COMPREHENSIVE MOCK EXAM 3

Supplemental to: Neonatology Board Review 2026-2027

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American Board of Pediatrics Neonatal-Perinatal Medicine Subspecialty Certifying Examination - 200-question supplemental simulation, A-D single-best-answer format, domain-weighted to the approved ABP content outline

Board-Review Questions

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

A. Gastroschisis with uncovered bowel protruding through a right paraumbilical defect

B. Omphalocele with a midline membrane-covered sac containing abdominal organs

C. Umbilical hernia with skin-covered bowel and spontaneous reducibility

D. Congenital diaphragmatic hernia with bowel displaced into the thoracic cavity

2. A newborn screen suggests sickle cell disease in an infant who is clinically well. Why are vaso-occlusive symptoms uncommon in the immediate neonatal period?

A. Neonatal erythrocytes do not contain any beta-globin chains until one year of age

B. Maternal normal hemoglobin permanently replaces the infant's red cells during gestation

C. High fetal hemoglobin levels inhibit polymerization of sickle hemoglobin

D. Sickle hemoglobin is not produced until iron stores become depleted after six months

3. A preterm infant on CPAP develops recurrent desaturation and bradycardia. The abdomen is distended, temperature is unstable, and the infant is less active than usual. What is the best next step regarding the apnea?

A. Evaluate for secondary causes such as sepsis or gastrointestinal disease before labeling it apnea of prematurity

B. Administer a sedative to reduce energy expenditure and prevent recurrent apnea

C. Assume all apnea before 37 weeks is physiologic and requires no diagnostic evaluation

D. Increase caffeine indefinitely without investigating the new systemic findings

4. A male newborn has hypertelorism, low-set ears, redundant neck skin, pectus deformity, and pulmonary valve stenosis. Karyotype is normal. Which syndrome is most likely?

A. Turner syndrome requiring complete or partial monosomy X

B. Beckwith-Wiedemann syndrome producing neonatal overgrowth and macroglossia

C. Noonan syndrome involving the RAS-MAPK signaling pathway

D. Trisomy 18 producing rocker-bottom feet and overlapping fingers

5. A 3-week-old infant has persistent conjugated hyperbilirubinemia, pale stools, dark urine, and hepatomegaly. Which diagnosis must be excluded urgently because early surgery improves outcome?

A. Breast milk jaundice causing isolated unconjugated hyperbilirubinemia in a thriving infant

B. Biliary atresia causing neonatal cholestasis with acholic stools and dark urine

C. Gilbert syndrome causing mild intermittent unconjugated hyperbilirubinemia without pale stools

D. Physiologic jaundice caused by unconjugated bilirubin after birth

6. A newborn has annular erythematous skin lesions, thrombocytopenia, and congenital complete heart block. The mother has anti-Ro/SSA antibodies. Which diagnosis is most likely?

A. Omenn syndrome causing erythroderma from severe combined immunodeficiency and heart block

B. Kawasaki disease causing neonatal rash, thrombocytopenia, and atrioventricular block

C. Wiskott-Aldrich syndrome causing eczema, thrombocytopenia, and complete heart block

D. Neonatal lupus from transplacental maternal autoantibodies

7. A male fetus has oligohydramnios, bilateral hydronephrosis, a thick-walled bladder, and a keyhole appearance of the posterior urethra. What is the most likely diagnosis?

A. Posterior urethral valves causing congenital bladder outlet obstruction

B. Congenital nephrotic syndrome causing fetal protein loss and polyhydramnios

C. Multicystic dysplastic kidney affecting one kidney without lower urinary tract obstruction

D. Bilateral renal agenesis causing absent bladder filling throughout gestation

8. A 27-week infant develops grunting, retractions, and increasing oxygen need immediately after birth. Chest radiograph shows diffuse reticulogranular opacities with air bronchograms and low lung volumes. What is the most likely diagnosis?

A. Meconium aspiration syndrome with patchy air trapping and coarse infiltrates

B. Respiratory distress syndrome from surfactant deficiency and alveolar collapse

C. Congenital lobar overinflation causing focal hyperlucency and mediastinal shift

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

9. A 2-day-old infant has a heart rate fluctuating between 170 and 205 beats/min during crying and feeding, with visible P waves preceding each QRS complex. Which interpretation is most likely?

A. Sinus tachycardia driven by physiologic stress rather than fixed reentrant SVT

B. Reentrant supraventricular tachycardia with a relatively fixed rate near 200 beats/min

C. Ectopic atrial tachycardia with an abnormal atrial focus and persistent tachycardia

D. Atrial flutter with rapid atrial activity and variable atrioventricular conduction

10. A neonate has recurrent inspiratory stridor that is worse when supine and with feeding but improves when prone. Growth and oxygenation are otherwise preserved. Flexible laryngoscopy shows collapse of supraglottic tissues during inspiration. What is the diagnosis?

A. Tracheoesophageal fistula causing aspiration without inspiratory supraglottic collapse

B. Laryngomalacia with dynamic supraglottic collapse

C. Bilateral choanal atresia causing fixed posterior nasal obstruction

D. Subglottic stenosis causing a fixed biphasic airway obstruction after intubation

11. A ventilated infant has adequate chest movement but persistent hypoxemia. Arterial PaO2 does not rise as expected despite increasing inspired oxygen, and echocardiography shows severe right-to-left shunting. Which physiologic problem best explains the poor oxygen response?

A. Isolated metabolic acidosis that prevents oxygen from entering pulmonary capillary blood

B. Pure hypoventilation with no shunt and a normal alveolar-arterial oxygen gradient

C. Extrapulmonary shunting that bypasses ventilated alveoli

D. Mild anemia alone, which causes a large fall in arterial oxygen partial pressure

12. Prenatal imaging shows a lumbosacral open neural tube defect with exposed neural tissue. After birth, what is the immediate priority before definitive neurosurgical repair?

A. Cover the lesion with dry sterile gauze and change it frequently before surgery

B. Place the infant supine to stabilize the spine while awaiting neurosurgical repair

C. Protect the lesion with sterile moist nonadherent coverage and avoid pressure or contamination

D. Apply topical antibiotic ointment to the exposed tissue and defer protective positioning

13. A nonvigorous term infant is born through meconium-stained fluid. The infant is apneic with a heart rate of 80 beats/min. Which approach is preferred?

A. Perform routine tracheal suctioning until no meconium is recovered before any ventilation

B. Initiate ventilation promptly rather than delaying for routine tracheal suctioning

C. Withhold ventilation until the infant has been observed for spontaneous respirations for 60 seconds

D. Provide chest compressions first because meconium makes face-mask ventilation ineffective

14. A medication error reaches a newborn and causes transient hypotension but no lasting harm. The team has corrected the problem. What is the most appropriate communication with the family?

A. Disclose the error and known effects transparently, explain corrective steps, and answer questions

B. Alter the medical record to remove the error so the family is not distressed

C. Tell the family only if they independently discover the event after discharge

D. Do not mention the error because no permanent injury occurred

15. A symptomatic newborn is being evaluated for early-onset bacterial sepsis. Which empiric antibiotic combination is traditionally used to cover common pathogens while cultures are pending?

A. Oral azithromycin alone because atypical organisms cause most early neonatal sepsis

B. Metronidazole alone because anaerobic bacteria are the dominant early-onset pathogens

C. Ampicillin plus an aminoglycoside such as gentamicin, adjusted to local resistance patterns

D. Vancomycin alone because gram-negative organisms are rarely important at birth

16. A 30-week infant with RDS receives surfactant and then rapidly improves in lung compliance. What ventilator adjustment is especially important to prevent subsequent injury?

A. Maintain the current ventilator pressures until the next scheduled blood gas is obtained

B. Increase positive end-expiratory pressure to offset a presumed transient loss of recruitment

C. Promptly reduce pressure or volume delivery as compliance improves

D. Increase inspired oxygen preemptively while leaving pressure and volume delivery unchanged

17. A newborn has truncus arteriosus, hypocalcemia, absent thymic shadow, and feeding difficulties. Which genetic test would most directly evaluate the suspected syndrome?

A. Testing for a paternal 15q11-q13 deletion

B. Testing for a 22q11.2 deletion

C. Testing for a beta-globin gene mutation

D. Testing for an FGFR3 gain-of-function variant

18. A newborn with trisomy 21 develops signs of heart failure. Echocardiography shows a common atrioventricular valve and a defect involving both the atrial and ventricular septa. What is the diagnosis?

A. Complete atrioventricular septal defect associated with endocardial cushion maldevelopment

B. Tetralogy of Fallot with overriding aorta and right ventricular outflow obstruction

C. Total anomalous pulmonary venous return with pulmonary veins draining to systemic veins

D. Isolated muscular ventricular septal defect without atrioventricular valve involvement

19. A female newborn has marked dorsal hand and foot edema, a webbed neck, widely spaced nipples, and coarctation of the aorta. Which diagnosis is most likely?

A. Noonan syndrome caused most often by a RAS-MAPK pathway variant

B. Beckwith-Wiedemann syndrome involving disordered imprinting at chromosome 11p15

C. Turner syndrome due to complete or partial monosomy X

D. Trisomy 21 with an extra chromosome 21

20. A ventilated preterm infant has pH 7.18, PCO2 72 mm Hg, and adequate oxygenation. Which ventilator change most directly increases carbon dioxide elimination if lung mechanics allow?

A. Lower minute ventilation so carbon dioxide has more time to diffuse out of the blood

B. Reduce respiratory rate and accept smaller tidal volumes to accelerate carbon dioxide clearance

C. Decrease alveolar ventilation while increasing inspired oxygen concentration alone

D. Increase effective minute ventilation by raising tidal ventilation or respiratory rate

21. An extremely preterm infant receives aggressive early nutrition after a period of significant intrauterine undernutrition. Serum phosphorus falls while potassium and magnesium also trend downward. Which process should be considered?

A. Primary hyperparathyroidism causing simultaneous retention of phosphorus, potassium, and magnesium

B. Refeeding-like intracellular shifts during rapid anabolic nutrition

C. Renal failure causing obligatory urinary wasting of all three electrolytes during oliguria

D. Physiologic postnatal adaptation in which low phosphorus proves nutrition should be stopped entirely

22. Parents at 22 weeks gestation ask about resuscitation options after counseling reveals substantial uncertainty in survival and long-term outcomes. Which approach best reflects family-centered neonatal ethics?

A. Withhold prognostic uncertainty so the family is not burdened by difficult information

B. Present resuscitation as mandatory because parental values are irrelevant at the limits of viability

C. Ask the family to choose without clinician guidance because recommendations are ethically inappropriate

D. Use shared decision-making that presents prognosis honestly and incorporates the family's values within medically reasonable options

23. A wet preterm infant is placed in a cool room after delivery and becomes hypothermic, hypoglycemic, and acidotic. Which physiologic response to cold stress contributes to this deterioration?

A. Shivering thermogenesis rapidly generates heat without increasing oxygen consumption

B. Peripheral vasodilation conserves core heat and reduces metabolic demand

C. Cold exposure suppresses catecholamine release and decreases brown-fat metabolism

D. Nonshivering thermogenesis increases oxygen and glucose consumption

24. A 2-week-old infant has microcephaly, petechiae, hepatosplenomegaly, and sensorineural hearing loss. Congenital cytomegalovirus is suspected. Which test best establishes congenital rather than postnatal infection?

A. CMV-specific IgM serology obtained during the second week of life

B. CMV PCR from urine first obtained at 6 weeks of age

C. CMV PCR from saliva or urine obtained within the first 21 days of life

D. Maternal CMV IgG avidity testing performed after the infant has been discharged

25. A post-term infant is born through thick meconium-stained fluid and develops severe respiratory distress. Chest radiograph shows patchy bilateral infiltrates with areas of hyperinflation. Which mechanism is most important?

A. Airway obstruction with ball-valve air trapping plus chemical pneumonitis from aspirated meconium

B. Diffuse lymphatic obstruction causing fetal pleural effusions without airway disease

C. Uniform alveolar collapse caused exclusively by primary surfactant absence in the mature lung

D. Isolated pulmonary venous congestion caused by a large left-to-right ductal shunt

26. A term newborn fails to pass meconium and develops abdominal distention. Contrast enema shows a small-caliber unused colon, and thick meconium obstructs the terminal ileum. Which underlying disease should be suspected?

