatic liver disease causing direct bilirubin toxicity

B. Acute bilirubin encephalopathy

C. Physiologic jaundice without neurologic risk

D. Iron deficiency from chronic phlebotomy

77. Parents of an infant born at the threshold of viability are being counseled before delivery. Which communication approach is most appropriate?

A. Avoid discussing uncertainty because it may distress the parents

B. Base the initial recommendation primarily on birth weight and survival statistics

C. Use shared decision-making that explains prognosis, uncertainty, and reasonable care options

D. Present a single mandatory plan without discussing family values

78. A well newborn develops erythematous papules and pustules on the trunk on day 2, sparing the palms and soles. A smear shows eosinophils. What is the most likely diagnosis?

A. Erythema toxicum neonatorum

B. Congenital candidiasis

C. Incontinentia pigmenti

D. Neonatal herpes simplex infection

79. A newborn has rhythmic limb jerking that cannot be stopped by holding the limb and is accompanied by ocular deviation. What is the best interpretation?

A. The movements are typical benign jitteriness

B. The movements are concerning for neonatal seizures

C. The pattern is most consistent with active-sleep motor activity

D. The movements reflect peripheral neuromuscular hyperexcitability

80. A newborn has coloboma, choanal atresia, characteristic external ears, and a conotruncal cardiac defect. Which diagnosis best unifies these findings?

A. CHARGE syndrome

B. Beckwith-Wiedemann syndrome

C. Tuberous sclerosis complex

D. Prader-Willi syndrome

81. A 30-week infant is extubated to noninvasive support after improvement in respiratory distress syndrome. Which modality can provide distending pressure while allowing spontaneous breathing?

A. Negative-pressure ventilation using a neonatal cuirass

B. Nasal continuous positive airway pressure

C. Unhumidified low-flow oxygen without positive pressure

D. Intermittent blow-by oxygen delivered near the face

82. A preterm infant receiving fortified breast milk develops persistent hyponatremia with high urine sodium losses and otherwise normal renal function for gestation. Which nutritional intervention may be appropriate?

A. Restrict sodium intake while reducing total fluid volume

B. Replace feeds with free water until sodium normalizes

C. Use intermittent hypertonic saline supplementation with feeds

D. Provide individualized enteral sodium supplementation

83. A neonate has recurrent catalase-positive bacterial and fungal infections. Testing shows an abnormal oxidative burst in neutrophils. Which diagnosis is most likely?

A. Common variable immunodeficiency

B. Chronic granulomatous disease

C. Severe congenital neutropenia

D. Complement C1 inhibitor deficiency

84. A former 26-week infant has normal gross motor milestones but persistent feeding difficulty, poor growth, and sensory aversion. Which principle is most appropriate?

A. Feeding aversion is primarily gastrointestinal rather than developmental

B. Normal gross motor function excludes meaningful developmental disability

C. Formal cognitive testing later in childhood is the main developmental measure

D. Neurodevelopment includes feeding, sensory, language, cognitive, and motor domains

85. A newborn has profound cyanosis and echocardiography shows pulmonary atresia with an intact ventricular septum. Which immediate physiologic goal is most important?

A. Restrict systemic blood flow to raise right ventricular pressure

B. Close the ductus to reduce pulmonary overcirculation

C. Maintain ductal patency to support pulmonary blood flow

D. Lower systemic vascular resistance with aggressive vasodilators

86. A very preterm infant is clinically stable and approaching the age for retinopathy screening. What determines the timing of the first examination?

A. Current body weight combined with oxygen requirement

B. Whether the infant has already reached term-equivalent age

C. Whether the infant has visible eye redness

D. Gestational age at birth and postnatal age according to screening criteria

87. A 40-week infant remains cyanotic and bradycardic during mask ventilation. The team is uncertain whether the heart rate is 55 or 75/min by auscultation. Chest compressions may be needed. Which method is most useful for rapid continuous heart-rate assessment at this point?

A. Pulse oximetry alone without any cardiac monitoring

B. Frequent umbilical pulse palpation during each ventilation cycle

C. Capillary refill time measured after each ventilation cycle

D. Electrocardiographic monitoring with leads placed promptly

88. A preterm infant develops frequent apnea two days after caffeine was discontinued. The infant has otherwise been stable, but is now hypothermic and feeding less well. What is the best next step?

A. Attribute the events to recurrent apnea of prematurity and restart caffeine

B. Evaluate for a new pathologic trigger such as sepsis before simply restarting caffeine

C. Begin chronic home oxygen without additional evaluation

D. Perform immediate tracheostomy for recurrent respiratory pauses

89. A 28-week infant develops abdominal distention, bloody stools, lethargy, and temperature instability. Abdominal radiography shows pneumatosis intestinalis. Which diagnosis is most likely?

A. Physiologic gastroesophageal reflux

B. Necrotizing enterocolitis

C. Hypertrophic pyloric stenosis

D. Meckel diverticulum with painless bleeding

90. A newborn becomes progressively lethargic and tachypneic on day 3. Ammonia is 600 micromol/L, glucose is normal, and there is respiratory alkalosis without a significant anion-gap acidosis. Which disorder is most likely?

A. Urea cycle defect

B. Distal renal tubular acidosis

C. Diabetic ketoacidosis

D. Methylmalonic acidemia

91. A diagnostic test for neonatal sepsis has very high sensitivity but modest specificity. Which use best fits this property?

A. The test will produce almost no false-positive results

B. A negative result can be useful for ruling out disease when pretest probability is appropriate

C. A positive result is usually sufficient to confirm infection in a symptomatic infant

D. Sensitivity determines how disease prevalence changes after testing

92. A newborn has purpura, thrombocytopenia, hepatosplenomegaly, and a congenital infection. Which mechanism most commonly explains the low platelet count in this setting?

A. Platelet consumption and impaired production during systemic infection

B. Isolated vitamin K deficiency reducing platelet number

C. Excess erythropoietin directly destroying megakaryocytes

D. Transient thrombocytopenia related to perinatal stress

93. A newborn with severe oligohydramnios has Potter facies and profound respiratory failure. Ultrasound shows absent kidneys bilaterally. Which immediate problem is the major cause of death?

A. Pulmonary hypoplasia from prolonged oligohydramnios

B. Pulmonary overcirculation from a large ductus arteriosus

C. Isolated anemia from deficient renal erythropoietin

D. Hyperinsulinemic hypoglycemia from renal agenesis

94. A term newborn is delivered through thick meconium-stained amniotic fluid. The infant is vigorous with good tone and a strong respiratory effort. What is the most appropriate immediate airway approach?

A. Intubate immediately and suction below the vocal cords

B. Administer prophylactic surfactant before spontaneous breathing

C. Provide routine initial care without routine tracheal suctioning

D. Perform deep oropharyngeal suctioning before stimulation

95. A very-low-birth-weight infant develops persistent candidemia despite antifungal therapy. Which additional evaluation is important?

A. Avoid removing potentially infected central venous access

B. Persistent candidemia may represent catheter colonization without invasion

C. Discontinue antifungal treatment once the infant is afebrile

D. Assess for disseminated infection involving organs such as the eye and central nervous system

96. A newborn has electrographic seizures after hypoxic-ischemic injury. Which medication is commonly used as first-line antiseizure therapy in the neonatal period?

A. Phenobarbital as initial antiseizure therapy

B. Acetazolamide as definitive anticonvulsant therapy

C. Ethosuximide for presumed absence seizures

D. Carbamazepine as initial therapy for focal electrographic seizures

97. A newborn has multiple fractures, blue sclerae, and generalized osteopenia. There is a family history of recurrent fractures. Which disorder is most likely?

A. Achondroplasia

B. Osteogenesis imperfecta

C. Congenital muscular dystrophy

D. Hypochondroplasia

98. A 28-week infant has abdominal distention, visible bowel loops, bloody stools, and pneumatosis intestinalis. Which feeding exposure is associated with the lowest risk of this complication?

A. Human milk rather than bovine formula

B. Hyperosmolar clear liquid feeds without milk proteins

C. Concentrated cow-milk formula from the first day of life

D. Delay feeding advancement until later postmenstrual maturation

99. A 36-week infant with meconium aspiration has severe persistent pulmonary hypertension despite optimized ventilation and oxygenation. Blood pressure is adequate. Which therapy selectively lowers pulmonary vascular resistance without causing major systemic vasodilation?

A. Intramuscular epinephrine

B. Inhaled nitric oxide

C. Intravenous nitroprusside

D. Oral propranolol

100. A ventilated 25-week infant acutely deteriorates with bradycardia, hypotension, and asymmetric chest movement. Breath sounds are absent on the right, and transillumination is bright on that side. What is the next step?

A. Immediate decompression of a suspected tension pneumothorax

B. Increase positive end-expiratory pressure to recruit the right lung

C. Wait for a confirmatory chest radiograph before intervening

D. Administer surfactant before evaluating for an air leak

101. A newborn with congenital diaphragmatic hernia remains severely hypoxemic despite optimized ventilation. Which associated physiology commonly drives the refractory hypoxemia?

A. Persistent pulmonary hypertension from hypoplastic pulmonary vasculature

B. Primary surfactant overproduction causing diffuse alveolar flooding

C. Excessive pulmonary blood flow caused by an isolated large atrial defect

D. Complete absence of fetal pulmonary vascular smooth muscle

102. A 25-week infant with a symptomatic patent ductus arteriosus has significant oliguria and rising creatinine. Which pharmacologic option is often preferred when concern about further renal vasoconstriction is high?

A. Indomethacin because it increases renal blood flow

B. Prostaglandin E1 because it constricts ductal smooth muscle

C. Acetaminophen as an alternative ductal-closing agent

D. Dopamine because it directly closes the ductus

103. A 31-week infant has gastroesophageal reflux episodes but is growing well and has no apnea temporally linked to reflux. Which management approach is most appropriate?

A. Begin acid suppression for presumed acid-mediated reflux symptoms

B. Use conservative feeding strategies and avoid routine acid suppression

C. Begin chronic metoclopramide as first-line therapy

D. Pause enteral feeds and provide postpyloric nutrition temporarily

104. A 25-week infant has a sudden drop in hematocrit and increasing apnea on day 3. Cranial ultrasound shows blood within the germinal matrix and lateral ventricle without ventricular dilation. Which diagnosis is most likely?

A. Neonatal arterial ischemic stroke

B. Periventricular leukomalacia

C. Germinal matrix-intraventricular hemorrhage

D. Dandy-Walker malformation

105. A newborn has disproportionately short proximal limbs, macrocephaly, frontal bossing, and a relatively narrow thorax but normal intelligence is expected. Which diagnosis is most likely?

A. Hypophosphatasia

B. Osteogenesis imperfecta type II

C. Achondroplasia

D. Thanatophoric dysplasia

106. A 34-week infant is born after prolonged rupture of membranes. Prenatal ultrasound had shown severe oligohydramnios beginning at 20 weeks. At birth, the infant has a small thorax and severe hypoxemic respiratory failure despite effective ventilation. Which prenatal complication most likely produced this presentation?

A. Transient fetal airway obstruction from swallowed amniotic debris

B. Primary pulmonary lymphangiectasia caused by fetal anemia

C. Delayed surfactant synthesis from maternal corticosteroid exposure

D. Pulmonary hypoplasia from prolonged loss of amniotic fluid

107. A mother is known to be colonized with group B Streptococcus. Which intervention most effectively reduces early-onset GBS disease in the newborn?

A. Immediate neonatal vaccination against group B Streptococcus

B. Antepartum maternal antibiotics after a positive screening culture

C. Appropriate intrapartum maternal antibiotic prophylaxis

D. Short-course neonatal penicillin prophylaxis after birth

108. A newborn screen suggests congenital hypothyroidism. The infant appears clinically normal. Why is prompt confirmatory testing and treatment still important?

A. Clinical symptoms usually become obvious during the first postnatal week

B. Newborn screening cannot detect hypothyroidism before symptoms develop

C. Early thyroid hormone deficiency can cause preventable neurodevelopmental impairment

D. Treatment can be deferred until biochemical abnormalities persist on repeat testing

109. Investigators compare two NICU infection-prevention bundles using a before-and-after design. Infection rates fall after implementation, but hand-hygiene compliance also improved during the same period. What is the major threat to causal inference?

A. Randomization with balanced baseline characteristics

B. Blinding that eliminates secular trends

C. Allocation concealment that prevents cointerventions

D. Confounding by concurrent changes in care

110. A newborn develops marked jaundice after exposure to an oxidant medication. The blood smear shows bite cells and Heinz bodies. Which enzyme deficiency is most likely?