A. Cystic fibrosis causing meconium ileus with inspissated terminal ileal contents

B. Necrotizing enterocolitis causing pneumatosis and portal venous gas in a preterm infant

C. Hirschsprung disease causing aganglionosis with a rectosigmoid transition zone

D. Pyloric stenosis causing gastric outlet obstruction without distal meconium impaction

27. A former preterm infant is evaluated at 18 months corrected age with a standardized developmental instrument assessing cognitive, language, and motor domains. Which type of tool is being used?

A. General Movements Assessment focused on spontaneous motor repertoire in early infancy

B. Hammersmith Infant Neurological Examination focused on neurologic signs and motor function

C. A standardized infant developmental assessment such as the Bayley scales

D. Ages and Stages Questionnaire used as a parent-completed developmental screening tool

28. A newborn has a cleft palate and micrognathia with recurrent airway obstruction from posterior displacement of the tongue. Which immediate supportive measure is often helpful while a definitive airway plan is developed?

A. Use positioning that reduces glossoptosis and monitor airway and feeding safety closely

B. Avoid feeding assessment because cleft palate does not influence aspiration or airway management

C. Begin blind deep oral suctioning continuously because suction corrects mandibular hypoplasia

D. Force the infant supine because posterior tongue displacement improves in that position

29. A preterm infant develops a pulmonary hemorrhage with sudden bloody fluid from the endotracheal tube, falling hematocrit, and severe respiratory deterioration. Which associated hemodynamic condition should be considered?

A. Isolated systemic hypertension without any change in pulmonary circulation

B. Complete closure of all fetal shunts with unusually low pulmonary capillary pressure

C. Critical pulmonary stenosis with severely reduced pulmonary blood flow

D. A large patent ductus arteriosus with excessive pulmonary blood flow

30. A term infant becomes tachypneic and poorly perfused at 6 hours. The mother was colonized with group B Streptococcus and had prolonged rupture of membranes without intrapartum antibiotics. Which pathogen is a leading concern?

A. Mycoplasma pneumoniae causing classic early-onset neonatal meningitis

B. Bordetella pertussis causing immediate congenital pneumonia at birth

C. Group B Streptococcus causing early-onset neonatal sepsis

D. Clostridioides difficile causing bloodstream infection from maternal colonization

31. A 3-week-old infant has tachypnea, diaphoresis with feeds, poor weight gain, hepatomegaly, and a harsh holosystolic murmur at the left lower sternal border. Which lesion is most likely?

A. Small secundum atrial septal defect producing severe heart failure in the first week

B. Mild peripheral pulmonary stenosis causing systemic venous congestion and hepatomegaly

C. Patent foramen ovale producing a large fixed left-to-right shunt with feeding intolerance

D. A large ventricular septal defect producing pulmonary overcirculation as resistance falls

32. A growing 29-week infant is receiving mostly breast milk near discharge. The care team is planning home nutrition. Which principle is most appropriate?

A. Individualize fortification or post-discharge supplementation based on growth and nutrient needs

B. Discontinue fortifier at 36 weeks postmenstrual age once the infant has reached 2 kilograms

C. Replace breast milk with post-discharge preterm formula to standardize caloric intake

D. Continue the full NICU fortification regimen unchanged through 12 months corrected age

33. A breastfed term infant receives substantial mucosal immune protection from human milk. Which component is particularly important at mucosal surfaces?

A. Maternal IgM that crosses intestinal epithelium into the infant's circulation in large amounts

B. Neutrophils that permanently engraft in the infant's bone marrow and replace host immunity

C. Secretory IgA that helps limit pathogen adherence and invasion

D. Complement C9 that is absorbed intact and provides lifelong systemic complement activity

34. A 28-week infant is ventilated with high pressures and later develops a persistent need for oxygen. Which respiratory-care principle most directly reduces ventilator-induced lung injury?

A. Use higher positive end-expiratory pressure while accepting larger delivered tidal volumes

B. Use pressure-limited ventilation without monitoring the tidal volume delivered as compliance changes

C. Target persistent hypocapnia to reduce spontaneous respiratory effort and ventilator dyssynchrony

D. Use lung-protective ventilation that limits excessive tidal volume and pressure exposure

35. Monochorionic twins are delivered after chronic unbalanced placental transfusion. One infant is pale with hemoglobin 8 g/dL and the co-twin is plethoric with hemoglobin 23 g/dL. Which diagnosis best describes the pair?

A. Twin anemia-polycythemia sequence from chronic small-vessel placental transfusion

B. Neonatal alloimmune hemolysis because maternal platelet antibodies cause discordant hemoglobin levels

C. Both twins have physiologic anemia because fetal hemoglobin falls immediately after delivery

D. Both twins have fetomaternal hemorrhage because placental vascular connections cannot transfer blood between twins

36. A 39-week infant has severe encephalopathy after placental abruption. Cord pH is 6.88, the infant required prolonged resuscitation, and neurologic examination at 3 hours shows lethargy, hypotonia, and seizures. Which therapy should be considered urgently?

A. Delay neurologic treatment until MRI confirms established hypoxic-ischemic injury

B. Induce mild hyperthermia to increase cerebral blood flow during the first 24 hours

C. Initiate protocol-based therapeutic hypothermia within the eligibility window

D. Start prophylactic high-dose corticosteroids to prevent post-asphyxial cerebral edema

37. A newborn screen shows absent T-cell receptor excision circles. The infant has persistent lymphopenia but is not yet clinically infected. Which diagnosis requires urgent evaluation?

A. Hereditary spherocytosis with neonatal hemolysis and normal lymphocyte development

B. Chronic granulomatous disease with isolated neutrophil oxidative burst failure

C. Severe combined immunodeficiency with impaired T-cell development

D. Transient tachypnea of the newborn with delayed lung-fluid clearance

38. A term infant has severe hypoxemia despite ventilation. Preductal oxygen saturation is 95% in the right hand and postductal saturation is 82% in a foot. Echocardiography shows no major structural heart defect. Which condition is most likely?

A. Coarctation of the aorta causing equal desaturation in all four extremities

B. Persistent pulmonary hypertension of the newborn with right-to-left ductal shunting

C. Large ventricular septal defect causing isolated left-to-right intracardiac shunting

D. Supraventricular tachycardia causing a fixed preductal-postductal saturation gradient

39. A 39-week infant requires respiratory support immediately after birth. There is no evidence of congenital heart disease or severe blood loss. What initial oxygen concentration is generally appropriate?

A. Begin respiratory support with room air and titrate oxygen using pulse oximetry

B. Begin with 60% oxygen because fetal hemoglobin requires a high inspired oxygen fraction

C. Avoid pulse oximetry and titrate oxygen according to visible cyanosis alone

D. Begin with 100% oxygen for all term infants and reduce only after five minutes

40. Prenatal imaging shows a midline abdominal wall defect at the umbilical ring with liver and bowel contained within a membrane-covered sac. Which diagnosis is most likely?

A. Umbilical hernia with intact skin and no herniated liver in a membrane sac

B. Gastroschisis with uncovered bowel usually protruding to the right of the umbilicus

C. Omphalocele with a midline membrane-covered abdominal wall sac at the umbilicus

D. Bladder exstrophy with exposed bladder mucosa below the umbilicus

41. A 25-week infant is hemodynamically stable on day 2 and has no evidence of intestinal obstruction or severe hypoperfusion. What is the principal purpose of small-volume trophic feeds?

A. Meet most daily calorie and protein requirements enterally while maintaining a very small feed volume

B. Provide enough luminal substrate to replace mineral supplementation during the first postnatal days

C. Minimize intestinal peristalsis while preserving mucosal contact with small amounts of milk

D. Provide enteral stimulation that supports gastrointestinal maturation while nutrition is advanced

42. A vigorous 36-week infant is born after an uncomplicated delivery and does not require resuscitation. Which umbilical cord strategy is most consistent with current neonatal resuscitation guidance?

A. Defer cord clamping for at least about 60 seconds while supporting normal transition

B. Clamp the cord at about 30 seconds in a vigorous late-preterm infant to reduce polycythemia risk

C. Use intact cord milking as the routine substitute for deferred clamping in this vigorous 36-week infant

D. Clamp immediately so thermal care and routine assessment can begin under the radiant warmer

43. A 25-week infant is receiving gentamicin. Why are dosing intervals often longer than in older children?

A. Preterm infants have markedly accelerated glomerular filtration that clears aminoglycosides too rapidly

B. Aminoglycosides are stored in fetal hemoglobin and released only once each day

C. Gentamicin is eliminated mainly by the liver, which is hyperfunctional in extreme prematurity

D. Immature renal clearance prolongs aminoglycoside elimination in very preterm infants

44. A term infant with meconium aspiration has severe PPHN and labile oxygenation despite optimized ventilation. Echocardiography confirms high pulmonary pressures and adequate left ventricular function. Which pulmonary vasodilator is commonly used?

A. Begin enteral sildenafil as the primary rapid rescue therapy for severe labile hypoxemia

B. Use intravenous milrinone as the sole pulmonary vasodilator despite preserved left ventricular function

C. Infuse prostaglandin E1 primarily to lower pulmonary vascular resistance in otherwise isolated PPHN

D. Inhaled nitric oxide to selectively reduce pulmonary vascular resistance

45. A newborn has micrognathia, glossoptosis, and a U-shaped cleft palate with recurrent upper-airway obstruction. Which description is most accurate?

A. Treacher Collins syndrome with mandibular hypoplasia and associated upper-airway obstruction

B. Isolated cleft palate with secondary posterior tongue displacement despite normal mandibular size

C. Pierre Robin sequence in which mandibular hypoplasia leads to posterior tongue displacement

D. CHARGE syndrome in which choanal atresia is the principal cause of the neonatal airway obstruction

46. A growth-restricted newborn has clenched hands with overlapping fingers, micrognathia, rocker-bottom feet, and a ventricular septal defect. Which diagnosis best fits?

A. Trisomy 21 with hypotonia and an atrioventricular septal defect

B. Turner syndrome with lymphedema and coarctation of the aorta

C. Trisomy 13 with holoprosencephaly and postaxial polydactyly

D. Trisomy 18 producing the characteristic overlapping fingers and rocker-bottom feet

47. A neonate has poor feeding, vomiting, lethargy, ketosis, hyperammonemia, and a high anion-gap metabolic acidosis. Which category of inherited disease is most likely?

A. Central diabetes insipidus causing hypernatremia and dilute urine

B. Urea-cycle disorder causing isolated respiratory alkalosis without metabolic acidosis

C. Organic acidemia causing accumulation of toxic organic acids

D. Congenital hypothyroidism causing bradycardia and prolonged jaundice without anion-gap acidosis

48. A premature infant has persistent oxygen requirement and echocardiographic evidence of pulmonary hypertension in the setting of established bronchopulmonary dysplasia. Which principle is most important in management?

A. Optimize lung disease and oxygenation while evaluating and treating pulmonary vascular disease in a multidisciplinary manner

B. Stop nutritional support because somatic growth increases pulmonary vascular resistance

C. Intentionally maintain chronic hypoxemia because oxygen promotes pulmonary vasoconstriction

D. Close all intracardiac shunts immediately without assessing their hemodynamic significance

49. A term newborn has profound hypotonia, weak suck, poor arousal, and cryptorchidism. Chromosomal testing later shows loss of paternally expressed genes in 15q11-q13. Which diagnosis is present?

A. Prader-Willi syndrome from loss of paternally expressed genes at chromosome 15q11-q13

B. Noonan syndrome from a pathogenic RAS-MAPK pathway variant

C. Spinal muscular atrophy from biallelic SMN1 deletion

D. Angelman syndrome from loss of maternally expressed genes in the same region

50. A 1-day-old infant has a serum creatinine of 1.0 mg/dL, the same as the mother's value before delivery. Urine output is normal and the infant is clinically well. Which interpretation is most appropriate?

A. Interpret the value as a direct measurement of neonatal glomerular filtration on the first day

B. Assume maternal creatinine no longer influences the infant once placental circulation has ended

C. Early neonatal creatinine initially reflects maternal creatinine and should be trended over time

D. Diagnose intrinsic neonatal kidney injury because the creatinine exceeds a fixed newborn threshold

51. A 3-day-old infant becomes progressively lethargic and tachypneic. Ammonia is 520 micromol/L, glucose is normal, and there is respiratory alkalosis without a major anion-gap metabolic acidosis. Which category of disorder is most likely?

A. Congenital lactic acidosis causing very high lactate with metabolic acidosis

B. Renal tubular acidosis causing hyperchloremic acidosis without severe hyperammonemia

C. Organic acidemia causing prominent anion-gap metabolic acidosis and ketosis

D. Urea-cycle disorder causing impaired nitrogen disposal

52. A newborn has persistent right-sided nasal obstruction and unilateral mucoid discharge. A small catheter cannot be passed through the right nostril but passes easily through the left. Which diagnosis is most likely?

A. Unilateral choanal atresia producing fixed obstruction of one posterior nasal passage

B. Nasal septal deviation causing partial right-sided obstruction but allowing catheter passage with repositioning

C. Congenital nasolacrimal duct obstruction causing tearing and discharge without posterior nasal blockage

D. Pyriform aperture stenosis causing anterior bony narrowing that is usually bilateral rather than unilateral

53. A newborn has truncus arteriosus, hypocalcemia, and characteristic facial features. Which chromosomal abnormality should be suspected?

A. A 5p deletion causing cri-du-chat syndrome and a high-pitched cry

B. A 15q11-q13 paternal deletion causing Prader-Willi syndrome and neonatal hypotonia

C. A 22q11.2 deletion affecting pharyngeal arch and conotruncal development

D. A 7q11.23 deletion causing Williams syndrome and supravalvar aortic stenosis

54. A 25-week infant on day 4 has a widened pulse pressure, bounding pulses, hyperdynamic precordium, and increasing ventilator needs. Echocardiography shows a large left-to-right ductal shunt. Which diagnosis best explains the findings?

A. Hemodynamically significant patent ductus arteriosus with pulmonary overcirculation

B. Tetralogy of Fallot with dynamic right ventricular outflow tract obstruction

C. Persistent pulmonary hypertension with predominant right-to-left ductal shunting

D. Critical pulmonary valve stenosis with markedly reduced pulmonary blood flow

55. A septic newborn is receiving gentamicin for several days. Which monitoring strategy best reduces toxicity while preserving antibacterial exposure?

A. Follow serum creatinine and urine output alone and omit gentamicin concentrations if both remain stable

B. Measure only a post-infusion peak concentration because accumulation is reflected by peak values alone

C. Use a single untimed gentamicin concentration without reference to when the preceding dose was given

D. Use appropriately timed serum drug concentrations together with renal function and dosing history

56. A preterm infant passes the newborn hearing screen but had prolonged ventilation, meningitis, and ototoxic medication exposure. Which follow-up principle is most appropriate?

A. Repeat hearing screening only if the infant develops visible external-ear abnormalities

B. Continue audiologic surveillance because some high-risk infants develop delayed-onset hearing loss

C. No further hearing evaluation is needed because a passed newborn screen excludes later hearing loss

D. Use only parental response to sound because objective testing has no role after NICU discharge

57. A 29-week infant on full feeds has poor weight gain and serum sodium of 128 mEq/L without edema. Urine sodium is elevated, and renal function is otherwise stable. Which mechanism is most likely?

A. Excess aldosterone action causing pathologic retention of sodium and free water

B. Iatrogenic sodium overload causing dilutional hyponatremia in the absence of fluid excess

C. Central diabetes insipidus causing pure water loss and hypernatremia

D. Renal sodium wasting from immature tubular reabsorption in prematurity

58. A term infant has petechiae and a platelet count of 9,000/microliter immediately after birth but is otherwise well. The mother has a normal platelet count. A previous sibling had an intracranial hemorrhage as a neonate. Which diagnosis is most likely?

A. Maternal immune thrombocytopenia, which usually produces severe maternal thrombocytopenia as the defining clue

B. Disseminated intravascular coagulation from severe neonatal sepsis with multiorgan instability

C. Neonatal alloimmune thrombocytopenia from maternal antibodies against a fetal platelet antigen

D. Thrombocytopenia of placental insufficiency, which is usually mild and develops only after several weeks

59. After a difficult vacuum-assisted delivery, a term infant becomes pale and tachycardic. Examination shows an enlarging diffuse boggy scalp swelling that crosses cranial sutures, and the hematocrit is falling. Which extracranial hemorrhage is most concerning for major blood loss?

A. Cephalohematoma confined beneath periosteum and therefore limited by suture lines

B. Subgaleal hemorrhage with blood collecting in the large potential space beneath the galea

C. Aplasia cutis congenita causing absence of scalp tissue without a blood collection

D. Caput succedaneum consisting of superficial scalp edema with minimal blood loss

60. A 24-week infant develops serum potassium of 7.1 mEq/L on day 2 despite good urine output and no potassium in intravenous fluids. The ECG shows peaked T waves. Which phenomenon is characteristic of extreme prematurity?

A. Pseudohyperkalemia from heel-stick hemolysis despite the associated electrocardiographic changes

B. Adrenal mineralocorticoid deficiency causing impaired potassium excretion in the first 48 hours

C. Nonoliguric hyperkalemia from limited cellular potassium uptake and renal immaturity

D. Oliguric acute kidney injury causing potassium retention despite the documented brisk urine output

61. Investigators enroll preterm infants at birth, classify them according to an exposure, and follow both groups to determine subsequent BPD incidence. What study design is this?

A. Prospective cohort study following exposure groups forward over time

B. Case series without a comparison group

C. Case-control study beginning with infants who already have BPD

D. Randomized crossover trial in which each infant receives both exposures

62. A newborn screen detects homozygous deletion of SMN1 before symptoms develop. Which diagnosis has been identified?

A. Duchenne muscular dystrophy from a dystrophin gene defect

B. Spinal muscular atrophy from biallelic loss of SMN1 function

C. Pompe disease caused by lysosomal acid alpha-glucosidase deficiency

D. Myotonic dystrophy caused by a CTG repeat expansion in DMPK

63. After shoulder dystocia, a newborn holds one arm adducted and internally rotated with the elbow extended and forearm pronated. Hand grasp is preserved. Which injury is most likely?

A. Radial nerve injury causing wrist drop without shoulder abduction weakness

B. Lower brachial plexus injury involving C8-T1 roots with predominant hand weakness

C. Upper brachial plexus injury involving C5-C6 roots, producing Erb palsy

D. Spinal cord transection causing isolated unilateral arm posture with normal legs

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

A. Trisomy 13 producing severe midline malformations with clefting and postaxial polydactyly

B. Monosomy X with neonatal lymphedema and a webbed neck

C. Trisomy 18 with clenched overlapping fingers and rocker-bottom feet

D. Trisomy 21 with a complete atrioventricular septal defect

65. A newborn has profound cyanosis and respiratory distress. Chest radiograph shows pulmonary edema. Echocardiography demonstrates all pulmonary veins entering a vertical vein with severe obstruction before reaching the systemic venous circulation. Which diagnosis is present?

A. Critical pulmonary stenosis causing oligemic rather than edematous pulmonary vascular markings

B. Unobstructed partial anomalous pulmonary venous return causing mild right-sided volume loading

C. Isolated patent ductus arteriosus causing systemic diastolic runoff without venous obstruction

D. Obstructed total anomalous pulmonary venous return requiring urgent intervention

66. A term infant has severe respiratory distress and cyanosis immediately after birth. The abdomen is scaphoid, breath sounds are markedly decreased on the left, and bowel sounds are heard in the left chest. What is the best initial respiratory strategy?

A. Begin nasal CPAP only and defer gastric decompression until radiography is completed

B. Intubate the trachea and decompress the stomach while avoiding bag-mask ventilation

C. Perform immediate thoracentesis because bowel sounds in the chest indicate a pneumothorax

D. Provide vigorous bag-mask ventilation to expand the compressed left lung rapidly

67. A preterm infant with chronic lung disease receives repeated loop diuretic therapy. Which adverse effect deserves specific surveillance?

A. Nephrocalcinosis and electrolyte losses associated with prolonged loop diuretic exposure

B. Permanent closure of the ductus arteriosus from direct cyclooxygenase inhibition

C. Severe hypercalcemia from renal calcium retention caused by loop diuretics

D. Congenital hypothyroidism from suppression of neonatal thyroid hormone synthesis

68. A former 25-week infant undergoes serial retinal examinations. Abnormal vascular proliferation is progressing toward the posterior pole. Which disease is being monitored?

A. Congenital cataract caused by opacity of the lens at birth

B. Congenital glaucoma causing corneal enlargement and tearing

C. Retinoblastoma presenting as leukocoria from an intraocular tumor

D. Retinopathy of prematurity with abnormal retinal vascular proliferation in a very preterm infant

69. A 26-week infant has frequent extubation failures because of increased work of breathing. There is no major airway lesion, sepsis, or cardiac shunt. Which feature of extreme prematurity contributes most to this problem?

A. Highly compliant chest wall with immature respiratory muscles and limited endurance

B. Excessive respiratory-muscle mass that causes hyperventilation and respiratory alkalosis

C. A rigid calcified chest wall that prevents inward recoil during inspiration

D. Mature diaphragmatic fatigue resistance that prevents work-of-breathing limitation

70. A term infant has progressive abdominal distention and bilious emesis and has not passed meconium by 48 hours. Rectal examination produces an explosive discharge of stool and gas. Which diagnosis is most likely?

A. Hirschsprung disease due to distal intestinal aganglionosis

B. Meconium ileus from cystic fibrosis with inspissated distal ileal contents

C. Duodenal atresia causing immediate proximal obstruction with a double-bubble pattern

D. Hypertrophic pyloric stenosis causing nonbilious vomiting after several weeks

71. A newborn with trisomy 21 has leukocytosis with circulating blasts, hepatosplenomegaly, and thrombocytopenia. The blast count falls spontaneously over the next weeks. Which condition is most likely?

A. Congenital acute leukemia with persistent clonal blasts and progressive organ infiltration

B. A severe leukemoid response to infection with predominantly mature neutrophil precursors

C. Transient abnormal myelopoiesis associated with trisomy 21 and neonatal megakaryoblastic proliferation

D. Myeloid leukemia of Down syndrome with a persistent progressive megakaryoblastic clone

72. A newborn has bilateral cataracts, a continuous murmur from patent ductus arteriosus, and failed hearing screening. Which congenital infection best explains this triad?

A. Congenital cytomegalovirus with periventricular calcifications and petechiae

B. Congenital toxoplasmosis with hydrocephalus and intracranial calcifications

C. Congenital rubella syndrome causing cataracts, ductal heart disease, and hearing impairment

D. Congenital syphilis with snuffles and metaphyseal bone lesions

73. A study reports P = 0.03 for the difference in a primary outcome between two randomized groups. Which statement best describes this P value?

A. The treatment effect is necessarily large enough to be clinically important

B. If the null hypothesis were true, data this extreme or more would occur about 3% of the time under the model

C. There is a 3% probability that the null hypothesis itself is true

D. There is a 97% probability that the treatment is clinically effective

74. A 30-week infant is delivered after the mother received magnesium sulfate shortly before birth for severe preeclampsia. The newborn is hypotonic with shallow respirations and diminished deep tendon reflexes. Which explanation is most likely?

A. Placental calcium transfer causing acute neonatal hypercalcemia with central apnea

B. Fetal catecholamine excess causing transient depletion of acetylcholine after birth

C. Maternal beta-blocker exposure causing isolated neuromuscular junction blockade

D. Transplacental magnesium exposure causing neonatal neuromuscular depression

75. A 28-week infant is tolerating full volumes of expressed human milk but has poor linear growth and low serum phosphorus. What nutritional intervention is usually needed?

A. Stop enteral feeding and use dextrose-only intravenous fluids until growth improves

B. Dilute human milk with sterile water to reduce renal solute load and phosphorus intake

C. Fortify human milk to increase protein, mineral, and caloric delivery

D. Replace all human milk with unfortified term formula because fortifiers are contraindicated in prematurity

76. A profoundly cyanotic newborn has pulmonary atresia with an intact ventricular septum and minimal antegrade pulmonary blood flow. Which medication should be started while planning definitive management?

A. Prostaglandin E1 to maintain ductal pulmonary blood flow

B. Indomethacin to close the ductus and reduce pulmonary artery pressure

C. Adenosine to recruit pulmonary blood flow by slowing atrioventricular conduction

D. Furosemide to create a compensatory right-to-left shunt through the ductus

77. A 24-week infant with a central venous catheter and prolonged broad-spectrum antibiotics develops thrombocytopenia and persistent sepsis with blood cultures growing Candida albicans. Which complication should be actively considered?