A. Factor VIII deficiency causing a bleeding disorder

B. G6PD deficiency causing oxidant hemolysis

C. Protein C deficiency causing neonatal thrombosis

D. Pyruvate kinase deficiency causing chronic hemolysis

111. A term infant with perinatal asphyxia develops oliguria and rising creatinine. Urinalysis shows granular casts. Which renal lesion is most likely?

A. Acute tubular injury from ischemia

B. Autosomal dominant polycystic kidney disease

C. Congenital nephrotic syndrome

D. Isolated distal renal tubular acidosis

112. During resuscitation of a term infant, the heart rate remains 45/min despite effective ventilation through an endotracheal tube and coordinated chest compressions. An umbilical venous catheter is in place. Which medication is indicated?

A. Intravenous atropine to reverse vagally mediated bradycardia

B. Intravenous calcium gluconate as first-line chronotropic therapy

C. Intravenous epinephrine while ventilation and compressions continue

D. Intravenous adenosine to terminate an occult reentrant rhythm

113. A newborn has abdominal distention and failure to pass meconium. Radiography shows a microcolon and inspissated meconium obstructing the terminal ileum. Which underlying disease should be investigated?

A. Galactosemia

B. Cystic fibrosis

C. Congenital hypothyroidism alone

D. Congenital chloride diarrhea

114. A mechanically ventilated 28-week infant has a $PaCO_2$ of 68 mm Hg, acceptable oxygenation, and chest radiography showing adequate expansion. Which ventilator change most directly increases minute ventilation?

A. Reduce pressure support to decrease alveolar ventilation

B. Decrease ventilator rate while leaving tidal volume unchanged

C. Increase delivered tidal volume or effective ventilator rate

D. Lower inspiratory pressure despite inadequate carbon dioxide clearance

115. A newborn has persistent thrush, severe viral infections, chronic diarrhea, and profound lymphopenia. Which disorder should be considered urgently?

A. Transient neonatal neutrophilia

B. Isolated complement C5 deficiency

C. Hereditary spherocytosis

D. Severe combined immunodeficiency

116. Parents of an extremely preterm infant ask whether a normal cranial ultrasound guarantees normal development. What is the best response?

A. Severe cranial ultrasound abnormalities are the principal determinant of outcome

B. Later follow-up is unnecessary when neonatal imaging is normal

C. Normal imaging is reassuring but does not eliminate risk of later developmental difficulties

D. Normal ultrasound guarantees normal cognition and motor function

117. A very preterm infant has low blood pressure but warm extremities, brisk capillary refill, adequate urine output, and normal lactate. Which principle should guide management?

A. Treat perfusion and end-organ function rather than a numeric pressure alone

B. Start a vasoactive infusion because the systolic pressure is below a numeric threshold

C. Assume occult blood loss whenever mean pressure is below 35 mm Hg

D. Give repeated fluid boluses until the mean pressure exceeds gestational age

118. A newborn has stridor, feeding difficulty, and a high-pitched cry. Flexible laryngoscopy shows an omega-shaped epiglottis with inspiratory collapse of supraglottic tissues. Which diagnosis is most likely?

A. Bilateral choanal atresia causing nasal obstruction

B. Vocal-cord paralysis with fixed cords

C. Subglottic hemangioma causing fixed airway narrowing

D. Laryngomalacia with dynamic supraglottic collapse

119. A 27-week infant's mother is unable to provide sufficient milk. Which enteral substitute is generally preferred over preterm formula when available for a very-low-birth-weight infant?

A. Unmodified whole cow milk

B. Electrolyte solution without protein or fat

C. Rice-based beverage fortified with iron

D. Pasteurized donor human milk

120. A ventilated preterm infant suddenly has markedly diminished breath sounds on the left but remains hemodynamically stable. The endotracheal tube depth is greater than before. What is the most likely cause?

A. Right mainstem bronchial intubation

B. Left tension pneumothorax with severe obstructive shock

C. Pulmonary hemorrhage isolated to the right lung

D. Acute bilateral surfactant inactivation

121. A term infant meets criteria for moderate hypoxic-ischemic encephalopathy at 2 hours of life. Which treatment has the strongest evidence for improving neurodevelopmental outcome?

A. Prophylactic high-dose corticosteroids for seven days

B. Continuous anticonvulsants despite absence of seizures

C. Use controlled hyperventilation to lower $PaCO_2$ during the cooling period

D. Whole-body or selective head therapeutic hypothermia initiated promptly

122. A growth-restricted newborn has a prominent occiput, micrognathia, low-set ears, clenched fists with overlapping fingers, and rocker-bottom feet. Which chromosomal disorder is most likely?

A. Trisomy 18

B. Trisomy 21

C. Noonan syndrome

D. Klinefelter syndrome

123. A term infant is born to a mother with poorly controlled diabetes. The infant is plethoric and macrosomic and develops tachypnea shortly after birth. Which fetal adaptation most directly explains the infant's increased risk for hypertrophic cardiomyopathy?

A. Chronic fetal hyperglucagonemia from placental dysfunction

B. Fetal hyperinsulinemia stimulated by maternal hyperglycemia

C. Reduced fetal insulin-like growth factor signaling

D. Maternal insulin crossing the placenta in high concentrations

124. A newborn has periventricular calcifications, microcephaly, sensorineural hearing loss, and petechiae. Which congenital infection is most likely?

A. Toxoplasma gondii

B. Group B Streptococcus

C. Parvovirus B19

D. Cytomegalovirus

125. A 700-g infant requires a medication dosed in mg/kg. Which prescribing practice best reduces medication error?

A. Calculate from a current verified weight and independently check dose and concentration

B. Use the standard NICU concentration without a second concentration check

C. Scale an established pediatric dose downward by body weight

D. Round the infant's weight to the nearest 5 kg before calculating

126. A newborn's transcutaneous bilirubin is close to the hour-specific phototherapy threshold. What is the most appropriate next test before making a treatment decision?

A. Obtain the direct bilirubin fraction as the confirmatory test

B. Obtain a total serum bilirubin measurement

C. Repeat visual inspection under fluorescent lighting

D. Wait several days because transcutaneous values are definitive

127. A critically ill newborn has a poor prognosis, and the parents request continuation of every possible intervention. The clinical team believes some treatments no longer offer physiologic benefit. What is the best next step?

A. Tell the parents that their values are irrelevant to neonatal decisions

B. Continue current life-sustaining treatments while postponing goal reassessment

C. Limit interventions judged nonbeneficial before another family meeting

D. Hold a structured interdisciplinary meeting to clarify goals, benefits, burdens, and options

128. A very preterm infant has extremely fragile, gelatinous skin with marked transepidermal water loss. Which care strategy is most helpful during the first days?

A. Use a humidified thermal environment and minimize skin trauma

B. Keep the incubator humidity low to prevent edema

C. Apply repeated alcohol cleansing to accelerate keratinization

D. Use a dry skin-care approach to reduce maceration and infection risk

129. A 27-week infant has an echocardiographically large PDA but is clinically stable on minimal respiratory support with good perfusion. What is the best general management principle?

A. Use surgical ligation before considering expectant management

B. Base treatment on clinical hemodynamic significance rather than size alone

C. Begin prostaglandin E1 whenever the ductus is larger than 2 mm

D. Treat a large echocardiographic ductus before clinical symptoms develop

130. A very preterm infant is receiving supplemental oxygen. Which practice best reduces the risk of oxygen-related retinopathy and lung injury?

A. Use pulse oximetry targets and avoid unnecessary sustained hyperoxia

B. Target a higher arterial oxygen tension to create a safety margin

C. Ignore brief oxygen changes because toxicity is unrelated to exposure

D. Maintain oxygen saturation at 100% continuously

131. A newborn has metabolic acidosis, lethargy, vomiting, and a markedly elevated anion gap after feeds are started. Ammonia is normal. Which broad diagnostic category should be strongly considered?

A. Nephrogenic diabetes insipidus

B. Organic acidemia

C. Isolated respiratory alkalosis

D. Urea cycle disorder

132. A cohort study finds an association between prolonged mechanical ventilation and neurodevelopmental impairment in extremely preterm infants. Sicker infants also receive longer ventilation. What type of bias is most important?

A. Lead-time bias from earlier cancer detection

B. Recall bias from inaccurate retrospective exposure reporting

C. Spectrum bias in a diagnostic test

D. Confounding by severity of illness

133. A 28-week infant develops hyponatremia during the second week of life while gaining weight appropriately. Urine sodium is high for the serum sodium. Which physiologic feature of prematurity is most likely responsible?

A. Limited renal tubular sodium reabsorption

B. Excess aldosterone sensitivity causing sodium retention

C. Complete absence of glomerular filtration

D. Reduced antidiuretic hormone secretion with excessive free-water excretion

134. A septic preterm infant has oozing from venipuncture sites, thrombocytopenia, prolonged PT and aPTT, low fibrinogen, and elevated D-dimer. Which diagnosis is most likely?

A. Hereditary spherocytosis with hemolysis

B. Hemophilia A with factor VIII deficiency

C. Disseminated intravascular coagulation

D. Neonatal polycythemia with hyperviscosity

135. A 30-week infant has reached full enteral feeds but is gaining weight slowly. Which measure best assesses whether growth is appropriate over time?

A. Absolute discharge weight compared with a standard target weight

B. One bedside glucose value after a feed

C. A single daily serum sodium concentration

D. Serial weight, length, and head-circumference trajectories

136. A term infant has severe hypotonia, facial weakness, feeding difficulty, and respiratory failure. The mother reports lifelong difficulty releasing her grip after handshakes. Which diagnosis is most likely?

A. Isolated spinal muscular atrophy type 3

B. Congenital myotonic dystrophy

C. Infant botulism acquired at birth

D. Prader-Willi syndrome

137. A newborn has micrognathia, glossoptosis, and a U-shaped cleft palate with episodic upper-airway obstruction when supine. Which sequence best describes this pattern?

A. VACTERL association

B. CHARGE syndrome

C. Pierre Robin sequence

D. Treacher Collins syndrome alone

138. A newborn has a heart rate of 50/min after 30 seconds of ventilation that produces clear bilateral chest movement and a rising oxygen saturation. What is the next step in resuscitation?

A. Continue ventilation alone for another two minutes

B. Defibrillate because profound bradycardia is an unstable rhythm

C. Begin coordinated chest compressions with ventilation

D. Administer naloxone because respiratory depression is likely

139. A term infant has failure to pass meconium, progressive abdominal distention, and explosive stool after rectal examination. Which diagnosis is most likely?

A. Hirschsprung disease

B. Hypertrophic pyloric stenosis

C. Duodenal atresia

D. Necrotizing enterocolitis from prematurity

140. A term newborn has severe respiratory distress, a scaphoid abdomen, and markedly decreased breath sounds over the left chest. The point of maximal cardiac impulse is shifted to the right. What is the most appropriate initial respiratory strategy?

A. Use noninvasive CPAP while monitoring for spontaneous improvement

B. Chest physiotherapy to mobilize presumed retained fetal lung fluid

C. Immediate endotracheal intubation with gentle ventilation and gastric decompression

D. Bag-mask ventilation with high peak pressures to expand the left lung

141. A stable newborn has supraventricular tachycardia at 270/min that persists despite vagal maneuvers. Intravenous access is available. Which medication is the preferred acute therapy?

A. Intravenous atropine bolus

B. Slow intravenous digoxin loading

C. Rapid intravenous adenosine

D. Continuous prostaglandin E1 infusion

142. A 30-week infant is delivered after the mother received a complete course of antenatal betamethasone 36 hours earlier. Which neonatal outcome is most consistently reduced by this treatment?

A. Respiratory distress syndrome due to surfactant deficiency

B. Late-onset sepsis caused by central venous catheters

C. Retinopathy of prematurity requiring laser therapy

D. Patent ductus arteriosus requiring pharmacologic closure

143. A 2-hour-old preterm infant has respiratory distress, hypotension, and temperature instability after maternal fever and prolonged rupture of membranes. Which organisms are among the most important causes of early-onset neonatal sepsis?

A. Vibrio cholerae and Shigella dysenteriae

B. Bordetella pertussis and Mycoplasma pneumoniae

C. Clostridioides difficile and Helicobacter pylori

D. Group B Streptococcus and Escherichia coli

144. A 900-g infant has alkaline phosphatase elevation and low serum phosphorus after prolonged parenteral nutrition and slow advancement of fortified feeds. Which complication is most likely?

A. Congenital hyperparathyroidism from maternal calcium excess

B. Metabolic bone disease of prematurity

C. Scurvy from isolated vitamin C deficiency

D. Osteopetrosis with excessive bone mineral density

145. A term newborn with respiratory distress has an oxygen saturation of 74% in the right hand and 88% in a foot. Which physiologic pattern best explains this reverse differential cyanosis?

A. Simple coarctation with left-to-right ductal shunting

B. Isolated persistent pulmonary hypertension with a normal great-artery relationship

C. Uncomplicated transient tachypnea after cesarean delivery

D. Transposition physiology with pulmonary hypertension and ductal streaming

146. A newborn with suspected inborn error of metabolism has hypoglycemia, cardiomyopathy, and low ketone production during fasting. Which pathway defect is most consistent with this pattern?

A. Classic galactosemia presenting with hepatic dysfunction

B. Urea cycle dysfunction with preserved fatty-acid use

C. Fatty-acid oxidation disorder

D. Phenylalanine hydroxylase deficiency

147. A newborn has unilateral absence of facial movement when crying, but the forehead also does not move and eye closure is incomplete on the affected side after a forceps delivery. Which diagnosis is most likely?

A. Peripheral facial nerve palsy

B. Normal facial asymmetry of the newborn

C. Isolated absence of the depressor anguli oris muscle

D. Bilateral cortical stroke

148. A newborn has recurrent bacterial infections, omphalitis with delayed cord separation, and marked neutrophilia. Which immune defect is most likely?

A. X-linked agammaglobulinemia

B. Selective IgA deficiency

C. Leukocyte adhesion deficiency

D. Hereditary angioedema

149. A former preterm infant passes a routine neurologic examination but has a history of severe bronchopulmonary dysplasia and prolonged hospitalization. What follow-up is most appropriate?

A. Limit developmental follow-up to infants who develop seizures or motor abnormalities

B. Continue multidisciplinary developmental surveillance despite a normal single examination

C. Use growth measurements alone as the developmental assessment

D. Discharge from developmental follow-up because one examination is normal

150. A term infant has severe hypoxemia with a preductal saturation of 94% and postductal saturation of 82%. Echocardiography shows structurally normal heart anatomy and elevated pulmonary artery pressure. Which diagnosis is most likely?

A. Total anomalous pulmonary venous return without obstruction

B. Persistent pulmonary hypertension of the newborn

C. Large ventricular septal defect with unrestricted pulmonary flow

D. Tetralogy of Fallot with severe right ventricular outflow obstruction

151. A newborn fails bilateral automated auditory brainstem response screening. What is the most appropriate next step?

A. Ignore the result because newborn hearing screening has no value

B. Repeat hearing screening during later routine childhood surveillance

C. Arrange timely diagnostic audiologic follow-up rather than assuming deafness

D. Diagnose permanent deafness based on the screening test alone

152. A newborn has hypotonia, upslanting palpebral fissures, a single transverse palmar crease, and an atrioventricular septal defect. Which chromosomal abnormality is most likely?

A. Monosomy X

B. Trisomy 21

C. 22q11.2 deletion

D. Trisomy 13

153. A randomized trial of a new neonatal respiratory therapy reports a relative risk of 0.80 for bronchopulmonary dysplasia compared with standard care. How should this result be interpreted?

A. The treated group had about a 20% relative reduction in risk

B. The relative risk estimate is sufficient to establish treatment benefit

C. The therapy prevented bronchopulmonary dysplasia in 80% of infants

D. The absolute risk decreased by exactly 80 percentage points

154. A newborn with Down syndrome has leukocytosis with circulating blasts, hepatomegaly, and thrombocytopenia. The blasts later disappear spontaneously. Which neonatal condition is most likely?

A. Transient abnormal myelopoiesis

B. Stress-related neonatal leukemoid reaction

C. Juvenile myelomonocytic leukemia

D. Acute lymphoblastic leukemia unrelated to trisomy 21

155. A 25-week infant loses 12% of birth weight during the first four days and develops hypernatremia. Urine output remains brisk. Which mechanism most contributes to this pattern?

A. Obligate sodium retention from mature distal tubules

B. Inappropriately low insensible water loss in an incubator

C. Excess renal concentrating ability causing water retention

D. High transepidermal water loss through immature skin

156. A term infant born by elective cesarean without labor develops tachypnea shortly after birth. Oxygen need is mild, and chest radiography shows prominent perihilar streaking with fluid in the interlobar fissures. What is the most likely diagnosis?

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

B. Congenital diaphragmatic hernia with bowel in the chest

C. Respiratory distress syndrome from severe surfactant deficiency

D. Pulmonary interstitial emphysema from ventilator injury

157. A 26-week infant is clinically stable on the first day of life. The mother has begun expressing colostrum. Which enteral feeding approach is generally preferred when there is no contraindication?

A. Use sterile water feeds first to test gastrointestinal function

B. Delay enteral feeding until respiratory support has substantially decreased

C. Begin small volumes of human milk and advance as tolerance permits

D. Start full-volume formula feeds immediately without gradual advancement

158. A monochorionic diamniotic twin pregnancy is complicated by polyhydramnios in one sac and oligohydramnios in the other. The recipient twin is hydropic, while the donor twin is growth restricted. Which placental finding underlies this disorder?

A. Unbalanced vascular anastomoses between the two fetal circulations

B. Complete separation of the placental vascular territories

C. Failure of fusion between the chorion and amnion

D. Placental infarction limited to the recipient twin territory

159. A newborn is exposed to maternal varicella when the mother develops a rash from five days before to two days after delivery. Why is this timing particularly concerning?

A. The main neonatal concern is bacterial infection from maternal skin lesions

B. The infant may have high-risk neonatal varicella with little transferred maternal antibody

C. Transplacental maternal antibody is expected to provide substantial protection

D. Varicella cannot be transmitted around the time of delivery

160. A newborn with trisomy 21 has tachypnea and a systolic murmur. Echocardiography shows a common atrioventricular valve and a large primum atrial and inlet ventricular septal defect. What is the diagnosis?

A. Total anomalous pulmonary venous connection

B. Double-outlet right ventricle with subaortic stenosis

C. Truncus arteriosus with a single semilunar valve

D. Complete atrioventricular septal defect

161. A 39-week infant receives face-mask positive-pressure ventilation after birth. The heart rate remains 70/min and there is no visible chest movement. What is the best next step?

A. Observe for one minute because the heart rate is above 60/min

B. Perform ventilation corrective maneuvers to achieve chest movement

C. Give sodium bicarbonate to correct presumed metabolic acidosis

D. Begin chest compressions immediately without further ventilation

162. A 35-week infant develops respiratory distress after maternal chorioamnionitis. Radiography shows diffuse bilateral opacities that could represent pneumonia or respiratory distress syndrome. Which approach is most appropriate initially?

A. Provide respiratory support while evaluating and empirically treating possible infection

B. Treat as surfactant deficiency while observing for infectious progression

C. Treat as transient tachypnea based on late-preterm gestation and radiographic pattern

D. Delay antibiotics until a blood culture becomes positive

163. A newborn has bilious emesis soon after birth. Abdominal radiography shows a double-bubble pattern with no distal bowel gas. Which diagnosis is most likely?

A. Jejunal atresia

B. Duodenal atresia

C. Meconium ileus

D. Pyloric stenosis

164. A term infant develops focal clonic seizures at 12 hours of life. The pregnancy and delivery were uncomplicated. MRI shows a left middle cerebral artery infarction. Which diagnosis is most likely?

A. Hypoxic-ischemic encephalopathy from global asphyxia

B. Benign neonatal sleep myoclonus

C. Periventricular leukomalacia of prematurity

D. Neonatal arterial ischemic stroke

165. A newborn has craniosynostosis, broad radially deviated thumbs, and broad great toes. Which syndrome is most likely?

A. Cornelia de Lange syndrome

B. Noonan syndrome

C. Marfan syndrome

D. Pfeiffer syndrome

166. A very-low-birth-weight infant receiving aggressive parenteral nutrition develops hyperglycemia. Which mechanism is most characteristic in extremely preterm infants?

A. Complete absence of hepatic glucose production

B. Obligate renal glucose retention despite high plasma glucose

C. Limited insulin secretion and peripheral insulin resistance

D. Excess pancreatic beta-cell mass with persistent hyperinsulinemia

167. A large-for-gestational-age infant of a diabetic mother is jittery at 45 minutes of life. Bedside glucose is very low. What is the primary mechanism?

A. Excess neonatal glucagon release causing rapid glucose uptake

B. Persistent fetal hyperinsulinemia after maternal glucose supply stops

C. Failure of maternal insulin to cross the placenta after delivery

D. Congenital absence of pancreatic beta cells

168. A neonatal treatment lowers the relative risk of an outcome substantially, but the baseline event rate is only 1%. Why is the absolute effect important for counseling?

A. Relative risk can be converted directly into the number needed to treat

B. The absolute risk reduction determines how many infants are likely to benefit

C. A low baseline risk guarantees no adverse effects from treatment

D. Absolute risk is unrelated to clinical decision-making

169. A newborn has an abdominal mass and ultrasound shows severe bilateral hydronephrosis with a thick-walled bladder. The infant is male and has a weak urinary stream. What is the most likely cause?

A. Posterior urethral valves causing bladder outlet obstruction

B. Transient antenatal urinary tract dilation that resolves after birth

C. Unilateral ureteropelvic junction obstruction

D. Bilateral renal agenesis with a distended bladder

170. A newborn has anemia, jaundice, splenomegaly, spherocytes on smear, and a negative direct antiglobulin test. A parent had a splenectomy for anemia. Which diagnosis is most likely?

A. Iron deficiency anemia

B. Hereditary spherocytosis

C. Glucose-6-phosphate dehydrogenase deficiency causing nonimmune hemolysis

D. ABO hemolytic disease

171. A 26-week infant has persistent oxygen need and frequent desaturations. Echocardiography shows elevated right ventricular pressure and septal flattening at 38 weeks postmenstrual age. Which complication should be suspected?

A. Spontaneous closure of the ductus with systemic hypertension

B. Isolated left ventricular hypertrophy from maternal diabetes

C. Pulmonary hypertension associated with bronchopulmonary dysplasia

D. Acute viral myocarditis without pulmonary vascular disease

172. A premature infant with respiratory distress syndrome has a continuous murmur, bounding pulses, widened pulse pressure, and increasing ventilator requirements on day 4. Which hemodynamic consequence best explains the respiratory deterioration?

A. Fixed obstruction of left ventricular inflow across the mitral valve

B. Pulmonary overcirculation from a left-to-right ductal shunt

C. Decreased pulmonary blood flow from premature ductal closure

D. Systemic venous congestion from isolated right-to-left ductal flow

173. A term infant is born after maternal use of an angiotensin-converting enzyme inhibitor throughout pregnancy. The infant has hypotension, oliguria, and renal insufficiency. Which fetal effect of this exposure is most characteristic?

A. Immune-mediated glomerulonephritis from maternal antibodies

B. Bilateral renal vein thrombosis from fetal polycythemia

C. Renal tubular dysgenesis with impaired fetal renal perfusion

D. Autosomal recessive polycystic kidney disease phenotype

174. A 3-week-old infant presents with fever, irritability, poor feeding, and a bulging fontanelle. Blood culture grows group B Streptococcus. Which complication should be actively evaluated?

A. Pyloric stenosis caused by bacterial toxin

B. Tetanus from placental transmission

C. Congenital rubella syndrome from the same organism

D. Meningitis as a manifestation of late-onset GBS disease

175. A preterm infant receiving morphine becomes increasingly somnolent with shallow respirations after renal function worsens. Which pharmacologic principle best explains the change?

A. Impaired clearance can increase exposure to drug or active metabolites

B. Rapid pharmacologic tolerance after repeated neonatal opioid exposure

C. Protein binding becomes so complete that free drug disappears

D. Increased opioid elimination caused by reduced renal tubular reabsorption

176. A newborn under intensive phototherapy has a falling total bilirubin. Which mechanism accounts for phototherapy's effect?

A. Conversion of bilirubin into more water-soluble photoisomers

B. Physical removal of bilirubin from blood through the skin surface

C. Inhibition of heme oxygenase through systemic ultraviolet absorption

D. Increased hepatic conjugating enzyme synthesis within minutes

177. A medically stable very preterm infant is ready for discharge but the parents are anxious about feeding, medications, and emergency signs. Which discharge practice is most effective?