A. Transient culture contamination because Candida in neonatal blood cultures is usually clinically insignificant

B. Localized oral thrush only because Candida rarely invades the bloodstream of preterm infants

C. Disseminated candidiasis with possible central nervous system, renal, or ocular involvement

D. Isolated viral meningitis because fungemia does not spread hematogenously to neonatal organs

78. A 24-week infant receiving mechanical ventilation develops progressive oxygen need. Chest radiograph shows multiple irregular linear and cystic lucencies extending from the hila through the lung fields. Which complication is most likely?

A. Pulmonary interstitial emphysema from air dissecting into the pulmonary interstitium

B. Pleural effusion producing homogeneous dependent opacity without intrapulmonary lucencies

C. Atelectasis from endotracheal tube obstruction producing uniform volume loss of both lungs

D. Diffuse pulmonary hemorrhage producing only bilateral alveolar opacities without air dissection

79. Two NICUs use the same screening test, but disease prevalence is much higher in NICU A. Assuming identical sensitivity and specificity, which test characteristic is expected to be higher in NICU A?

A. Specificity

B. Negative likelihood ratio

C. Sensitivity

D. Positive predictive value

80. A 12-hour-old infant has rapidly rising unconjugated bilirubin, anemia, reticulocytosis, and a positive direct antiglobulin test. Which process is most likely?

A. Immune-mediated hemolysis with antibody-coated neonatal erythrocytes

B. Breast milk jaundice causing reticulocytosis and a positive direct antiglobulin test

C. Biliary atresia causing isolated conjugated hyperbilirubinemia and pale stools

D. Physiologic bilirubin production with no increase in red-cell destruction

81. A former 25-week infant is approaching NICU discharge after a complicated course. Which follow-up plan best addresses the infant's developmental risk?

A. Delay all developmental assessment until school age so early variability does not affect interpretation

B. Routine primary care only because prematurity-related developmental risk ends at hospital discharge

C. Structured high-risk follow-up with standardized developmental surveillance and early-intervention referral

D. Refer for therapy only after a fixed major disability is established and cannot be modified

82. A preterm infant receiving prolonged fat-free parenteral nutrition develops dermatitis, thrombocytopenia, poor growth, and alopecia. Which deficiency is most likely?

A. Vitamin B12 excess from over-supplementation of parenteral nutrition

B. Excessive medium-chain triglyceride intake causing loss of essential fatty acids

C. Isolated sodium deficiency causing a characteristic eczematous rash and alopecia

D. Essential fatty acid deficiency caused by prolonged nutrition without an adequate lipid source

83. A newborn develops poor feeding, cool lower extremities, weak femoral pulses, metabolic acidosis, and shock as the ductus arteriosus begins to close. Which initial therapy is most appropriate while defining the anatomy?

A. Start prostaglandin E1 to reopen or maintain ductal systemic blood flow

B. Restrict fluids and begin diuresis before restoring systemic perfusion

C. Give indomethacin to accelerate ductal closure and reduce systemic steal

D. Administer inhaled nitric oxide as sole therapy for presumed pulmonary vasoconstriction

84. A screening test for congenital disease is positive in 95 of 100 affected infants and negative in 810 of 900 unaffected infants. What is the sensitivity of the test?

A. 50%

B. 90%

C. 99%

D. 95%

85. A 39-week infant born by elective cesarean delivery without labor develops tachypnea soon after birth. Oxygen need is minimal, and chest radiograph shows hyperinflation, perihilar streaking, and fluid in the fissures. Which diagnosis is most likely?

A. Respiratory distress syndrome caused by severe surfactant deficiency in a term infant

B. Persistent pulmonary interstitial emphysema from prolonged high-pressure ventilation

C. Transient tachypnea of the newborn due to delayed fetal lung fluid absorption

D. Congenital diaphragmatic hernia with bowel occupying the left hemithorax

86. A 28-week infant develops early-onset sepsis with shock and meningitis after prolonged rupture of membranes. Which gram-negative organism is particularly important in very preterm infants?

A. Vibrio cholerae causing neonatal meningitis through ascending genital tract infection

B. Escherichia coli as an important gram-negative cause of early-onset sepsis in very preterm infants

C. Pseudomonas aeruginosa acquired almost exclusively through breast milk at delivery

D. Salmonella Typhi as the dominant cause of routine early-onset sepsis in preterm infants

87. In a randomized trial of a feeding strategy, 20% of controls and 10% of treated infants develop NEC. What is the number needed to treat to prevent one NEC case?

A. 2

B. 20

C. 5

D. 10

88. A 29-week infant born to a mother with severe preeclampsia has an absolute neutrophil count of 700/microliter on day 1 but is clinically well. Which association is most likely?

A. Congenital leukemia causing isolated mild neutropenia without blasts or organomegaly

B. Severe combined immunodeficiency causing selective neutropenia while lymphocyte counts remain normal

C. Transient neonatal neutropenia associated with maternal hypertension and placental insufficiency

D. Vitamin K deficiency causing suppression of neutrophil production in the neonatal marrow

89. A newborn has diffuse maculopapular rash involving palms and soles, hepatosplenomegaly, anemia, and radiographic metaphyseal abnormalities. The mother had untreated syphilis during pregnancy. Which diagnosis is most likely?

A. Congenital toxoplasmosis with hydrocephalus and chorioretinitis

B. Congenital syphilis causing rash, hepatosplenomegaly, cytopenias, and long-bone abnormalities

C. Congenital rubella syndrome with cataracts and patent ductus arteriosus

D. Neonatal herpes simplex infection with grouped vesicles and temporal-lobe encephalitis

90. A newborn has severe cyanosis, cardiomegaly, and a holosystolic murmur. The mother used lithium during early pregnancy. Echocardiography shows apical displacement of the tricuspid valve with atrialization of the right ventricle. What is the diagnosis?

A. Complete atrioventricular septal defect with a common atrioventricular valve

B. Tetralogy of Fallot with a large ventricular septal defect and pulmonary stenosis

C. Critical aortic stenosis with left ventricular outflow obstruction and ductal dependence

D. Ebstein anomaly with apical displacement of the tricuspid valve and atrialization of the right ventricle

91. A term newborn has electrographic seizures after HIE. Glucose, calcium, sodium, and magnesium are normal. Which antiseizure medication is commonly used as first-line therapy for acute neonatal seizures?

A. Phenobarbital as commonly used first-line therapy for acute neonatal electrographic seizures

B. Levetiracetam as the preferred first-line drug for all acute neonatal seizures after HIE

C. Fosphenytoin as the routine first-line agent before phenobarbital for HIE-associated seizures

D. Continuous midazolam infusion as initial monotherapy before a loading antiseizure medication

92. A former 25-week infant has received prolonged furosemide therapy. Renal ultrasonography shows bilateral medullary echogenic foci. Which complication is most likely?

A. Nephrocalcinosis related to hypercalciuria from loop diuretic exposure

B. Renal vein thrombosis caused directly by loop diuretic inhibition of platelet function

C. Posterior urethral valves caused by postnatal diuretic exposure during nephron maturation

D. Polycystic kidney disease caused by acquired collecting-duct cyst formation from furosemide

93. A neonate has vertebral segmentation defects, imperforate anus, a ventricular septal defect, tracheoesophageal fistula, renal dysplasia, and radial ray abnormality. Which diagnostic label is most appropriate?

A. CHARGE syndrome with coloboma and choanal atresia as defining features

B. Beckwith-Wiedemann syndrome with overgrowth and macroglossia

C. Noonan syndrome with pulmonary stenosis and a RAS-MAPK disorder

D. VACTERL association involving multiple characteristic component anomalies

94. A term infant is born after maternal fever, uterine tenderness, fetal tachycardia, and prolonged rupture of membranes. The infant becomes tachypneic and poorly perfused at 2 hours of age. Which maternal condition most increases concern for early-onset neonatal sepsis?

A. Isolated maternal iron-deficiency anemia treated with oral supplementation in pregnancy

B. Elective cesarean delivery before labor with intact membranes and no maternal infection

C. Uncomplicated gestational hypertension without maternal fever or membrane rupture

D. Intraamniotic infection associated with maternal fever and prolonged rupture of membranes

95. A 26-week infant has much lower serum IgG than a term infant. Which developmental fact best explains this difference?

A. Placental IgG transport is greatest early in gestation and diminishes progressively during the third trimester

B. Most transplacental maternal IgG transfer occurs during the third trimester

C. Fetal B cells cannot express any immunoglobulin until after 40 weeks gestation

D. Maternal IgM crosses the placenta mainly during the first trimester and replaces IgG

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

A. Congenital cytomegalovirus with predominantly periventricular calcifications and microcephaly

B. Congenital rubella with cataracts, patent ductus arteriosus, and hearing impairment

C. Congenital toxoplasmosis producing hydrocephalus, intracranial calcifications, and chorioretinitis

D. Congenital syphilis with snuffles, rash, hepatosplenomegaly, and long-bone abnormalities

97. A 9-day-old infant has fever, lethargy, focal seizures, and clusters of vesicles on the scalp. The mother had no recognized genital lesions at delivery. What treatment should be started immediately?

A. Empiric antibacterial therapy alone while HSV testing is pending because maternal lesions were absent

B. Intravenous acyclovir while evaluating for neonatal herpes simplex virus infection

C. Oral acyclovir while awaiting cerebrospinal fluid HSV testing in the clinically ill newborn

D. Topical acyclovir for the vesicles with observation if the initial neurologic examination is reassuring

98. A newborn is irritable, tachycardic, and feeding poorly. The mother has Graves disease treated before pregnancy and currently has normal thyroid function. The infant's free thyroxine is elevated and TSH is suppressed. What is the most likely cause?

A. Neonatal pituitary TSH overproduction causing secondary elevation of circulating thyroxine

B. Transplacental maternal TSH-receptor-stimulating antibodies causing neonatal thyrotoxicosis

C. Transplacental maternal thyroid hormone causing prolonged neonatal hyperthyroidism after birth

D. Placental transfer of antithyroid medication causing autonomous neonatal thyroid hormone release

99. Prenatal ultrasonography demonstrates a multicystic mass within one fetal lung. After birth, the infant has respiratory distress and imaging confirms multiple air-filled cysts supplied by the pulmonary circulation. Which diagnosis is most likely?

A. Transient tachypnea caused by delayed absorption of fetal alveolar fluid

B. Bronchopulmonary sequestration supplied exclusively by the normal pulmonary arterial tree

C. Congenital pulmonary airway malformation producing a multicystic lesion supplied by pulmonary circulation

D. Pulmonary interstitial emphysema caused by postnatal positive-pressure ventilation

100. A preterm infant on full enteral feeds has faltering growth despite adequate volume. Which assessment best distinguishes true nutrient insufficiency from simple fluid-related weight change?

A. Trend weight velocity and serum prealbumin without measuring length or head circumference

B. Follow serial weight, length, and head circumference together with actual nutrient intake

C. Use weight-for-age z score alone because serial length measures are too variable to guide nutrition

D. Use weekly blood urea nitrogen alone as a surrogate for both protein and energy adequacy

101. A growth-restricted newborn is plethoric and sleepy. Venous hematocrit is 72% and glucose is low. Which complication is most directly related to the high red-cell mass?

A. Severe bleeding from dilutional deficiency of clotting factors caused by too few erythrocytes

B. High-output heart failure caused by profound anemia and low oxygen-carrying capacity

C. Isolated unconjugated bilirubin deficiency because increased red cells reduce bilirubin production

D. Hyperviscosity with impaired microvascular perfusion caused by the markedly increased neonatal red-cell mass

102. A newborn exposed in utero to chronic maternal opioid therapy develops tremors, high-pitched crying, loose stools, poor sleep, and increased muscle tone. Which pathophysiologic process is most likely?

A. Selective serotonin toxicity caused by increased central serotonin after placental separation

B. Autoimmune destruction of neonatal autonomic ganglia triggered by maternal opioid exposure

C. Withdrawal after abrupt interruption of chronic fetal opioid receptor stimulation

D. Persistent opioid intoxication causing progressive hypotonia and absent gastrointestinal motility

103. A family with limited English proficiency is discussing withdrawal of life-sustaining treatment for their critically ill newborn. Which communication strategy is best?