A. Concentrate caregiver teaching in a structured session on the day of discharge

B. Use teach-back, written plans, medication reconciliation, and coordinated follow-up

C. Rely on the primary care clinician to reconstruct the NICU course independently

D. Provide verbal instructions once without confirming understanding

178. A healthy term infant develops numerous small pustules on a nonerythematous base at birth. The pustules rupture, leaving hyperpigmented macules with a collarette of scale. What is the diagnosis?

A. Transient neonatal pustular melanosis

B. Neonatal herpes simplex infection

C. Erythema toxicum neonatorum

D. Bullous impetigo

179. A newborn has a lumbosacral myelomeningocele with exposed neural tissue. Which immediate care measure is most appropriate?

A. Place the infant supine directly on the lesion

B. Cover the lesion with a sterile moist nonadherent dressing and avoid pressure

C. Clean the neural tissue repeatedly with alcohol

D. Begin oral feeding before evaluating associated hydrocephalus

180. A female newborn has diffuse edema of the hands and feet, a webbed neck, widely spaced nipples, and a left-sided obstructive cardiac lesion. Which diagnosis is most likely?

A. Fragile X syndrome

B. Prader-Willi syndrome

C. Trisomy 13

D. Turner syndrome

181. A preterm infant on conventional ventilation has severe respiratory distress syndrome and requires high peak pressures to achieve adequate tidal volumes. Which ventilator strategy is most consistent with lung-protective care?

A. Normalize PaCO2 aggressively even if very large tidal volumes are required

B. Maintain sustained hyperoxia to prevent intermittent desaturation

C. Use minimal positive end-expiratory pressure to reduce barotrauma risk

D. Use the lowest pressures and tidal volumes that provide adequate gas exchange

182. A mother asks why her milk is preferred for her 27-week infant even after pasteurization is available for donor milk. Which benefit is most directly associated with mother's own milk?

A. It contains no variability in protein or caloric content

B. It markedly lowers the risk of late-onset sepsis in very preterm infants

C. It provides species-specific nutrients and bioactive immune components

D. Its nutrient content is sufficient for extremely preterm growth without fortification

183. A former extremely preterm infant has persistent abnormal fidgety movements and poor postural control at 4 months corrected age. What is the most appropriate next step?

A. Defer formal cerebral palsy assessment until motor patterns stabilize in later childhood

B. Discontinue follow-up if weight gain is normal

C. Wait until school age because early motor findings are not useful

D. Refer promptly for standardized developmental assessment and early intervention

184. A newborn with 22q11.2 deletion has a conotruncal cardiac defect and profound T-cell lymphopenia. Which embryologic defect explains the immune abnormality?

A. Absence of splenic macrophages from mesodermal failure

B. Thymic hypoplasia from abnormal pharyngeal pouch development

C. Failure of fetal bone marrow to produce neutrophils

D. Excess maternal IgG transfer causing lymphocyte destruction

185. A term infant has cyanosis that worsens with crying. Echocardiography shows a common arterial trunk arising from the heart above a large ventricular septal defect. Which congenital lesion is present?

A. D-transposition of the great arteries

B. Persistent truncus arteriosus

C. Pulmonary atresia with intact ventricular septum

D. Critical valvar aortic stenosis

186. A newborn has a neck mass that transilluminates and is soft, multiloculated, and located in the posterior triangle. Which diagnosis is most likely?

A. Congenital muscular torticollis

B. Thyroglossal duct cyst

C. Lymphatic malformation

D. Second branchial cleft anomaly

187. A term infant has severe perinatal depression and requires prolonged resuscitation. Ten minutes later the infant is breathing spontaneously but has abnormal tone and reduced consciousness. Which next step is most important if the infant meets established criteria?

A. Begin prophylactic phenobarbital before any seizures occur

B. Delay neurologic assessment until after 24 hours of life

C. Rapidly assess eligibility for therapeutic hypothermia

D. Normalize body temperature above 38 degrees Celsius

188. A 1-day-old infant has bilious vomiting and a soft, minimally distended abdomen. Prenatal imaging was normal. Which diagnosis must be excluded urgently because bowel ischemia can develop rapidly?

A. Delayed physiologic meconium passage in a term infant

B. Uncomplicated gastroesophageal reflux

C. Hypertrophic pyloric stenosis

D. Malrotation with midgut volvulus

189. A preterm infant with evolving bronchopulmonary dysplasia has repeated episodes of pulmonary edema and poor lung compliance. Which intervention may improve short-term pulmonary mechanics but requires careful monitoring of electrolytes and bone health?

A. Diuretic therapy in carefully selected symptomatic infants

B. Reduce caloric fortification to decrease carbon dioxide production

C. Use prolonged anti-inflammatory macrolide therapy for evolving lung disease

D. High-volume crystalloid administration to expand pulmonary blood flow

190. A 2-day-old infant has vomiting, weight loss, hyperkalemia, hyponatremia, and shock. The external genitalia are virilized in a genetic female. Which disorder is most likely?

A. Pseudohypoaldosteronism with excess cortisol production

B. Congenital hyperthyroidism causing sodium retention

C. 11-beta-hydroxylase deficiency causing mineralocorticoid excess

D. 21-hydroxylase deficiency causing salt-wasting congenital adrenal hyperplasia

191. Two observers independently grade retinopathy of prematurity images. Which statistic is commonly used to quantify agreement beyond chance for categorical ratings?

A. Pearson correlation coefficient for association between ratings

B. Cohen kappa statistic for interrater agreement

C. Kaplan-Meier survival estimate

D. Number needed to treat

192. A male newborn has persistent bleeding after circumcision. Platelet count and prothrombin time are normal, but aPTT is prolonged. Which inherited disorder is most likely?

A. Disseminated intravascular coagulation

B. Immune thrombocytopenia with low platelets

C. Vitamin K deficiency bleeding

D. Hemophilia A from factor VIII deficiency

193. A preterm infant receiving indomethacin for PDA closure develops decreased urine output and a transient rise in serum creatinine. Which mechanism is most likely?

A. Direct immune destruction of the glomerular basement membrane

B. Reduced renal prostaglandin-mediated blood flow

C. Obligate osmotic diuresis from excessive bicarbonate generation

D. Increased renal vasodilation from cyclooxygenase inhibition

194. A 35-week infant is delivered by urgent cesarean because of a placental abruption. Cord blood shows severe metabolic acidosis, and the infant is pale with poor perfusion. Which additional neonatal problem should be specifically considered because of the abruption?

A. Acute fetal blood loss with hypovolemic shock

B. Isolated thrombocytosis from fetal stress

C. Chronic iron overload from placental transfusion

D. Hypervolemia caused by maternal-to-fetal hemorrhage

195. A 26-week infant with a central venous catheter develops new apnea, thrombocytopenia, and glucose instability at 3 weeks of age. Which diagnosis should be considered first?

A. Normal maturation of autonomic control

B. Recurrent apnea of prematurity triggered by recent caffeine discontinuation

C. Congenital pneumonia acquired before delivery

D. Late-onset sepsis related to nosocomial bloodstream infection

196. A newborn has profound hypotonia, absent deep tendon reflexes, weak cry, and tongue fasciculations but remains alert. Which disorder should be considered?

A. Congenital hyperthyroidism

B. Benign neonatal sleep myoclonus

C. Spinal muscular atrophy

D. Hypoxic-ischemic encephalopathy with hyperreflexia

197. A large-for-gestational-age newborn has macroglossia, omphalocele, ear creases, and recurrent hypoglycemia due to hyperinsulinism. Which syndrome is most likely?

A. Cri-du-chat syndrome

B. Smith-Lemli-Opitz syndrome

C. Turner syndrome

D. Beckwith-Wiedemann syndrome

198. A 29-week infant on parenteral nutrition develops progressive direct hyperbilirubinemia after several weeks with minimal enteral feeding. Which strategy most directly reduces the risk of parenteral-nutrition-associated cholestasis?

A. Increase intravenous glucose delivery to stimulate insulin secretion

B. Reduce intravenous lipid substantially while maintaining essential fatty acid intake

C. Advance enteral feeding as feasible and minimize unnecessary parenteral nutrition

D. Give prolonged broad-spectrum antibiotics to sterilize the intestine

199. A term newborn becomes profoundly cyanotic several hours after birth. Oxygen saturation is 68% despite supplemental oxygen. The second heart sound is single, and chest radiography shows a narrow mediastinum. Which intervention should be started while arranging definitive cardiac evaluation?

A. Prostaglandin E1 infusion to maintain ductal patency

B. Indomethacin to promote ductal closure

C. High-dose furosemide to reduce pulmonary edema

D. Inhaled nitric oxide as definitive treatment

200. A newborn has recurrent cyanotic spells that improve when crying. A catheter cannot be passed through either nostril. Which respiratory diagnosis is most likely?

A. Pulmonary interstitial emphysema

B. Bilateral choanal atresia

C. Unilateral diaphragmatic paralysis

D. Congenital lobar overinflation

Answers

1. C. Meconium aspiration can obstruct small airways, producing ball-valve air trapping, atelectasis, and air leak, while also causing chemical pneumonitis and surfactant dysfunction. The radiograph often has patchy opacities with hyperinflation. Persistent pulmonary hypertension may complicate severe cases.

2. B. A very rapid, regular, narrow-complex tachycardia with abrupt onset and termination strongly suggests supraventricular tachycardia. Sinus tachycardia typically varies with physiologic stimuli and rarely reaches such high fixed rates in a neonate. Complete heart block causes bradycardia rather than tachycardia.

3. C. Preterm infants have lower iron stores at birth and substantial iron needs for rapid growth and erythropoiesis, especially after phlebotomy losses. Enteral iron supplementation is commonly required once feeds are established, with dosing individualized to gestation, transfusion history, and diet.

4. D. Cystic periventricular leukomalacia represents destructive white-matter injury near the lateral ventricles and is strongly associated with motor impairment, particularly spastic diplegia. The descending corticospinal fibers to the lower extremities are especially vulnerable in this distribution.

5. D. A 22q11.2 deletion can cause conotruncal cardiac defects, thymic hypoplasia with T-cell dysfunction, hypocalcemia from parathyroid hypoplasia, and palatal abnormalities that impair feeding. The constellation is often recognized in the neonatal period.

6. A. Infants born after severe preeclampsia and placental insufficiency commonly have transient neutropenia, thought to reflect impaired neutrophil production. The finding is usually self-limited but can complicate early sepsis assessment. Chronic fetal hypoxemia more often contributes to polycythemia than to neutrophilia.

7. B. Neonatal enterovirus infection can be acquired perinatally and may cause fulminant sepsis-like disease with hepatitis, myocarditis, coagulopathy, and multiorgan failure. Maternal symptoms may be mild or nonspecific. Diagnosis relies on molecular testing from appropriate sites and supportive critical care.

8. C. Hyperinsulinism suppresses lipolysis and ketogenesis while driving glucose into tissues, so hypoglycemia occurs with inappropriately low ketones and free fatty acids. A high glucose requirement is another clue. Persistent cases require specialized endocrine and genetic evaluation.

9. D. An underpowered study has an increased risk of failing to detect a real treatment effect, which is a type II error. A nonsignificant result in a small study should therefore not automatically be interpreted as proof of equivalence or absence of benefit.

10. A. Hypernatremia with excessive dilute urine indicates inability to conserve water and should raise concern for central or nephrogenic diabetes insipidus. SIADH causes water retention, hyponatremia, and relatively concentrated urine. Evaluation includes serum and urine osmolality and assessment of the clinical context.

11. B. Vitamin K deficiency bleeding can occur in newborns who do not receive prophylaxis because vitamin K stores are low and intestinal bacterial synthesis is limited. Bleeding may involve the gastrointestinal tract, skin, or intracranial space. Intramuscular vitamin K at birth is highly effective prevention.

12. B. In neonatal resuscitation, effective lung ventilation is the priority when an infant is apneic, gasping, or has a heart rate below 100/min after initial steps. Chest compressions are reserved for a heart rate below 60/min despite adequate ventilation. Epinephrine and volume expansion come later when indicated.

13. A. Older strategies used aggressive hyperventilation and alkalosis to lower pulmonary vascular resistance, but profound hypocapnia and alkalosis can reduce cerebral blood flow and cause harm.

Contemporary management emphasizes adequate lung recruitment, oxygenation, ventilation, and selective pulmonary vasodilation while avoiding extreme gas targets.

14. C. Pneumoperitoneum in an infant with necrotizing enterocolitis indicates gastrointestinal perforation until proven otherwise and is a classic indication for urgent surgical involvement. The infant also requires resuscitation, antibiotics, and gastric decompression. This is not a benign sign of improving motility.

15. B. Maternal IgG is actively transported across the placenta, with transfer increasing substantially during the third trimester. Very preterm infants therefore begin life with lower IgG concentrations and reduced passive humoral protection. Maternal IgM does not normally cross the placenta in significant amounts.

16. A. Severe intraventricular hemorrhage, especially with parenchymal injury, and cystic periventricular leukomalacia are major predictors of later cerebral palsy and motor impairment. Neurodevelopmental outcome is multifactorial, so imaging must be integrated with clinical course and longitudinal assessment.

17. D. Retinopathy of prematurity is multifactorial, with extreme prematurity and low birth weight being dominant risks. Excessive oxygen exposure and large oxygen fluctuations can contribute to abnormal retinal vascular development, so oxygen is carefully titrated while avoiding prolonged hypoxemia.

18. A. Apnea is a well-recognized adverse effect of prostaglandin E1, particularly in neonates receiving higher doses or during transport. The infant should be closely monitored with readiness for respiratory support or intubation. The effect does not indicate treatment failure or ductal closure.

19. C. Coordinated suck-swallow-breathe patterns mature during late gestation, and preterm infants may have feeding-related desaturation or bradycardia before these skills are reliable. Feeding therapy, pacing, positioning, and maturation are often needed. The issue is not a universal inability to digest human milk.

20. D. Exogenous surfactant replaces deficient surface-active material and improves compliance and oxygenation in preterm respiratory distress syndrome. Modern strategies often combine early CPAP with selective surfactant when oxygen or work-of-breathing thresholds are reached. Oxygen alone does not correct the primary pathophysiology.

21. A. Maternal magnesium crosses the placenta and can transiently depress neonatal neuromuscular transmission, contributing to hypotonia and respiratory depression when levels are elevated. Supportive care is usually sufficient as magnesium is cleared, although severe toxicity may require targeted management.

22. C. Trisomy 13 is classically associated with midline craniofacial abnormalities, holoprosencephaly, microphthalmia, cleft lip/palate, and postaxial polydactyly. Trisomy 18 more often features clenched hands with overlapping fingers, rocker-bottom feet, and growth restriction.

23. B. Repeated intrauterine transfusions replace fetal red cells and improve oxygen delivery, which suppresses endogenous erythropoietin and fetal marrow activity. Consequently, infants may have anemia with an unexpectedly low reticulocyte count after birth. This differs from untreated hemolysis, in which reticulocytosis is usually prominent.

24. A. Congenital syphilis can present with rhinitis, hepatosplenomegaly, generalized lymphadenopathy, mucocutaneous rash including palms and soles, anemia, thrombocytopenia, and characteristic long-bone changes. Evaluation depends on maternal treatment history, infant examination, and serologic findings.

25. D. Gentamicin is eliminated primarily by the kidneys, and preterm infants have reduced glomerular filtration and renal clearance. Extended dosing intervals are therefore commonly used and adjusted with gestational/postnatal age, renal function, and drug levels. Neonatal pharmacokinetics change rapidly with maturation.

26. C. Transparent disclosure and a safety-focused review are appropriate after a medication error, even when no immediate harm is evident. High-quality systems examine contributing factors, strengthen safeguards, and support a culture of safety rather than concealment or reflexive blame.

27. C. Jaundice in the first 24 hours is considered abnormal and warrants prompt bilirubin measurement and evaluation for causes such as hemolysis. Management depends on total serum bilirubin, age in hours, gestational age, and neurotoxicity risk factors. Treatment should not be based on visual assessment alone.

28. D. Aplasia cutis congenita is congenital absence of skin, most commonly affecting the scalp. Lesions vary from small superficial defects to larger defects involving deeper structures. Evaluation should consider associated anomalies and syndromes, while management depends on lesion size and depth.

29. C. Tetralogy of Fallot consists of a ventricular septal defect, an overriding aorta, right ventricular outflow obstruction, and right ventricular hypertrophy. Severe pulmonary obstruction can make pulmonary blood flow ductal dependent. The other combinations do not define tetralogy.

30. C. Pulmonary interstitial emphysema occurs when air dissects from overdistended alveoli into the interstitial tissues, often in very preterm infants receiving mechanical ventilation. Radiographs show characteristic linear or cystic lucencies. Management focuses on reducing ventilator injury and optimizing lung volumes.

31. D. Neonatal hyperglycemia commonly reflects prematurity, excessive glucose delivery, stress, medications, or infection. Initial management includes confirming the glucose value, reviewing the glucose infusion rate, and addressing underlying illness. Insulin may be considered for persistent severe hyperglycemia but is not automatically first-line.

32. C. When a 95% confidence interval for a relative measure crosses 1, or for a difference crosses 0, the result is not statistically significant at the conventional two-sided 0.05 level. The interval also

describes the range of effect sizes compatible with the data and should be considered alongside clinical importance.

33. B. Anemia of prematurity reflects a blunted erythropoietin response to anemia, shortened red-cell lifespan, rapid growth, and frequent laboratory blood loss. Reticulocyte production is often inappropriately low. Management emphasizes minimizing phlebotomy, adequate nutrition, and transfusion when clinically indicated.

34. A. Distal renal tubular acidosis results from impaired distal acid secretion, producing a normal-anion-gap metabolic acidosis with inappropriately high urine pH. Nephrocalcinosis can occur because of hypercalciuria and hypocitraturia. Diabetes insipidus causes water loss rather than metabolic acidosis.

35. A. Extremely preterm infants have high protein requirements and rapidly become catabolic without amino acid provision. Early parenteral amino acids help support nitrogen balance and growth while enteral nutrition is advancing. They do not eliminate the need for enteral feeding or prevent every metabolic complication.

36. B. Myelomeningocele is frequently associated with Chiari II malformation and hydrocephalus. Increasing head circumference, a tense fontanelle, and sunset eye deviation suggest rising intracranial pressure. Serial head measurements and neuroimaging guide management.

37. A. Genetic evaluation begins with careful phenotyping, prenatal and family history, and selection of tests that address the clinical differential. Chromosomal analysis, microarray, targeted testing, or sequencing may be appropriate depending on the presentation. Physical findings alone should not be used to assume a karyotype.

38. B. For term and preterm infants who do not require immediate resuscitation, deferred cord clamping for at least approximately 60 seconds is beneficial in most circumstances. Immediate clamping is not routinely required. Intact cord milking is not a universal substitute for deferred clamping.

39. D. Respiratory distress syndrome in a very preterm infant results from surfactant deficiency, which increases surface tension and promotes widespread atelectasis. The lungs are poorly compliant, producing low volumes, diffuse granular opacities, and air bronchograms. Hyperinflation is more typical of obstructive processes.

40. B. Physiologic and breast-milk jaundice are predominantly unconjugated processes. Direct hyperbilirubinemia, dark urine, and pale or acholic stools indicate cholestasis and require prompt evaluation for hepatobiliary disease, including biliary atresia, infection, metabolic disease, and parenteral-nutrition-associated cholestasis.

41. A. In hypoplastic left heart syndrome, the right ventricle supports both pulmonary and systemic circulation. Systemic blood flow depends on right-to-left flow across the ductus arteriosus into the aorta, so prostaglandin E1 is essential before staged palliation. Adequate atrial-level mixing is also required.

42. A. Absent or reversed end-diastolic umbilical artery flow reflects markedly increased placental vascular resistance and is associated with fetal compromise, acidosis, and stillbirth risk. It is not a benign finding in a growth-restricted fetus. Cerebral Doppler changes are assessed in different vessels.

43. D. Endotracheal tubes become colonized, so a positive tracheal culture alone does not establish pneumonia or ventilator-associated infection. Diagnosis requires integration of respiratory deterioration, imaging, inflammatory or systemic findings, and culture data. Unnecessary antibiotics promote resistance and other adverse effects.

44. D. Preterm infants may have limited capacity to clear intravenous triglycerides, especially during illness or with excessive lipid delivery. Significant hypertriglyceridemia should prompt reassessment and reduction or temporary adjustment of the lipid dose while preserving essential fatty acid needs when possible.

45. B. Absent exhaled carbon dioxide together with gastric breath sounds and clinical deterioration strongly suggests esophageal intubation. The tube should be removed and effective ventilation re-established immediately. Right mainstem intubation would usually still generate exhaled carbon dioxide.

46. B. Very preterm infants lose heat rapidly because of a high surface-area-to-mass ratio, thin immature skin, low fat stores, and limited thermogenic capacity. Hypothermia increases oxygen consumption and can worsen hypoglycemia and metabolic acidosis. Thermal protection is therefore a major part of delivery-room and transport care.

47. C. A significant intrapartum hypoxic event followed by early encephalopathy and seizures is characteristic of hypoxic-ischemic encephalopathy. Neurologic examination, blood gases, organ injury, EEG monitoring, and imaging help establish severity and exclude mimics. Eligible infants should be assessed urgently for therapeutic hypothermia.

48. C. Corrected age accounts for the number of weeks the infant was born before term and is commonly used when assessing growth and development in former preterm infants during the first years of life. Chronologic age remains important for immunizations and many routine care decisions.

49. D. Fetal and neonatal alloimmune thrombocytopenia occurs when maternal antibodies target a fetal platelet antigen inherited from the father. The mother can have a normal platelet count because her own platelets lack the target antigen. Severe fetal thrombocytopenia and intracranial hemorrhage can recur in subsequent pregnancies.

50. C. A thyroglossal duct cyst is a midline neck mass arising from persistence of the tract followed by the developing thyroid. It characteristically moves with swallowing or tongue protrusion. Imaging should confirm the presence and location of normal thyroid tissue before excision.

51. B. Critical coarctation may present as the ductus arteriosus closes, causing abrupt reduction in lower-body perfusion and shock. Upper-extremity hypertension relative to the legs and weak femoral pulses are important clues. Small septal defects do not cause this pattern of systemic hypoperfusion.

52. B. Chromosomal microarray detects many clinically significant copy-number variants that are too small to be seen on a conventional karyotype. It is commonly used in infants with multiple congenital anomalies or unexplained dysmorphism when aneuploidy testing is unrevealing. It does not replace targeted testing when a specific single-gene disorder is strongly suspected.

53. D. Quality improvement commonly uses iterative plan-do-study-act cycles: plan a change, implement it, study the results, and adapt the process. The goal is rapid local learning and system improvement, although rigorous measurement is still needed to determine whether changes are meaningful and sustained.

54. B. Hyponatremia with edema and weight gain suggests excess total body water rather than simple sodium depletion. Management begins with assessment of fluid balance, renal function, and sources of free water, with fluid restriction or adjustment as appropriate. Hypertonic saline is reserved for selected severe or symptomatic cases.

55. C. Neonatal polycythemia increases blood viscosity and can impair microvascular perfusion, causing neurologic, respiratory, metabolic, and gastrointestinal symptoms. A venous hematocrit is used for confirmation because capillary values may overestimate the level. Management depends on symptoms and severity.

56. A. Inspiratory stridor and a weak cry after intubation suggest an upper-airway problem such as laryngeal edema or vocal-cord injury. Distal parenchymal disorders cause tachypnea, retractions, or hypoxemia rather than isolated stridor. Airway assessment is warranted if symptoms are significant or persistent.

57. B. Human milk alone does not reliably provide enough protein, calcium, phosphorus, and other nutrients for very preterm infants at standard volumes. Human-milk fortification improves nutrient density and supports growth and bone mineralization. Dilution would worsen the nutrient deficit.

58. A. Congenital infections acquired early in gestation can cause severe structural injury because organogenesis and neuronal proliferation are actively occurring. Later infections may transmit more efficiently for some pathogens but often cause less severe malformation. Maternal antibody transfer generally provides passive protection rather than amplifying infection.

59. A. The classic triad of congenital toxoplasmosis includes chorioretinitis, hydrocephalus, and intracranial calcifications that are often diffuse. Congenital CMV more often produces periventricular calcifications and microcephaly. Diagnosis uses maternal, neonatal, serologic, and molecular information.

60. D. When hypotension is associated with clinical evidence of hypovolemia and an underfilled heart, a cautious isotonic fluid bolus is reasonable. Neonatal hypotension should be treated according to the underlying physiology rather than by blood pressure alone. Diuresis would worsen true volume depletion.