A. Avoid discussing prognosis until a family member becomes fluent in the clinical team's language

B. Use an older sibling as the primary interpreter because family members understand medical nuance best

C. Use a qualified medical interpreter and speak directly with the parents in clear, respectful language

D. Rely on automated translation alone for consent because professional interpreters can introduce bias

104. A 3-week-old infant remains jaundiced. Total bilirubin is 7 mg/dL and direct bilirubin is 4 mg/dL. The infant has dark urine and poor weight gain. Which statement is most accurate?

A. This is physiologic jaundice because direct bilirubin normally predominates after two weeks

B. No evaluation is needed until the direct bilirubin exceeds the total bilirubin concentration

C. Breast milk jaundice usually causes marked direct hyperbilirubinemia with dark urine

D. Conjugated hyperbilirubinemia is abnormal and requires evaluation for neonatal cholestasis

105. A former 25-week infant is now 36 weeks postmenstrual age and still requires supplemental oxygen after weeks of mechanical ventilation. Chest imaging shows coarse interstitial markings and areas of hyperinflation. What diagnosis best explains the chronic lung disease?

A. Bronchopulmonary dysplasia related to arrested lung development and neonatal lung injury

B. Persistent pulmonary hypertension causing hypoxemia without explaining the chronic parenchymal changes

C. Postinfectious bronchiolitis obliterans following an unrecognized neonatal lower respiratory infection

D. Congenital lobar overinflation causing a focal hyperlucent lobe rather than diffuse chronic lung disease

106. Prenatal ultrasonography shows severe oligohydramnios, echogenic kidneys, and poor calvarial ossification after maternal use of an angiotensin-converting enzyme inhibitor late in pregnancy. Which fetal effect is most characteristic?

A. Increased fetal renal blood flow causing polyhydramnios and excessive urine production

B. Premature closure of the ductus arteriosus from inhibition of prostaglandin synthesis

C. Direct fetal pancreatic toxicity causing hyperinsulinemia and macrosomia

D. Reduced fetal renal perfusion with renal dysfunction and oligohydramnios sequence

107. A newborn has granulomatous skin lesions, respiratory distress, sepsis, and a maternal history of febrile gastroenteritis with consumption of unpasteurized dairy products. Which organism should be considered?

A. Listeria monocytogenes causing transplacental or perinatal sepsis with granulomatous skin lesions

B. Streptococcus pneumoniae causing a pathognomonic congenital skin granuloma syndrome

C. Neisseria gonorrhoeae causing disseminated infection with granulomas at birth

D. Bordetella pertussis causing congenital granulomatous disease through placental transmission

108. A preterm infant with severe intraventricular hemorrhage develops rapidly increasing head circumference, a full fontanelle, and progressive ventricular enlargement on ultrasound. Which complication is most likely?

A. Posthemorrhagic ventricular dilation from impaired cerebrospinal fluid circulation and resorption

B. Craniosynostosis causing premature suture closure and enlarged ventricles from excess skull volume

C. Subgaleal hemorrhage causing intraventricular dilation through scalp blood accumulation

D. Benign macrocephaly caused by accelerated skull growth without ventricular enlargement

109. A 26-week infant has adequate weight gain but poor length and head-circumference growth. Which nutritional component is especially important for lean-tissue and linear growth?

A. High dextrose delivery alone with minimal amino acid intake

B. Restriction of all dietary protein to reduce blood urea nitrogen

C. Adequate protein delivery together with sufficient nonprotein energy

D. Water supplementation without additional calories or amino acids

110. A term infant has episodes of abnormal movements, but bedside observation is uncertain. Which test is the gold standard for determining whether the events are epileptic seizures?

A. Continuous multichannel video EEG monitoring to correlate clinical events with cortical electrical activity

B. Amplitude-integrated EEG alone without simultaneous conventional multichannel video EEG

C. Cranial ultrasonography performed during the abnormal movements to look for acute hemorrhage

D. Cardiorespiratory event monitoring with pulse oximetry and electrocardiography but no brain recording

111. A neonate with severe respiratory failure is being considered for extracorporeal support. Which underlying feature generally makes ECMO least feasible?

A. Meconium aspiration in a term infant with potentially reversible lung disease

B. Congenital diaphragmatic hernia in a selected infant managed at an experienced center

C. Extreme prematurity with very high risk of catastrophic intracranial hemorrhage

D. Reversible pulmonary hypertension in a near-term infant despite maximal conventional therapy

112. A 20-day-old infant has fever and poor feeding. Urinalysis shows pyuria and a catheterized urine culture grows a single uropathogen. Which additional evaluation is commonly considered after a confirmed neonatal urinary tract infection?

A. Renal and bladder ultrasonography to assess for structural urinary tract abnormalities

B. Voiding cystourethrography during the acute infection before any screening renal ultrasonography

C. Dimercaptosuccinic acid renal scan during the acute episode as the first structural study

D. Repeat urine culture after therapy without urinary tract imaging after a first neonatal infection

113. In a cohort study, BPD occurs in 30 of 100 exposed infants and 15 of 100 unexposed infants. What is the relative risk associated with the exposure?

A. 0.5

B. 15.0

C. 1.5

D. 2.0

114. A term newborn is apneic after initial warming, positioning, drying, and stimulation. Heart rate is 80 beats/min. What is the most important next action?

A. Observe for another minute because the heart rate remains above 60 beats/min

B. Begin effective positive-pressure ventilation and assess for a rising heart rate

C. Give intravenous epinephrine before establishing effective lung inflation

D. Start chest compressions immediately before attempting assisted ventilation

115. A newborn has severe anemia, jaundice within the first hours of life, and a strongly positive direct antiglobulin test. The mother is Rh(D)-negative and was previously sensitized. Which process best explains the infant's disease?

A. Fetal antibodies crossed into the maternal circulation and destroyed maternal erythrocytes

B. Maternal complement proteins crossed the placenta and directly lysed fetal erythrocytes without antibody

C. Maternal IgM antibodies crossed the placenta and caused intravascular fetal hemolysis

D. Maternal IgG antibodies crossed the placenta and destroyed fetal erythrocytes

116. A newborn has a firm scalp swelling over the right parietal bone after a difficult delivery. The swelling does not cross suture lines and becomes more apparent several hours after birth. Which diagnosis is most likely?

A. Cephalohematoma, a subperiosteal blood collection that remains limited by cranial suture lines

B. Caput succedaneum that is present at birth and crosses suture lines as superficial edema

C. Subgaleal hemorrhage that crosses sutures and can cause massive blood loss

D. Encephalocele containing intracranial tissue through a skull defect

117. A 32-week infant with RDS is stabilized on noninvasive support but requires increasing inspired oxygen. The team wants to give surfactant while minimizing exposure to prolonged invasive ventilation. Which strategy best fits this goal?

A. Intubate for surfactant and continue conventional ventilation until the oxygen requirement is minimal

B. Increase noninvasive pressure alone and defer surfactant despite a steadily rising oxygen requirement

C. Use a minimally invasive surfactant approach through a thin catheter while maintaining spontaneous breathing

D. Switch to high-flow nasal cannula and reserve surfactant for later recurrent apnea or severe failure

118. A dark-skinned term newborn has superficial pustules present at birth that rupture easily, leaving hyperpigmented macules with a collarette of scale. The infant is otherwise well. What is the most likely diagnosis?

A. Erythema toxicum neonatorum, which usually begins after birth on an erythematous base

B. Bullous impetigo producing flaccid bullae with bacterial illness rather than benign pigmented macules

C. Neonatal HSV infection producing grouped vesicles that heal without residual pigmentation

D. Transient neonatal pustular melanosis with fragile pustules that leave pigmented macules and collarettes

119. A 10-day-old male infant has vomiting, dehydration, hypotension, sodium 122 mEq/L, potassium 7.0 mEq/L, and glucose 48 mg/dL. Which diagnosis should be suspected?

A. Central diabetes insipidus causing hypernatremic dehydration with dilute urine

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

C. Congenital hypothyroidism causing isolated low thyroxine without electrolyte crisis

D. Primary hyperaldosteronism causing hypertension, hypokalemia, and sodium retention

120. A stable neonate suddenly develops a regular narrow-complex tachycardia at 250 beats/min. Vagal maneuvers fail, blood pressure remains adequate, and an intravenous line is present. What is the preferred next treatment?

A. Administer prostaglandin E1 because all neonatal tachyarrhythmias are ductal dependent

B. Give slow intravenous atropine to block atrioventricular nodal conduction over several minutes

C. Begin unsynchronized defibrillation despite preserved perfusion and a regular narrow complex

D. Give rapid intravenous adenosine for presumed supraventricular tachycardia

121. A very preterm infant on fortified human milk has elevated alkaline phosphatase and low serum phosphorus. Radiographs show metaphyseal demineralization. Which diagnosis is most likely?

A. Metabolic bone disease of prematurity from inadequate mineral accretion

B. Scurvy from isolated vitamin C deficiency causing generalized increased bone density

C. Achondroplasia caused by impaired endochondral growth without mineral deficiency

D. Osteopetrosis from excessive calcium deposition throughout the skeleton

122. A treatment trial reports a relative risk of death of 0.82 with a 95% confidence interval from 0.66 to 1.04. Which interpretation is most appropriate?

A. The treatment definitely increases mortality because the upper confidence limit exceeds 1.0

B. The interval includes 1.0, so the result is not statistically significant at the conventional 0.05 level

C. The result proves equivalence because the confidence interval spans both benefit and harm

D. The treatment definitely reduces mortality because the point estimate is below 1.0

123. A dehydrated newborn with sepsis develops gross hematuria, thrombocytopenia, an enlarged tender left kidney, and a new left-sided abdominal mass. Which diagnosis is most likely?

A. Nephrocalcinosis causing bilateral medullary calcifications without renal enlargement

B. Posterior urethral valves causing isolated lower urinary tract obstruction

C. Minimal-change nephrotic syndrome causing isolated proteinuria without hematuria or thrombocytopenia

D. Renal vein thrombosis producing hematuria, thrombocytopenia, and unilateral renal enlargement

124. A family of a technology-dependent former preterm infant reports difficulty attending multiple specialty visits and obtaining therapy services. Which intervention best supports neurodevelopmental outcomes?

A. Coordinate multidisciplinary follow-up and address transportation, caregiver, language, and social barriers to care

B. Focus only on growth measurements because family stress and access do not affect developmental outcomes

C. Require each specialty to work independently so information is not shared across disciplines

D. Reduce follow-up because social barriers indicate the family is unlikely to benefit from developmental services

125. A newborn has intermittent tremulous movements that stop when the examiner gently flexes and holds the limb. The infant remains alert, and there is no eye deviation or autonomic change. Which phenomenon is most likely?

A. Tonic seizure because suppressibility with gentle restraint is diagnostic of epilepsy

B. Infantile spasms because neonatal seizures typically occur in symmetric flexor clusters

C. Focal clonic seizure because all neonatal tremulous movements are epileptic

D. Jitteriness rather than an epileptic seizure because the tremulous movement is stimulus-sensitive and suppressible

126. Parents of an extremely preterm infant ask why corrected age is used during early developmental follow-up. What is the best explanation?

A. Corrected age is used only to calculate medication doses and not developmental expectations

B. Corrected age adds the number of premature weeks to chronologic age, making the infant appear older

C. Corrected age accounts for the weeks of prematurity when comparing early development with term-born peers

D. Corrected age permanently replaces chronologic age for all medical decisions throughout adulthood

127. A critically ill preterm infant with sepsis develops oozing from venipuncture sites, thrombocytopenia, prolonged PT and aPTT, low fibrinogen, and elevated D-dimer. Which diagnosis is most likely?

A. Isolated vitamin K deficiency with normal fibrinogen and no evidence of fibrin breakdown

B. Disseminated intravascular coagulation with consumption of platelets and coagulation factors

C. Hemophilia A causing isolated prolongation of aPTT with normal platelets and fibrinogen

D. Neonatal alloimmune thrombocytopenia causing isolated severe thrombocytopenia in an otherwise well infant

128. A newborn's heart rate remains 50 beats/min despite effective positive-pressure ventilation with visible chest movement and ventilation corrective steps. What should be done next?

A. Begin coordinated chest compressions while continuing effective ventilation

B. Administer sodium bicarbonate as the first medication before starting compressions

C. Delay intervention because neonatal bradycardia is expected during the first 10 minutes

D. Stop ventilation and provide chest compressions alone until the heart rate rises

129. A newborn has asymmetric crying: the right lower face does not move, and the right eyelid cannot close completely. The forehead is also weak. Which lesion is most likely?