61. A. Preterm lungs may require adjustment of inflation pressure to achieve effective ventilation. Current guidance supports initial peak inflation pressures in the range of about 20 to 30 cm H2O, followed by adjustment to clinical response while avoiding excessive tidal volume. Ventilation should not be abandoned when the infant remains apneic or bradycardic.

62. B. Bronchopulmonary dysplasia is a chronic lung disease of prematurity arising from arrested lung development plus inflammatory and ventilator/oxygen injury. Ongoing oxygen or respiratory support near 36 weeks postmenstrual age is central to contemporary definitions. The radiographic pattern may be heterogeneous with hyperinflation and atelectatic areas.

63. D. Failure to pass an orogastric tube with pooling of secretions strongly suggests esophageal atresia. The most common form includes a distal tracheoesophageal fistula, which may allow gas into the stomach. Feeding should be stopped, secretions managed, and surgical evaluation obtained.

64. B. The classic Erb pattern from upper brachial plexus injury involves C5-C6, producing shoulder adduction/internal rotation and an extended elbow with relatively preserved hand function. Most neonatal brachial plexus injuries improve spontaneously, but serial examination and early therapy are important.

65. B. Prader-Willi syndrome results from absence of paternally expressed genes in 15q11-q13 and often presents in infancy with profound hypotonia and feeding difficulty. Hyperphagia and obesity develop later. Angelman syndrome involves loss of maternal expression in the same region and has a different neurodevelopmental phenotype.

66. A. Small gastric residuals are common in preterm infants because gastric emptying and motility are immature. Residual volume or color alone is a poor discriminator of necrotizing enterocolitis; the entire clinical picture matters, including abdominal findings, stool, perfusion, and systemic illness.

67. B. Hypomagnesemia can impair parathyroid hormone secretion and action, causing or perpetuating neonatal hypocalcemia. It should be checked when hypocalcemia is symptomatic, severe, or refractory to calcium replacement. Infants of diabetic mothers are at increased risk of early hypocalcemia and hypomagnesemia.

68. A. Absolute risk reduction is the control event rate minus the treatment event rate: 20% minus 15% equals 5 percentage points. The corresponding number needed to treat would be 1/0.05, or 20, assuming the observed effect is valid and applicable.

69. B. Severe isolated thrombocytopenia in an otherwise well newborn with a mother who has a normal platelet count strongly suggests fetal and neonatal alloimmune thrombocytopenia. Maternal antibodies target a fetal platelet antigen inherited from the father. Cranial imaging is important because intracranial hemorrhage can occur before birth.

70. D. Unilateral multicystic dysplastic kidney is often managed conservatively when the contralateral kidney is normal. Follow-up focuses on renal growth, blood pressure, urinary tract abnormalities, and

function of the solitary kidney. Routine nephrectomy or dialysis is not required in an asymptomatic infant.

71. C. Apnea of prematurity reflects immature respiratory control and is commonly treated with caffeine, which reduces apnea and facilitates extubation. Before attributing events to prematurity, clinicians should consider secondary causes such as sepsis, anemia, temperature instability, or seizures. Respiratory depressants would worsen the problem.

72. B. Obstructed total anomalous pulmonary venous return causes severe pulmonary venous hypertension, pulmonary edema, and cyanosis. Because pulmonary venous blood cannot return normally to the left atrium and obstruction limits volume loading, the heart may not be enlarged. This is a surgical emergency.

73. D. Maternal anti-Ro/SSA and anti-La/SSB antibodies can cross the placenta and injure the fetal conduction system, causing congenital complete atrioventricular block. The ventricular rate is typically persistently slow and dissociated from atrial activity. Other maternal medications can cause bradycardia, but the antibody association is classic.

74. D. Neonatal herpes simplex virus infection can present with skin, eye, and mouth disease, encephalitis, or disseminated illness, and maternal history may be absent. When HSV is suspected, intravenous acyclovir should begin promptly while diagnostic testing is obtained because treatment delay can worsen outcomes.

75. C. When protein binding is reduced, the unbound or pharmacologically active fraction of a highly bound drug may increase. Neonates also have different body-water composition, organ clearance, and protein-binding capacity from older children. Total measured drug concentration may therefore not perfectly reflect active exposure.

76. B. Poor feeding, abnormal tone, and a high-pitched cry in the setting of severe unconjugated hyperbilirubinemia are concerning for acute bilirubin encephalopathy. Hemolysis can cause a rapid bilirubin rise and lowers the threshold for urgent escalation of care. Severe cases may require exchange transfusion.

77. C. Periviability counseling should be individualized, evidence informed, and family centered. Clinicians should communicate the range of possible outcomes and uncertainty, explore family values and goals, and explain reasonable resuscitative and palliative options when medically appropriate. Prognosis should not be reduced to a single variable.

78. A. Erythema toxicum neonatorum is a benign, self-limited eruption that usually appears after birth in otherwise well infants. Lesions are erythematous macules, papules, or pustules and classically contain eosinophils. Palms and soles are usually spared.

79. B. Neonatal seizures may manifest as focal clonic movements and ocular phenomena, and epileptic movements generally are not abolished by passive restraint. Jitteriness is stimulus-sensitive, typically

lacks ocular deviation, and can often be suppressed by holding the limb. EEG confirmation is important because many neonatal seizures are electrographic only.

80. A. CHARGE syndrome is associated with coloboma, heart defects, choanal atresia, growth and developmental problems, genital anomalies, and ear abnormalities. Pathogenic variants in CHD7 are common. Airway, feeding, hearing, and cardiac issues often dominate neonatal care.

81. B. Nasal CPAP provides continuous distending pressure that helps maintain functional residual capacity and prevent alveolar collapse while the infant breathes spontaneously. It is commonly used after extubation and as primary support for many preterm infants. Blow-by or low-flow oxygen does not provide reliable distending pressure.

82. D. Preterm kidneys have limited sodium-conserving ability, and sodium losses can impair growth even when fluid balance appears acceptable. Selected infants benefit from individualized sodium supplementation guided by serum values, urine losses, growth, and clinical status. Routine severe restriction can worsen deficiency.

83. B. Chronic granulomatous disease results from defective NADPH oxidase function, impairing the phagocyte respiratory burst. Patients are particularly susceptible to catalase-positive organisms and may form granulomas. An oxidative-burst assay is a key diagnostic test.

84. D. Neurodevelopmental outcome is multidimensional. Former preterm infants may have feeding, sensory, executive, language, attention, or learning difficulties even without major motor impairment. Follow-up should evaluate the whole child and family rather than a single milestone domain.

85. C. With pulmonary atresia, pulmonary blood flow may depend on a patent ductus arteriosus. Prostaglandin E1 should be used to maintain ductal patency until definitive catheter-based or surgical management. Ductal closure can cause catastrophic hypoxemia.

86. D. ROP screening timing is based primarily on gestational age at birth and chronological age so that examinations occur when sight-threatening retinal disease can first be detected. Screening is performed even when the external eye examination appears normal. Follow-up frequency depends on retinal findings.

87. D. Electrocardiographic monitoring provides rapid, reliable heart-rate assessment during neonatal resuscitation and is especially useful when chest compressions are being considered or performed. Pulse oximetry is valuable for oxygen saturation but may acquire slowly in poor perfusion. Palpation and auscultation are less continuous and can be inaccurate.

88. B. A new increase in apnea after a period of stability should prompt evaluation for an acquired cause, including infection, anemia, metabolic disturbance, temperature instability, or neurologic disease. Apnea of prematurity is a diagnosis of exclusion when the clinical pattern changes. Caffeine may ultimately be restarted, but evaluation comes first.

89. B. Pneumatosis intestinalis in a preterm infant with systemic illness and bloody stools is highly characteristic of necrotizing enterocolitis. Management includes bowel rest, gastric decompression, antibiotics, supportive care, and surgical consultation when perforation or progressive disease is suspected.

90. A. Severe neonatal hyperammonemia with respiratory alkalosis is characteristic of a urea cycle defect. Organic acidemias more often cause high-anion-gap metabolic acidosis and secondary hyperammonemia. Very high ammonia levels are neurotoxic and require urgent metabolic therapy and sometimes dialysis.

91. B. A highly sensitive test has relatively few false negatives, so a negative result can help rule out disease when used in an appropriate clinical context. Modest specificity means false positives may be common. Predictive values also depend strongly on disease prevalence or pretest probability.

92. A. Congenital and severe systemic infections can cause thrombocytopenia through a combination of consumption, inflammation, marrow suppression, and hypersplenism. Vitamin K deficiency impairs clotting-factor activity but does not directly lower the platelet count. The clinical context guides evaluation.

93. A. Bilateral renal agenesis causes severe oligohydramnios because fetal urine is a major source of amniotic fluid in the second half of pregnancy. Prolonged oligohydramnios causes pulmonary hypoplasia, which is the dominant lethal problem at birth. Characteristic limb and facial deformation may also occur.

94. C. A vigorous newborn born through meconium-stained fluid should receive routine initial newborn care rather than routine endotracheal suctioning. Suctioning is reserved for suspected airway obstruction that interferes with ventilation. Unnecessary suctioning can delay effective ventilation and cause bradycardia.

95. D. Candida bloodstream infection in a neonate is clinically significant and can disseminate to the central nervous system, eyes, kidneys, heart, and other organs. Persistent candidemia should prompt evaluation for deep foci and attention to infected intravascular devices. Treatment duration depends on clearance and organ involvement.

96. A. Phenobarbital remains a commonly used first-line medication for acute neonatal seizures, although treatment is increasingly guided by etiology, EEG response, and evolving evidence. Neonatal seizures are usually acute symptomatic rather than classic childhood epilepsy syndromes. Continuous EEG or amplitude-integrated EEG can help assess response.

97. B. Osteogenesis imperfecta is a collagen disorder that can present with fractures, osteopenia, bone deformity, and blue sclerae. Severity varies widely by genotype. A family history supports a heritable collagen disorder rather than a primary chondrodysplasia.

98. A. Human milk is associated with a lower risk of necrotizing enterocolitis than bovine formula, particularly in very preterm infants. The protective effect is one reason maternal or donor human milk is emphasized in this population. Prolonged fasting is not a safe substitute for appropriate feeding.

99. B. Inhaled nitric oxide reaches ventilated lung regions and produces selective pulmonary vasodilation, making it useful in appropriately selected late-preterm and term infants with hypoxic respiratory failure and pulmonary hypertension. Systemic vasodilators can worsen systemic hypotension and ventilation-perfusion matching.

100. A. A tension pneumothorax can rapidly impair ventilation and venous return, causing life-threatening hypoxemia and shock. When clinical signs are compelling in an unstable infant, decompression should not be delayed for radiography. Increasing airway pressure may worsen the air leak.

101. A. Pulmonary hypoplasia in congenital diaphragmatic hernia is accompanied by abnormal pulmonary vascular development and elevated pulmonary vascular resistance. Persistent pulmonary hypertension and right-to-left shunting can therefore cause profound hypoxemia even after ventilation is optimized.

102. C. Cyclooxygenase inhibitors such as indomethacin can reduce renal perfusion and urine output. Acetaminophen is an alternative pharmacologic option for PDA closure and may be considered when renal or gastrointestinal contraindications limit NSAID use. Prostaglandin E1 maintains, rather than closes, ductal patency.

103. B. Gastroesophageal reflux is common in preterm infants and is often physiologic. Acid-suppressive drugs have not shown routine benefit for nonspecific symptoms and may increase infectious or gastrointestinal risks. Management should focus on feeding volume, pacing, positioning when safe and monitored, and evaluation for alternative causes of symptoms.

104. C. Very preterm infants are vulnerable to germinal matrix hemorrhage because of fragile vasculature and unstable cerebral blood flow. Blood can extend into the ventricular system. Cranial ultrasonography is the standard bedside screening tool, and severity is related to ventricular involvement and parenchymal injury.

105. C. Achondroplasia is the most common nonlethal skeletal dysplasia and causes rhizomelic limb shortening, macrocephaly, and characteristic craniofacial features. It results from activating variants in FGFR3. Severe respiratory compromise is more typical of lethal skeletal dysplasias with marked thoracic restriction.

106. D. Severe, prolonged oligohydramnios early in gestation can impair lung growth and cause pulmonary hypoplasia. Affected infants may have a small chest and profound respiratory failure that responds poorly to routine ventilation. Antenatal corticosteroids promote rather than delay surfactant maturation.

107. C. Intrapartum antibiotic prophylaxis for mothers at increased risk is the cornerstone of preventing early-onset group B streptococcal disease. Antibiotics given well before labor do not reliably eradicate colonization, and there is no routinely available neonatal GBS vaccine. Newborn management depends on gestation, clinical appearance, and maternal risk factors.