A. Isolated depressor anguli oris hypoplasia because eyelid closure and forehead function are preserved

B. Peripheral facial nerve palsy from birth-related nerve injury

C. Upper motor neuron lesion because forehead weakness is usually spared in a central facial palsy

D. Brachial plexus injury because the facial nerve travels within the brachial plexus

130. A 31-week infant receiving fortified human milk develops repeated emesis and abdominal distention. Examination shows tenderness and bloody stool, and radiography demonstrates pneumatosis intestinalis. What should happen to enteral nutrition?

A. Switch immediately to unfortified cow's milk and continue feeding through the acute illness

B. Continue the same feeds because blood in the stool is expected during routine fortification

C. Increase feed volume because pneumatosis indicates inadequate enteral stimulation

D. Stop enteral feeds and begin treatment for suspected necrotizing enterocolitis

131. A term infant with severe perinatal asphyxia develops hypotension, hepatomegaly, gallop rhythm, and elevated cardiac troponin. Echocardiography shows globally depressed ventricular function. What is the most likely mechanism?

A. Primary hypertrophic cardiomyopathy from maternal diabetes with isolated septal thickening

B. Hypoxic-ischemic myocardial injury causing transient ventricular dysfunction

C. Ductal steal from a small restrictive PDA causing severe biventricular myocardial necrosis

D. Congenital absence of the pericardium causing acute global systolic failure after birth

132. A breastfed preterm infant is approaching discharge after rapid growth. Which micronutrient issue requires particular attention because preterm infants have lower stores at birth and high erythropoietic needs?

A. Vitamin D supplementation because preterm infants also have limited stores and high skeletal needs

B. Folate supplementation because rapid erythropoiesis can increase folate requirements during growth

C. Iron supplementation because preterm infants have low stores and high erythropoietic requirements

D. Copper supplementation because deficiency can contribute to anemia and neutropenia in selected infants

133. In a randomized trial, several infants assigned to the intervention cross over and receive the control therapy. The primary analysis keeps them in the groups to which they were originally randomized. What principle is being used?

A. Per-protocol analysis

B. Intention-to-treat analysis

C. Interim subgroup analysis

D. Case-control matching

134. An infant of a diabetic mother develops jitteriness at 18 hours. Total calcium is low and magnesium is also reduced. Which endocrine disturbance is most likely?

A. Early neonatal hypocalcemia related to impaired parathyroid response, worsened by hypomagnesemia

B. Pseudohypoaldosteronism causing isolated calcium loss without sodium or potassium abnormalities

C. Vitamin D intoxication causing simultaneous hypercalcemia and hypermagnesemia

D. Primary hyperparathyroidism causing excessive calcium release from bone and hypercalcemia

135. A 28-week infant receives caffeine for apnea of prematurity. Which pharmacokinetic feature is typical in very preterm infants?

A. Rapid hepatic CYP1A2 metabolism producing a short elimination half-life in extreme prematurity

B. Marked first-pass hepatic metabolism causing poor bioavailability after enteral caffeine dosing

C. A very small volume of distribution because total body water is low in very preterm infants

D. A long elimination half-life because hepatic metabolism and renal elimination are immature

136. A newborn has a lethal genetic condition with escalating respiratory failure. The parents choose comfort-focused care after informed discussions. Which plan is most appropriate?

A. Stop all clinical care because choosing comfort means no medications or nursing interventions are appropriate

B. Provide active palliative care focused on relief of pain, dyspnea, agitation, and family support

C. Separate the infant from the family so staff can manage symptoms without emotional interference

D. Continue burdensome invasive procedures automatically because palliative care cannot coexist with neonatal treatment

137. A newborn has coloboma, congenital heart disease, choanal atresia, genital hypoplasia, and abnormal external ears. Which syndrome best unifies these findings?

A. VACTERL association, which requires vertebral, anal, cardiac, tracheoesophageal, renal, or limb anomalies

B. Pierre Robin sequence, defined by micrognathia, glossoptosis, and upper-airway obstruction

C. CHARGE syndrome, commonly associated with pathogenic CHD7 variants

D. Turner syndrome, characterized by monosomy X and lymphatic abnormalities

138. A newborn with gastroschisis has exposed bowel placed in a silo after delivery. Which nutritional plan is most appropriate initially?

A. Use only free water through a feeding tube until the abdominal wall is completely closed

B. Use parenteral nutrition until bowel function recovers, then introduce enteral feeds gradually

C. Avoid all nutrition for several weeks because parenteral amino acids impair bowel healing

D. Begin full-volume oral feeds immediately because exposed bowel has normal motility after reduction

139. A 3-week-old exclusively breastfed infant who did not receive vitamin K prophylaxis presents with intracranial bleeding. PT is markedly prolonged. Which diagnosis is most likely?

A. Vitamin K deficiency bleeding after omission of routine neonatal vitamin K prophylaxis

B. Hemophilia B causing procedural or spontaneous bleeding with an isolated prolonged aPTT

C. Disseminated intravascular coagulation from occult sepsis with thrombocytopenia and low fibrinogen

D. Neonatal liver failure causing reduced synthesis of multiple clotting factors and prolonged PT

140. A diagnostic test is negative in 880 of 900 infants who do not have the disease. What is the specificity?

A. 88.0%

B. 2.2%

C. 97.8%

D. 90.0%

141. A ventilated preterm infant with meningitis develops oliguria, serum sodium of 122 mEq/L, low serum osmolality, and inappropriately concentrated urine. Which disorder best explains the hyponatremia?

A. Renal sodium retention from primary hyperaldosteronism causing hypernatremia

B. Central diabetes insipidus causing excessive free-water loss and dilute urine

C. Syndrome of inappropriate antidiuretic hormone secretion

D. Physiologic contraction of extracellular volume causing maximally dilute urine despite hyponatremia

142. A pregnant patient at 29 weeks has preterm premature rupture of membranes and is expected to deliver within 48 hours. Which antenatal intervention most directly lowers the infant's risk of respiratory distress syndrome and intraventricular hemorrhage?

A. Administer magnesium sulfate primarily for fetal neuroprotection before the anticipated preterm birth

B. Administer a course of antenatal corticosteroids before the anticipated preterm delivery

C. Start latency antibiotics for preterm membrane rupture to reduce infection and prolong pregnancy

D. Use short-term tocolysis when appropriate to allow transfer and completion of other antenatal therapy

143. Investigators identify infants with NEC and matched infants without NEC, then compare prior transfusion exposure. What study design is this?

A. Prospective randomized trial assigning exposure before NEC outcomes develop

B. Cross-sectional survey measuring exposure and NEC status at one time point

C. Case-control study comparing prior exposure in infants with and without NEC

D. Ecologic time-series comparing unit-level exposure trends with NEC rates

144. A newborn on CPAP suddenly requires more oxygen. Breath sounds remain symmetric, the abdomen is markedly distended, and the diaphragm is elevated on radiograph without an air leak. Which simple intervention may improve ventilation?

A. Decompress the stomach with an orogastric tube while continuing respiratory support

B. Increase CPAP because the rising oxygen need is most likely from alveolar derecruitment alone

C. Reduce CPAP and change to high-flow support to decrease swallowed air without gastric decompression

D. Hold feeds and prioritize evaluation for necrotizing enterocolitis before addressing the distended stomach

145. A 5-day-old exclusively breastfed term infant has lost 13% of birth weight and is lethargic. Serum sodium is 158 mEq/L, and the mother reports painful latch and very few wet diapers. What is the most likely primary problem?

A. Excess mineralocorticoid secretion causing sodium retention despite abundant free-water intake

B. Inadequate milk transfer causing free-water deficit and hypernatremic dehydration

C. Renal salt wasting from congenital adrenal hyperplasia causing hypernatremia and weight loss

D. Syndrome of inappropriate antidiuretic hormone secretion causing concentrated sodium in the serum

146. A 25-week infant has a routine cranial ultrasound on day 3 showing hemorrhage confined to the germinal matrix without ventricular extension. Which diagnosis is most accurate?

A. Diffuse hypoxic-ischemic cortical injury typical of a term infant

B. Grade I germinal matrix-intraventricular hemorrhage

C. Subdural hemorrhage caused by traumatic separation of dural layers

D. Periventricular leukomalacia with cystic white-matter injury and no hemorrhage

147. A 1-hour-old term infant born to a mother with poorly controlled pregestational diabetes is jittery. The infant is large for gestational age and has a plasma glucose concentration of 28 mg/dL. Which maternal-fetal mechanism most directly explains this finding?

A. Persistent fetal hyperinsulinemia after interruption of the maternal glucose supply

B. Transient neonatal cortisol excess after placental separation from maternal hormones

C. Reduced fetal glycogen deposition caused by chronic maternal hyperglycemia during pregnancy

D. Abrupt loss of maternally transferred insulin after delivery with delayed pancreatic secretion

148. A newborn develops widespread blistering and skin erosions with minimal friction during routine handling. Similar disease affected several relatives. Which inherited disorder is most likely?

A. Nevus simplex causing blanching vascular patches from superficial capillary dilation

B. Erythema toxicum causing sterile pustules without true skin fragility

C. Epidermolysis bullosa causing mechanical fragility at the dermal-epidermal junction

D. Milia caused by keratin-filled cysts that rupture with routine pressure

149. A healthy term infant develops scattered erythematous macules and papules with small central pustules on the trunk at 36 hours of age. Palms and soles are spared, and the infant is afebrile and feeding well. Which diagnosis is most likely?

A. Staphylococcal scalded skin syndrome with tenderness and widespread superficial desquamation

B. Neonatal herpes simplex infection with grouped vesicles and possible systemic illness

C. Congenital candidiasis with diffuse pustules often present at or soon after birth

D. Erythema toxicum neonatorum, a benign pustular eruption in an otherwise well newborn

150. A 35-week infant has respiratory distress after a prolonged difficult delivery. Chest radiograph shows elevation of the right hemidiaphragm with paradoxical motion on ultrasonography. Which diagnosis is most likely?

A. Respiratory distress syndrome causing focal phrenic nerve dysfunction after delivery

B. Bilateral choanal atresia causing diaphragmatic eventration on one side

C. Transient tachypnea causing isolated paradoxical movement of a hemidiaphragm

D. Phrenic nerve injury causing diaphragmatic paralysis

151. A newborn has recurrent bacterial and fungal infections, minimal pus formation, and delayed separation of the umbilical cord. Which immune defect should be suspected?

A. Selective IgA deficiency causing absent neutrophil adhesion and delayed cord separation

B. Leukocyte adhesion deficiency causing impaired neutrophil migration into tissues

C. X-linked agammaglobulinemia causing neonatal neutrophil adhesion failure before maternal IgG wanes

D. Terminal complement deficiency causing absent neutrophil rolling in all tissues

152. A 900-g infant on parenteral nutrition has persistent glucose values of 190 to 220 mg/dL. Which initial nutritional consideration is most appropriate?

A. Give repeated large insulin boluses without first reviewing dextrose delivery or clinical causes

B. Stop all amino acids because protein is the usual direct cause of neonatal hyperglycemia

C. Increase glucose infusion because hyperglycemia indicates inadequate cellular glucose exposure

D. Review the glucose infusion rate and reduce excessive dextrose delivery while maintaining needed nutrition

153. A 27-week infant on enteral feeds develops abdominal distention, bloody stools, temperature instability, and apnea. Abdominal radiograph shows pneumatosis intestinalis. Which diagnosis is most likely?

A. Necrotizing enterocolitis with systemic illness, bloody stool, abdominal distention, and pneumatosis intestinalis

B. Hirschsprung disease causing isolated distal obstruction without bowel-wall pneumatosis

C. Physiologic feeding intolerance causing bloody stool with pathognomonic intramural gas

D. Pyloric stenosis causing nonbilious projectile vomiting and hypochloremic alkalosis

154. A term infant after severe hypoxic-ischemic injury develops brisk urine output of 8 mL/kg/h, rising serum sodium, high serum osmolality, and very dilute urine. Which diagnosis is most likely?

A. Syndrome of inappropriate antidiuretic hormone secretion causing water retention and concentrated urine

B. Cerebral salt wasting causing renal sodium loss, polyuria, and progressive hyponatremia

C. Osmotic diuresis from hyperglycemia causing polyuria with a relatively concentrated solute-rich urine

D. Central diabetes insipidus due to impaired antidiuretic hormone secretion

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

A. Deletion 22q11.2 syndrome with conotruncal heart disease and hypocalcemia

B. Monosomy X Turner syndrome with lymphedema and left-sided heart disease

C. Trisomy 21 with neonatal hypotonia and an atrioventricular septal defect

D. Trisomy 18 with growth restriction, overlapping fingers, and rocker-bottom feet

156. A neonate has generalized erythroderma, chronic diarrhea, lymphadenopathy, eosinophilia, and very high IgE in the setting of profound T-cell dysfunction. Which syndrome should be considered?

A. Physiologic neonatal immune immaturity causing isolated low complement levels

B. Transient neonatal pustular melanosis with benign self-limited skin findings

C. Hereditary spherocytosis causing hemolysis without cellular immune dysfunction

D. Omenn syndrome, an atypical form of severe combined immunodeficiency

157. A cyanotic newborn has a single loud second heart sound and minimal response to supplemental oxygen. Echocardiography shows transposition of the great arteries with a restrictive atrial communication. What is the most urgent intervention after starting prostaglandin E1?

A. Perform balloon atrial septostomy to improve mixing between the circulations

B. Delay intervention until pulmonary vascular resistance falls naturally over several days

C. Close the ductus arteriosus pharmacologically to reduce recirculation through the lungs

D. Start indomethacin to decrease pulmonary blood flow before definitive repair

158. A newborn screen shows markedly elevated TSH and low thyroxine in an asymptomatic 5-day-old infant. What is the most important next step?

A. Confirm promptly and start levothyroxine without delaying treatment for prolonged diagnostic workup

B. Wait several months because congenital hypothyroidism usually resolves spontaneously

C. Start propylthiouracil to reduce thyroid hormone production while awaiting imaging

D. Provide iodine restriction only because exogenous thyroid hormone impairs brain development

159. A very preterm infant's ROP progresses to treatment-requiring disease. Which therapeutic principle is commonly used to prevent retinal detachment?

A. Raise oxygen saturation targets to suppress retinal hypoxia-driven angiogenic signaling

B. Ablate or suppress pathologic retinal angiogenic drive using laser therapy or selected anti-VEGF treatment

C. Treat with systemic corticosteroids because the proliferative retinopathy is primarily inflammatory

D. Use cryotherapy only after advanced retinal detachment has already developed

160. A term infant has a soft systolic ejection murmur radiating to both axillae and the back. The infant is otherwise well, with normal pulses, oxygen saturation, and feeding. Which diagnosis is most likely?

A. Severe pulmonary valve atresia causing marked cyanosis and absent antegrade pulmonary flow

B. Critical coarctation of the aorta causing ductal-dependent systemic perfusion

C. Physiologic peripheral pulmonary artery stenosis of early infancy

D. Large ventricular septal defect causing pulmonary overcirculation and congestive symptoms

161. A large-for-gestational-age infant has macroglossia, omphalocele, lateralized overgrowth, and recurrent hypoglycemia. Which syndrome is most likely?

A. Beckwith-Wiedemann syndrome with dysregulated imprinting at 11p15

B. Prader-Willi syndrome with neonatal hypotonia and later hyperphagia

C. Cri-du-chat syndrome with a high-pitched cry and microcephaly

D. Turner syndrome with lymphatic edema and short stature

162. A former 28-week infant has a cranial ultrasound several weeks after birth showing bilateral periventricular cystic lesions. Which long-term motor outcome is particularly associated with this injury?

A. Pure cerebellar ataxia without corticospinal motor involvement

B. Isolated flaccid paralysis from anterior horn cell degeneration

C. Progressive muscular dystrophy caused by dystrophin deficiency

D. Spastic diplegic cerebral palsy from periventricular white-matter injury

163. A 6-day-old newborn becomes lethargic and develops poor feeding, dystonia, and a sweet maple-syrup odor of the urine. Which metabolic defect is most likely?

A. Deficiency of branched-chain alpha-ketoacid dehydrogenase causing maple syrup urine disease

B. Deficiency of phenylalanine hydroxylase causing classic phenylketonuria

C. Deficiency of medium-chain acyl-CoA dehydrogenase causing a fatty-acid oxidation disorder

D. Deficiency of galactose-1-phosphate uridyltransferase causing galactosemia

164. A neonate has multiple congenital anomalies involving the heart, kidneys, and craniofacial structures, but no single syndrome is obvious on examination. Which genomic test is commonly used as a first-tier evaluation for pathogenic copy-number changes?

A. Conventional karyotyping to identify large aneuploidies and major chromosome rearrangements

B. Trio exome sequencing to search broadly for pathogenic single-gene variants

C. Chromosomal microarray analysis to detect clinically important genomic copy-number changes

D. A targeted multigene panel selected from the infant's dominant anomaly pattern

165. A newborn has a sharply demarcated area of absent skin on the vertex with a thin translucent membrane and no surrounding inflammation. What diagnosis is most likely?

A. Subgaleal hemorrhage causing a fluctuant scalp mass that crosses suture lines

B. Aplasia cutis congenita presenting as a sharply demarcated congenital absence of scalp skin

C. Caput succedaneum causing diffuse edematous scalp swelling after labor

D. Cephalohematoma causing subperiosteal blood collection limited by sutures

166. A term infant is visibly jaundiced at 10 hours of age. Which interpretation is most appropriate?

A. Early physiologic jaundice from increased bilirubin production and immature hepatic conjugation

B. Jaundice in the first 24 hours is pathologic until proven otherwise and warrants prompt bilirubin evaluation

C. Breastfeeding-associated jaundice from suboptimal intake during establishment of milk supply

D. Immune hemolysis only if the direct antiglobulin test is positive on the first sample

167. A term infant has focal seizures on day 2. MRI shows a unilateral middle cerebral artery territory infarction. Pregnancy and delivery were otherwise unremarkable. Which diagnosis is most likely?

A. Perinatal arterial ischemic stroke producing focal neonatal seizures and a unilateral arterial-territory infarct

B. Benign neonatal sleep myoclonus, which produces no focal cerebral infarction

C. Diffuse hypoxic-ischemic encephalopathy, which typically produces symmetric global injury rather than a vascular territory lesion

D. Periventricular leukomalacia, which primarily affects preterm periventricular white matter bilaterally

168. A very preterm infant has required parenteral nutrition for several weeks because of severe intestinal disease. Direct bilirubin and gamma-glutamyl transferase are rising. Which complication is most likely?

A. Biliary atresia causing extrahepatic obstruction and progressive conjugated hyperbilirubinemia

B. Parenteral nutrition-associated cholestasis, especially with prolonged intestinal failure

C. Congenital infection producing hepatitis, cholestasis, and additional systemic findings

D. An inherited metabolic or genetic cholestatic disorder causing impaired bile formation or transport

169. A 27-week infant has refractory hypotension despite adequate ventilation and careful volume assessment. Echocardiography shows low systemic blood flow without a large ductal shunt. Which approach best guides further treatment?

A. Give another crystalloid bolus based on the low blood pressure despite an adequate preload assessment

B. Start dopamine empirically because blood pressure alone establishes the mechanism of neonatal shock

C. Use targeted hemodynamic assessment to distinguish low preload, myocardial dysfunction, and abnormal vascular tone

D. Give hydrocortisone for presumed adrenal insufficiency without first reassessing cardiac output and tone

170. A fetus has persistent bradycardia at 55 beats/min and complete atrioventricular dissociation on fetal echocardiography. The mother is asymptomatic but has positive anti-Ro/SSA antibodies. Which neonatal complication is most strongly associated with this maternal antibody?

A. Supraventricular tachycardia from an accessory atrioventricular conduction pathway

B. Congenital complete heart block from antibody-mediated injury to the conduction system

C. Hypertrophic cardiomyopathy from fetal hyperinsulinemia and septal overgrowth

D. Ductal constriction from fetal exposure to prostaglandin-inhibiting medication

171. A very preterm infant cannot receive enough mother's milk. Which substitute is generally preferred when the goal is to preserve a human-milk diet and reduce NEC risk?

A. Unmodified cow's milk because its protein concentration matches preterm requirements

B. Sterile water as the primary enteral fluid until maternal milk supply increases

C. Pasteurized donor human milk with appropriate fortification

D. Homemade formula prepared at bedside to increase caloric density

172. A newborn has a persistent unilateral white pupillary reflex on routine examination. What is the most appropriate interpretation?

A. Leukocoria is abnormal and requires urgent ophthalmologic evaluation

B. A white reflex is physiologic in newborns because the retina is incompletely vascularized

C. The finding confirms retinopathy of prematurity in any gestational age

D. No evaluation is needed unless visual behavior is abnormal after one year

173. A 4-day-old infant suddenly develops severe bilious vomiting, abdominal pain, and bloody stool. The abdomen becomes distended and the infant is shocked. Which diagnosis requires immediate surgical evaluation?

A. Physiologic stooling delay in a breastfed infant without intestinal obstruction

B. Midgut volvulus due to intestinal malrotation causing acute bilious emesis and intestinal ischemia

C. Uncomplicated gastroesophageal reflux causing painless nonbilious regurgitation

D. Hypertrophic pyloric stenosis causing progressive nonbilious vomiting at several weeks of age

174. A 4-day-old infant develops jaundice, vomiting, hepatomegaly, hypoglycemia, and Escherichia coli sepsis shortly after milk feeding is established. Which inborn error is most likely?

A. Phenylketonuria causing acute liver failure and gram-negative sepsis in the first days

B. Medium-chain acyl-CoA dehydrogenase deficiency triggered specifically by galactose intake

C. Classic galactosemia due to galactose-1-phosphate uridyltransferase deficiency

D. Maple syrup urine disease causing isolated hyperbilirubinemia after lactose exposure

175. A newborn remains pale, weakly perfused, and bradycardic despite effective ventilation, chest compressions, and epinephrine. There was a placental laceration with substantial blood loss at delivery. What additional intervention is most appropriate?

A. Stop resuscitation because persistent pallor excludes potentially reversible hypovolemia

B. Administer loop diuretic therapy to reduce pulmonary vascular resistance immediately

C. Restrict intravascular volume because neonatal myocardium cannot tolerate preload

D. Give intravascular volume expansion, using blood when acute blood loss is substantial

176. A newborn has multiple fractures, blue sclerae, and generalized osteopenia. Family history reveals an affected parent with recurrent fractures. Which disorder is most likely?

A. Hypophosphatasia caused by isolated neonatal vitamin D deficiency

B. Spinal muscular atrophy caused by loss of anterior horn cells

C. Achondroplasia caused by constitutive FGFR3 signaling

D. Osteogenesis imperfecta caused by abnormal type I collagen

177. A former 26-week infant at 6 weeks of age has pallor, tachycardia, low hemoglobin, low reticulocyte count, and no evidence of hemolysis or blood loss. Which diagnosis is most likely?

A. Vitamin K deficiency causing isolated low hemoglobin without bleeding or coagulation abnormalities

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

C. Autoimmune hemolytic anemia causing brisk reticulocytosis and a positive antiglobulin test

D. Polycythemia from chronic fetal hypoxia causing increased rather than decreased hemoglobin

178. A term infant has persistent upper-airway obstruction that improves when crying. A feeding tube cannot be passed through either nostril into the pharynx. Which anomaly is most likely?

A. Unilateral vocal cord paralysis causing isolated inspiratory stridor with patent nasal passages

B. Tracheoesophageal fistula causing coughing with feeds but no fixed nasal obstruction

C. Laryngomalacia causing dynamic supraglottic collapse but normal catheter passage through the nares

D. Bilateral choanal atresia causing obstruction in an obligate nasal breather

179. A male newborn has prolonged bleeding after circumcision. Platelet count and PT are normal, but aPTT is markedly prolonged. An uncle had similar bleeding. Which disorder is most likely?

A. Disseminated intravascular coagulation causing low fibrinogen, thrombocytopenia, and prolonged PT and aPTT

B. Hemophilia A due to factor VIII deficiency causing procedural bleeding with an isolated prolonged aPTT

C. Vitamin K deficiency causing predominant PT prolongation and multiple factor deficiencies

D. Neonatal alloimmune thrombocytopenia causing severe thrombocytopenia with normal coagulation times

180. A growth-restricted infant is born after a pregnancy complicated by severe placental insufficiency. At 2 hours, the hematocrit is 70% and the infant is ruddy and lethargic. What prenatal process most likely contributed to the polycythemia?