108. C. Most infants with congenital hypothyroidism have few obvious signs at birth because of maternal thyroid hormone transfer and the gradual evolution of symptoms. Delayed treatment can impair brain development. Prompt confirmatory serum testing and levothyroxine therapy are therefore essential.

109. D. Before-and-after studies are vulnerable to secular trends and concurrent interventions. If hand hygiene improved at the same time as the bundle, the observed infection decline cannot be confidently attributed to the bundle alone. Randomized or carefully controlled designs reduce this threat.

110. B. G6PD deficiency reduces red-cell protection from oxidative stress and can cause episodic hemolysis with neonatal hyperbilirubinemia. Heinz bodies and bite cells support oxidant injury. Recognition is important because severe hyperbilirubinemia can develop rapidly.

111. A. Hypoxic-ischemic injury can cause acute kidney injury, most commonly through renal hypoperfusion and acute tubular injury. Oliguria, rising creatinine, and granular casts support this mechanism. Management emphasizes perfusion, avoidance of nephrotoxins, and careful fluid and electrolyte control.

112. C. Epinephrine is indicated when the heart rate remains below 60/min despite effective ventilation and coordinated chest compressions. The preferred route is intravascular, commonly through an umbilical venous catheter. Atropine, adenosine, and calcium are not routine first-line therapies for neonatal resuscitation bradycardia.

113. B. Meconium ileus is strongly associated with cystic fibrosis and results from abnormally thick intestinal secretions causing distal small-bowel obstruction. Contrast enema may be both diagnostic and therapeutic in uncomplicated cases. The infant should undergo evaluation for cystic fibrosis.

114. C. Carbon dioxide elimination depends primarily on alveolar ventilation. Increasing effective tidal volume or respiratory rate generally increases minute ventilation and lowers $PaCO_2$, provided dead-space ventilation and lung mechanics are considered. Excessive changes should be avoided because volutrauma and hypocapnia can be harmful.

115. D. Severe combined immunodeficiency causes profound defects in cellular immunity, often with impaired humoral immunity as well. Early findings may include severe or unusual infections, persistent candidiasis, diarrhea, failure to thrive, and lymphopenia. Early diagnosis is crucial because definitive therapy is time sensitive.

116. C. Cranial imaging provides important prognostic information, but many developmental outcomes are influenced by gestational age, illness severity, environment, sensory function, and subtle brain injury

not captured by routine ultrasound. Continued developmental surveillance remains appropriate for high-risk infants.

117. A. Blood pressure is only one component of neonatal hemodynamic assessment. A preterm infant with apparently adequate perfusion, urine output, and metabolic status may not benefit from treatment directed solely at a low numeric pressure. Unnecessary volume and vasoactive therapy can cause harm.

118. D. Laryngomalacia is the most common cause of congenital stridor and results from dynamic inspiratory collapse of supraglottic tissues. Symptoms often worsen with feeding, agitation, or supine positioning. Most cases improve with growth, but severe feeding or airway compromise may require intervention.

119. D. Pasteurized donor human milk is preferred over formula for many very-low-birth-weight infants when maternal milk is unavailable because it lowers the risk of necrotizing enterocolitis. Because donor milk has variable and often lower nutrient content after processing, fortification is commonly needed for adequate growth.

120. A. Endotracheal tubes can migrate into the right mainstem bronchus, especially with head movement, producing asymmetric breath sounds and left-sided atelectasis. Tube position should be corrected and confirmed. The absence of shock makes tension pneumothorax less likely in this vignette.

121. D. Therapeutic hypothermia started within the accepted early window reduces death or major neurodevelopmental disability in appropriately selected term and near-term infants with moderate to severe hypoxic-ischemic encephalopathy. Supportive intensive care and seizure detection remain essential. Severe hypocapnia can worsen cerebral perfusion.

122. A. The described pattern is characteristic of Edwards syndrome, or trisomy 18. Severe growth restriction, prominent occiput, micrognathia, clenched hands with overlapping fingers, and rocker-bottom feet are classic findings. Significant congenital heart and renal anomalies are also common.

123. B. Maternal glucose crosses the placenta, whereas maternal insulin does not. Fetal hyperglycemia stimulates fetal pancreatic insulin secretion, and insulin acts as a growth-promoting hormone that contributes to macrosomia and septal hypertrophy. The same hyperinsulinemic state also increases the risk of postnatal hypoglycemia.

124. D. Congenital cytomegalovirus can cause growth restriction, petechiae, hepatosplenomegaly, microcephaly, periventricular calcifications, and sensorineural hearing loss. Toxoplasmosis more classically causes diffuse intracranial calcifications, hydrocephalus, and chorioretinitis.

125. A. Neonatal medication errors can have large consequences because doses and volumes are small. Safe prescribing uses a current verified weight, correct units, age-specific dosing references, concentration checks, and independent verification for high-alert medications. Adult-dose fractions are unreliable and unsafe.

126. B. When a transcutaneous bilirubin approaches the treatment threshold, a serum bilirubin should be obtained because treatment decisions are based on total serum bilirubin. The current AAP approach incorporates gestational age, postnatal age, and neurotoxicity risk factors rather than relying on visual assessment alone.

127. D. Conflict about treatment goals should prompt clear, compassionate communication and interdisciplinary review. The team should distinguish treatments that are burdensome or nonbeneficial from those that can support achievable goals, while incorporating parental values and considering ethics consultation when needed. Decisions should not be made through abandonment or coercion.

128. A. Extremely preterm skin has an immature barrier, causing high evaporative heat and water loss and increased vulnerability to injury. High environmental humidity and gentle handling support thermal and fluid stability. Harsh adhesives and repeated alcohol exposure can damage the skin further.

129. B. An echocardiographic PDA is common in very preterm infants, but treatment decisions should incorporate the infant's respiratory status, systemic perfusion, shunt physiology, and overall clinical course. Ductal size alone does not mandate pharmacologic or surgical closure. Expectant management is appropriate for many stable infants.

130. A. Excess oxygen exposure contributes to oxidative injury, including retinopathy of prematurity and chronic lung disease. Neonatal oxygen therapy should therefore be titrated to appropriate saturation targets, avoiding both prolonged hypoxemia and unnecessary hyperoxia. Routine saturation of 100% is not a safe universal goal.

131. B. Organic acidemias often present after protein feeding with high-anion-gap metabolic acidosis, ketosis, poor feeding, and encephalopathy. Urea cycle disorders classically cause marked hyperammonemia with respiratory alkalosis and little or no metabolic acidosis. Urgent metabolic evaluation and supportive care are required.

132. D. Severity of illness is associated with both the exposure, prolonged ventilation, and the outcome, neurodevelopmental impairment. It can therefore confound the observed relationship. Statistical adjustment or study design can reduce but not always eliminate residual confounding.

133. A. Preterm kidneys have immature tubular function and limited capacity to conserve sodium. Renal sodium losses can therefore lead to late hyponatremia, particularly during rapid growth. Sodium supplementation may be needed in selected infants after assessment of fluid status and intake.

134. C. Disseminated intravascular coagulation causes systemic coagulation activation with consumption of platelets and clotting factors, producing thrombocytopenia, prolonged coagulation studies, low fibrinogen, and elevated fibrin degradation products. Sepsis, severe asphyxia, and shock are common neonatal triggers.

135. D. Growth assessment in preterm infants should consider serial weight gain together with linear and head growth, interpreted on appropriate growth curves. A single weight or laboratory value cannot

characterize nutritional adequacy. Poor head or length growth may reveal deficits not obvious from weight alone.

136. B. Congenital myotonic dystrophy can cause profound neonatal hypotonia, facial and bulbar weakness, and respiratory failure. A key diagnostic clue is previously unrecognized maternal myotonia, because anticipation can produce severe disease in the infant. Genetic testing confirms the diagnosis.

137. C. Pierre Robin sequence consists of mandibular hypoplasia leading to posterior displacement of the tongue and often a cleft palate. The resulting glossoptosis can cause positional airway obstruction and feeding difficulty. It may occur in isolation or as part of another syndrome.

138. C. When the heart rate remains below 60/min after at least 30 seconds of effective ventilation, chest compressions are indicated. Neonatal compressions are coordinated with ventilation, traditionally at a 3:1 ratio. Profound neonatal bradycardia is usually caused by inadequate oxygenation rather than a shockable arrhythmia.

139. A. Hirschsprung disease results from absence of enteric ganglion cells in the distal bowel, causing functional obstruction. Delayed meconium passage, abdominal distention, and explosive stool after rectal examination are classic clues. Rectal suction biopsy is used for definitive diagnosis.

140. C. Congenital diaphragmatic hernia should be suspected with a scaphoid abdomen, unilateral decreased breath sounds, and mediastinal shift. Bag-mask ventilation can insufflate intrathoracic bowel and worsen compression, so prompt intubation, gentle ventilation, and gastric decompression are preferred. Definitive repair follows stabilization.

141. C. Adenosine is the preferred acute therapy for stable narrow-complex reentrant supraventricular tachycardia because its very short half-life transiently blocks atrioventricular nodal conduction. It should be given rapidly through a proximal IV with an immediate flush. Synchronized cardioversion is used when the infant is unstable.

142. A. Antenatal corticosteroids accelerate fetal lung maturation and reduce the incidence and severity of respiratory distress syndrome in preterm infants. They also reduce intraventricular hemorrhage and neonatal mortality. They do not directly prevent catheter-related sepsis, PDA, or retinopathy of prematurity.

143. D. Early-onset neonatal sepsis is commonly caused by organisms acquired around delivery, with group B Streptococcus and Escherichia coli among the major pathogens. Prematurity, maternal fever, and prolonged rupture of membranes increase risk. Empiric therapy should reflect local epidemiology and neonatal susceptibility.

144. B. Metabolic bone disease of prematurity reflects inadequate mineral accretion, especially calcium and phosphorus, in very preterm infants. Low phosphorus and elevated alkaline phosphatase are common clues. Adequate mineral and vitamin D intake and optimized enteral nutrition are important preventive strategies.

145. D. Reverse differential cyanosis, in which postductal saturation exceeds preductal saturation, is unusual and can occur in transposition of the great arteries with pulmonary hypertension and specific ductal streaming. Typical persistent pulmonary hypertension more often produces lower postductal than preductal saturation.

146. C. Fatty-acid oxidation defects impair the ability to generate energy and ketones during fasting, leading to hypoketotic hypoglycemia and, in some disorders, cardiomyopathy or liver dysfunction. The absence of appropriate ketosis during hypoglycemia is a key diagnostic clue.

147. A. Peripheral facial nerve injury can follow birth trauma and affects the entire ipsilateral face, including forehead movement and eye closure. Congenital absence or hypoplasia of the depressor anguli oris typically produces lower-lip asymmetry with preserved forehead and eye function.

148. C. Leukocyte adhesion deficiency impairs neutrophil migration from blood into tissues. Affected infants can have severe bacterial infections, poor pus formation, delayed umbilical cord separation, and striking neutrophilia. Antibody deficiencies generally present later after maternal IgG wanes.

149. B. High-risk neonatal conditions can be associated with later cognitive, language, behavioral, sensory, and motor difficulties even when early examinations appear reassuring. Longitudinal multidisciplinary surveillance detects emerging problems and connects families with therapies and educational resources.

150. B. Persistent pulmonary hypertension causes elevated pulmonary vascular resistance with right-to-left shunting through fetal channels, often producing differential cyanosis with lower postductal saturation. Structurally normal cardiac anatomy and evidence of pulmonary hypertension support the diagnosis.

151. C. Newborn hearing screening identifies infants who need further evaluation but is not itself a definitive diagnosis of hearing loss. Timely diagnostic audiology is essential because early identification and intervention support language development. Risk factors such as congenital CMV or prolonged NICU care may warrant continued surveillance.

152. B. The combination of characteristic facial features, neonatal hypotonia, and an atrioventricular septal defect strongly suggests Down syndrome due to trisomy 21. Congenital heart disease is common, especially atrioventricular septal defects. The other chromosomal disorders have different typical phenotypes.

153. A. A relative risk of 0.80 means the event risk in the treated group is 80% of that in the control group, corresponding to a 20% relative risk reduction. Clinical interpretation also requires the absolute risks, confidence interval, study quality, and potential adverse effects.

154. A. Transient abnormal myelopoiesis occurs almost exclusively in neonates with trisomy 21 and is associated with GATA1-mutated megakaryoblastic proliferation. It may resolve spontaneously, but

severe organ involvement can be life threatening, and affected children have an increased later risk of myeloid leukemia of Down syndrome.

155. D. Extremely preterm infants have thin, immature skin and a large surface-area-to-mass ratio, producing substantial transepidermal water loss. This can contribute to excessive weight loss and hypernatremia if fluid replacement is insufficient. Humidified incubators and careful fluid adjustment reduce these losses.

156. A. Transient tachypnea of the newborn is caused by delayed clearance of fetal lung fluid and is more common after cesarean delivery without labor. Typical imaging shows hyperinflation, perihilar streaking, and fissural fluid. Symptoms usually improve over the first one to three days.

157. C. Early trophic or minimal enteral feeding with human milk supports intestinal adaptation and is generally preferred in stable very preterm infants when no contraindication is present. Human milk is associated with lower necrotizing enterocolitis risk than formula. Feeding advancement is individualized to clinical status and tolerance.

158. A. Twin-twin transfusion syndrome results from unbalanced blood flow through placental vascular anastomoses in monochorionic pregnancies. The donor becomes relatively hypovolemic with oligohydramnios and growth restriction, whereas the recipient develops volume overload, polyhydramnios, and possible hydrops or cardiomyopathy.

159. B. Maternal varicella close to delivery can expose the infant to virus before sufficient protective maternal antibody has been transferred. This creates risk for severe neonatal varicella. Management depends on timing and exposure details and may include varicella-zoster immune globulin and antiviral therapy.

160. D. A complete atrioventricular septal defect includes a common atrioventricular junction and valve with atrial and ventricular septal components. It is strongly associated with trisomy 21. Pulmonary overcirculation and heart failure often become evident as pulmonary vascular resistance falls.

161. B. Failure of the heart rate to rise during positive-pressure ventilation usually indicates ineffective ventilation. The immediate priority is to correct mask leak, airway position, obstruction, or inadequate pressure and obtain chest movement. Compressions should not begin until effective ventilation has been established and the heart rate remains below 60/min.

162. A. Early-onset pneumonia can mimic other neonatal respiratory disorders, especially in preterm or late-preterm infants. Maternal chorioamnionitis increases infectious risk, so respiratory support and timely evaluation with empiric antibiotics are appropriate when clinical illness is present. A negative initial radiograph cannot exclude infection.

163. B. Duodenal atresia classically produces a double-bubble appearance representing the stomach and proximal duodenum, with little or no distal gas when obstruction is complete. It is associated with trisomy 21 and polyhydramnios. Bilious emesis occurs when the obstruction is distal to the ampulla.

164. D. Neonatal arterial ischemic stroke often presents with focal seizures in an otherwise relatively well term infant. MRI with diffusion-weighted imaging establishes the diagnosis and distribution. The presentation differs from global hypoxic-ischemic encephalopathy, which usually causes more diffuse neurologic dysfunction.

165. D. Pfeiffer syndrome is a craniosynostosis syndrome characterized by broad, medially deviated thumbs and great toes. Craniofacial anatomy can compromise the airway and feeding. Other craniosynostosis syndromes have overlapping cranial findings but different limb patterns.

166. C. Extremely preterm infants have limited capacity to regulate glucose because insulin secretion may be inadequate and peripheral insulin sensitivity is reduced. High glucose infusion rates, stress, medications, and sepsis can worsen hyperglycemia. Management usually begins by reassessing glucose delivery and clinical stressors.

167. B. Maternal hyperglycemia produces fetal hyperglycemia and compensatory fetal hyperinsulinemia. After cord clamping, the maternal glucose supply abruptly stops while insulin levels remain elevated, predisposing to early hypoglycemia. Symptomatic severe hypoglycemia requires prompt treatment.

168. B. Relative effects can appear large even when the event is uncommon. Absolute risk reduction incorporates baseline risk and allows calculation of the number needed to treat, making the expected magnitude of benefit easier to weigh against burdens, harms, and costs.

169. A. Posterior urethral valves are an important cause of lower urinary tract obstruction in male infants. Severe cases produce a thick-walled distended bladder, bilateral hydroureteronephrosis, renal dysplasia, and oligohydramnios. Prompt bladder drainage and urologic evaluation are required.

170. B. Hereditary spherocytosis can present neonatally with anemia and unconjugated hyperbilirubinemia. Spherocytes may be present, and a negative direct antiglobulin test helps distinguish it from immune hemolysis. Family history of anemia, gallstones, or splenectomy is an important clue.

171. C. Infants with moderate or severe bronchopulmonary dysplasia are at risk for pulmonary vascular disease and pulmonary hypertension. Echocardiographic evidence includes elevated right-sided pressures and septal flattening. Recognition matters because hypoxemia and poor growth can worsen pulmonary vascular stress.

172. B. A hemodynamically significant patent ductus arteriosus in a preterm infant commonly produces left-to-right shunting, pulmonary overcirculation, and systemic diastolic runoff. This can worsen lung compliance and ventilator dependence while producing bounding pulses and a widened pulse pressure.

173. C. ACE inhibitor or angiotensin receptor blocker exposure later in pregnancy can disrupt the fetal renin-angiotensin system, leading to renal tubular dysgenesis, oligohydramnios, hypotension, and renal failure. The mechanism is impaired renal perfusion and development, not immune injury or thrombosis.

174. D. Late-onset GBS disease commonly presents with bacteremia and may involve meningitis. Neurologic signs such as irritability or a bulging fontanelle increase concern and warrant cerebrospinal fluid evaluation when clinically safe. Intrapartum prophylaxis reduces early-onset disease but does not reliably prevent late-onset GBS infection.

175. A. Drug clearance in neonates depends on maturation and organ function. Renal or hepatic dysfunction can reduce elimination of a medication or its metabolites, increasing exposure and toxicity even when the prescribed dose has not changed. Doses and intervals must therefore be reassessed as clinical status changes.

176. A. Blue-green light changes unconjugated bilirubin into configurational and structural photoisomers that can be excreted without normal hepatic conjugation. Phototherapy does not physically extract bilirubin from the skin and does not depend on rapid induction of glucuronyl transferase.

177. B. Safe NICU discharge is a process rather than a single event. Teach-back, hands-on caregiver practice, medication reconciliation, clear warning signs, equipment training when relevant, and coordinated specialty and primary-care follow-up reduce preventable errors and improve family readiness.

178. A. Transient neonatal pustular melanosis is a benign condition present at birth, with superficial pustules that rupture and leave hyperpigmented macules and scale. Unlike erythema toxicum, lesions are not typically surrounded by erythema. The infant is otherwise well and no treatment is required.

179. B. An open neural tube defect should be protected from trauma, drying, and contamination with a sterile moist nonadherent dressing while the infant is positioned to avoid pressure on the sac. Neurosurgical evaluation is urgent, and associated hydrocephalus, Chiari II malformation, bladder dysfunction, and latex sensitivity require attention.

180. D. Turner syndrome, most often due to monosomy X or mosaicism, can present at birth with lymphedema of the hands and feet, a webbed neck, and characteristic chest shape. Coarctation of the aorta and bicuspid aortic valve are important associated cardiac lesions.

181. D. Lung-protective neonatal ventilation aims to avoid volutrauma, barotrauma, atelectrauma, and oxygen toxicity while maintaining acceptable gas exchange. Modest permissive hypercapnia may be tolerated in selected infants rather than using injuriously large tidal volumes. Appropriate PEEP helps maintain functional residual capacity.

182. C. Mother's own milk provides human-specific nutrients, immunologic factors, oligosaccharides, enzymes, and other bioactive components and is associated with important neonatal benefits. It does not completely prevent infection, and very preterm infants often still require fortification to meet protein and mineral needs.

183. D. High-risk infant follow-up and standardized early motor assessment can identify infants at increased risk for cerebral palsy and other developmental disorders before traditional late diagnoses. Early referral allows therapy during a period of high neuroplasticity and supports family-centered care.

184. B. The thymus develops from the pharyngeal pouches, and abnormal development in 22q11.2 deletion can cause thymic hypoplasia or aplasia with T-cell deficiency. The severity varies. This is mechanistically distinct from neutropenia or disorders of splenic function.

185. B. Persistent truncus arteriosus is characterized by a single arterial trunk supplying systemic, pulmonary, and coronary circulations, usually with a large ventricular septal defect. Mixing causes cyanosis, while falling pulmonary vascular resistance can lead to excessive pulmonary blood flow and heart failure.

186. C. A lymphatic malformation, historically called cystic hygroma when macrocystic, often presents as a soft, compressible, transilluminating neck mass that can cross tissue planes. Large lesions may compromise the airway. Association with chromosomal abnormalities is important when detected prenatally.

187. C. Therapeutic hypothermia reduces death or major disability in eligible term or near-term infants with moderate to severe hypoxic-ischemic encephalopathy when initiated within the therapeutic window. Eligibility depends on gestation, evidence of significant intrapartum hypoxia, and neurologic examination. Routine prophylactic antiseizure medication is not indicated in the absence of seizures.

188. D. Bilious emesis in a neonate is an emergency until an obstructive cause is excluded. Malrotation with midgut volvulus can compromise mesenteric blood flow and lead rapidly to intestinal necrosis, sometimes before marked distention develops. Prompt surgical evaluation and appropriate imaging are required.

189. A. Diuretics can transiently improve pulmonary mechanics in selected infants with bronchopulmonary dysplasia and pulmonary edema, although long-term outcome benefits are uncertain. They can cause electrolyte disturbances, nephrocalcinosis, and adverse bone effects. Therapy should therefore be individualized rather than routine.

190. D. Classic 21-hydroxylase deficiency reduces cortisol and aldosterone synthesis while increasing androgen production. Affected genetic females may have virilized genitalia, and both sexes can develop life-threatening salt wasting with hyponatremia, hyperkalemia, and shock. This is a neonatal endocrine emergency.

191. B. Cohen kappa measures agreement between observers for categorical classifications while accounting for the agreement expected by chance. Correlation measures association and can be high even when two raters systematically disagree. The choice of reliability statistic depends on the data type and number of raters.

192. D. Hemophilia A due to factor VIII deficiency causes an isolated prolonged aPTT with normal platelet count and PT. Bleeding after circumcision may be the first presentation in a newborn. Vitamin K deficiency typically prolongs PT first and can affect multiple vitamin K-dependent factors.

193. B. Indomethacin inhibits prostaglandin synthesis and can reduce renal blood flow and glomerular filtration, causing oliguria and increased creatinine. Renal function often improves after the drug is stopped, but dosing requires monitoring. The effect is hemodynamic rather than immune mediated.

194. A. Placental abruption can cause acute fetal hemorrhage as well as hypoxic-ischemic compromise. Pallor, poor perfusion, and metabolic acidosis should prompt assessment for hypovolemia and anemia. Maternal-to-fetal transfusion would cause volume gain rather than the characteristic concern for fetal blood loss.

195. D. Late-onset sepsis in very preterm infants may present nonspecifically with increased apnea, feeding intolerance, temperature or glucose instability, thrombocytopenia, or hemodynamic changes. Central vascular catheters increase bloodstream infection risk. A new change from baseline deserves prompt evaluation.

196. C. Spinal muscular atrophy is a lower motor neuron disorder that can present with severe hypotonia, weakness, absent reflexes, and tongue fasciculations while cognition and alertness are relatively preserved. Early molecular diagnosis is crucial because disease-modifying therapies are most effective when started promptly.

197. D. Beckwith-Wiedemann syndrome is an overgrowth disorder associated with macroglossia, abdominal-wall defects, organomegaly, neonatal hyperinsulinemic hypoglycemia, and characteristic ear findings. Affected children also require tumor surveillance because of increased embryonal tumor risk.

198. C. Prolonged parenteral nutrition, lack of enteral stimulation, infection, and prematurity all contribute to cholestasis. Advancing enteral nutrition when feasible and reducing parenteral exposure are central preventive measures. Lipid strategies may also matter, but total elimination risks essential fatty acid deficiency.

199. A. The presentation suggests d-transposition of the great arteries with inadequate mixing. Prostaglandin E1 maintains ductal patency and can improve mixing while urgent echocardiography and possible balloon atrial septostomy are arranged. Closing the ductus would worsen systemic oxygenation.

200. B. Newborns are preferential nasal breathers, so bilateral choanal atresia can cause severe distress and cyanosis that paradoxically improves with crying, when the mouth opens. Failure to pass a catheter through both nares supports the diagnosis. Immediate airway stabilization is required.