A. Chronic fetal hypoxemia stimulating erythropoietin-driven red-cell production

B. Suppression of fetal erythropoietin by placental vascular resistance and low oxygen delivery

C. Maternal folate excess causing accelerated fetal marrow maturation without hypoxemia

D. Placental transfer of mature maternal erythrocytes into the fetal circulation throughout gestation

181. Monochorionic twins are delivered at 31 weeks. Twin A is pale and growth restricted with a hematocrit of 31%, while twin B is plethoric with a hematocrit of 68%. Which placental process best explains this discordance?

A. Selective placental transfer of maternal erythrocytes only to the larger recipient twin

B. Unbalanced transfusion through placental vascular anastomoses between the twins

C. Fetal erythropoietin suppression in the recipient caused by placental separation before delivery

D. Independent maternal-fetal hemorrhage occurring equally from both twins into the maternal circulation

182. A newborn has rhizomelic limb shortening, macrocephaly, frontal bossing, and midface hypoplasia. Which molecular mechanism most commonly causes this disorder?

A. Deficiency of lysosomal acid alpha-glucosidase causing Pompe disease

B. Deletion of dystrophin causing Duchenne muscular dystrophy

C. Activating mutation in FGFR3 causing achondroplasia

D. Loss of collagen type I synthesis causing osteogenesis imperfecta

183. A stable 27-week infant is beginning enteral feeds. Which feeding choice is most strongly associated with a lower risk of necrotizing enterocolitis compared with bovine-formula feeding?

A. Nutrient-enriched preterm formula as the initial exclusive diet when maternal milk is available

B. Mother's own milk, with donor human milk used when maternal milk is unavailable

C. Pasteurized donor human milk in preference to the mother's own milk because pasteurization lowers NEC further

D. Extensively hydrolyzed preterm formula to reduce intestinal antigen exposure during early feeding

184. A jaundiced newborn of Mediterranean ancestry has a sudden bilirubin rise after exposure to an oxidant medication. Direct antiglobulin test is negative. Which disorder should be considered?

A. Biliary atresia causing episodic intravascular hemolysis after oxidant exposure

B. Gilbert syndrome causing severe hemolytic anemia with reticulocytosis after medication

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

D. Hereditary spherocytosis caused specifically by maternal red-cell antibodies

185. A mechanically ventilated 26-week infant suddenly becomes bradycardic and severely hypoxemic. The right chest is hyperexpanded with absent breath sounds and the point of maximal cardiac impulse shifts leftward. What is the most urgent treatment?

A. Place the infant prone and observe for spontaneous resolution of the event

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

C. Immediate decompression of a suspected right tension pneumothorax

D. Administer exogenous surfactant before evaluating for an air leak

186. A premature infant treated with high-dose chloramphenicol develops abdominal distention, vomiting, gray color, hypotension, and cardiovascular collapse. Which mechanism explains this classic toxicity?

A. Direct vitamin K antagonism causing isolated bleeding without cardiovascular toxicity

B. Immune hemolysis caused by universal maternal antibodies against chloramphenicol

C. Excess renal secretion causing rapid depletion of circulating chloramphenicol and shock

D. Drug accumulation because immature hepatic glucuronidation limits chloramphenicol clearance

187. A neonate with congenital complete heart block has a ventricular rate of 45 beats/min, poor perfusion, and signs of low cardiac output. Which therapy may be required?

A. Indomethacin because ductal closure will reliably increase the ventricular escape rate

B. Adenosine because the rhythm is most likely an atrioventricular nodal reentrant tachycardia

C. Inhaled nitric oxide because pulmonary vasodilation corrects atrioventricular dissociation

D. Cardiac pacing because symptomatic profound bradycardia may not respond to medications

188. A term newborn exposed to a selective serotonin reuptake inhibitor throughout late pregnancy has transient jitteriness, irritability, tachypnea, and feeding difficulty during the first day. Which diagnosis best fits this pattern?

A. Early-onset bacterial sepsis caused specifically by serotonergic medication exposure

B. Classic neonatal abstinence syndrome caused by chronic maternal opioid exposure

C. Poor neonatal adaptation after late-gestation serotonergic medication exposure

D. Congenital myasthenia from transplacental acetylcholine-receptor antibodies

189. An infant required prolonged ventilation, intubation, chest compressions, and epinephrine in the delivery room. The heart rate and oxygenation are now stable. Which early post-resuscitation assessment is especially important?

A. Discontinue cardiorespiratory monitoring once the delivery-room heart rate normalizes

B. Avoid checking glucose because stress hyperglycemia reliably protects against brain injury

C. Monitor blood glucose closely because dysglycemia may follow significant resuscitation

D. Withhold temperature monitoring because thermal instability is expected and self-correcting

190. A preterm infant has persistent non-anion-gap metabolic acidosis, normal lactate, and poor growth. Urine pH remains inappropriately high during systemic acidosis. Which renal problem should be considered?

A. Congenital lactic acidosis causing a large anion-gap acidosis with high lactate

B. Renal tubular acidification impairment causing renal tubular acidosis

C. Respiratory alkalosis from chronic hyperventilation causing low bicarbonate without renal acid loss

D. Urea-cycle dysfunction causing isolated hyperammonemia without metabolic acidosis

191. A 29-week infant has recurrent apnea lasting 25 seconds with bradycardia and desaturation. Examination between episodes is normal and evaluation finds no infection, anemia, metabolic abnormality, or airway obstruction. Which treatment is most appropriate?

A. Caffeine therapy for apnea of prematurity after secondary causes of recurrent apnea are excluded

B. Routine opioid sedation to suppress respiratory variability during sleep

C. Indomethacin therapy because recurrent apnea is diagnostic of a patent ductus arteriosus

D. Continuous neuromuscular blockade to prevent chest wall movement during apnea

192. A term infant fails automated auditory brainstem response screening in both ears before discharge. Which next step is most appropriate?

A. Ignore the result because inpatient hearing screening has no relationship to later hearing

B. Assume permanent deafness immediately and defer confirmatory testing until school age

C. Begin systemic corticosteroids because failed screening usually reflects inflammatory auditory neuropathy

D. Arrange timely repeat or diagnostic audiologic evaluation according to the newborn hearing pathway

193. A large-for-gestational-age newborn has recurrent severe hypoglycemia requiring a very high glucose infusion rate. During hypoglycemia, insulin is detectable, beta-hydroxybutyrate is suppressed, and free fatty acids are low. Which mechanism is most likely?

A. Cortisol excess stimulating excessive ketone production during hypoglycemia

B. Inappropriate insulin secretion suppressing lipolysis and ketogenesis

C. Growth hormone excess causing marked lipolysis and hyperketonemia

D. Primary fatty-acid oxidation failure causing high ketones during fasting

194. A 3-month corrected-age infant with prior cystic periventricular leukomalacia has persistent fisting, poor spontaneous movement repertoire, and abnormal general movements. What is the best response?

A. Refer promptly for neurologic assessment and early motor intervention rather than waiting for a definitive cerebral palsy label

B. Wait until independent walking age because abnormal movements are expected after prematurity

C. Avoid physical or occupational therapy because early intervention worsens spasticity

D. Reassure the family that motor signs before two years cannot predict later disability

195. A 30-week infant has tolerated small enteral feeds for 48 hours without significant residuals, abdominal signs, or instability. What is the rationale for advancing enteral nutrition rather than maintaining prolonged minimal volumes?

A. Maintain trophic volumes longer because slower advancement is expected to reduce NEC in a stable infant

B. Advancing feeds supports gut adaptation and growth while reducing dependence on parenteral nutrition

C. Advance only after gastric residuals are consistently absent because residual volume predicts bowel injury

D. Replace parenteral nutrition rapidly even if the infant has not demonstrated progressive enteral tolerance

196. A 34-week infant has differential cyanosis with lower-extremity oxygen saturation lower than right-hand saturation. Echocardiography shows right-to-left shunting across the ductus and foramen ovale with no structural defect. Which physiologic change would be expected to improve oxygenation?

A. Increase pulmonary vascular resistance through lower lung volume and greater hypoxic vasoconstriction

B. Lower systemic vascular resistance so ductal blood preferentially enters the pulmonary circulation

C. A fall in pulmonary vascular resistance that reduces right-to-left extrapulmonary shunting

D. Increase intrathoracic pressure to reduce pulmonary blood flow and limit right-sided volume loading

197. A term infant had placental abruption, severe metabolic acidosis, and prolonged resuscitation. At 4 hours the infant is lethargic with hypotonia, weak suck, and recurrent seizures. Which diagnosis is most likely?

A. Benign neonatal sleep myoclonus occurring only during sleep in an otherwise normal infant

B. Isolated peripheral facial nerve palsy causing global encephalopathy and seizures

C. Hypoxic-ischemic encephalopathy after a major perinatal hypoxic event with evolving seizures and depression

D. Transient tachypnea of the newborn causing diffuse neurologic depression

198. A newborn has excessive oral secretions, coughing and cyanosis with feeding, and inability to pass an orogastric tube into the stomach. Radiograph shows the tube coiled in the upper esophagus and gas in the abdomen. Which anomaly is most likely?

A. Isolated H-type tracheoesophageal fistula with a completely patent esophagus

B. Pure esophageal atresia without a fistula, which usually leaves the abdomen gasless

C. Hypertrophic pyloric stenosis causing distal gastric outlet obstruction

D. Esophageal atresia with a distal tracheoesophageal fistula

199. A 26-week infant's axillary temperature is 35.2 C after transport from delivery to the NICU. Which response is most appropriate?

A. Delay rewarming because neonatal hypothermia lowers metabolic demand without meaningful adverse effects

B. Rewarm in a controlled thermal environment while monitoring temperature, glucose, oxygenation, and perfusion

C. Use direct unregulated heating until the temperature exceeds 38 C to create a safety margin

D. Leave the infant uncovered because spontaneous shivering will restore temperature rapidly

200. A mother develops varicella rash 2 days before delivery. The newborn is initially well. Why is this timing especially concerning?

A. Varicella cannot cross the placenta, so only postnatal respiratory transmission is relevant

B. The infant may receive a high viral inoculum before enough protective maternal antibody has crossed the placenta

C. Maternal antibody transfer is maximal during the first 24 hours after maternal rash onset

D. Neonatal varicella is prevented completely by immediate routine live varicella vaccination at birth

Answers

1. A. Gastroschisis is usually a right paraumbilical full-thickness defect with exposed bowel and no covering membrane. Initial care includes protecting the bowel, preventing heat and fluid loss, gastric decompression, and surgical management. Omphalocele is midline and membrane covered.

2. C. Fetal hemoglobin is abundant at birth and interferes with HbS polymerization, so clinical manifestations of sickle cell disease usually emerge as HbF declines over the first months of life. The infant's own red cells already contain globin proteins; maternal erythrocytes do not provide long-term hematopoiesis.

3. A. Apnea of prematurity is a diagnosis of exclusion when new or worsening events occur. Temperature instability, abdominal distention, and lethargy should prompt evaluation for infection, NEC, metabolic disturbance, anemia, or other illness. Escalating stimulant therapy without addressing a new underlying cause can delay diagnosis.

4. C. Noonan syndrome can resemble Turner syndrome phenotypically but occurs in males and females and commonly includes pulmonary valve stenosis or hypertrophic cardiomyopathy. A normal male karyotype excludes classic Turner syndrome. The other listed syndromes have different growth and anomaly patterns.

5. B. Acholic stools, dark urine, hepatomegaly, and conjugated hyperbilirubinemia raise major concern for biliary atresia. Timely diagnosis matters because the effectiveness of Kasai portoenterostomy generally declines with delayed intervention. Unconjugated jaundice syndromes do not cause pale stools and direct hyperbilirubinemia.

6. D. Neonatal lupus results from transplacental anti-Ro/SSA or anti-La/SSB antibodies and may cause rash, cytopenias, hepatic abnormalities, and congenital heart block. The skin and hematologic manifestations usually resolve as maternal antibodies clear, whereas heart block may be permanent. The alternative immune disorders do not characteristically cause this maternal-antibody pattern.

7. A. Posterior urethral valves are a major cause of lower urinary tract obstruction in male fetuses and can cause megacystis, hydronephrosis, oligohydramnios, and renal dysplasia. Bilateral renal agenesis produces an absent or poorly visualized bladder rather than a distended thick-walled bladder. Unilateral cystic disease does not explain the keyhole sign.

8. B. Prematurity, early respiratory distress, low lung volumes, and a diffuse granular pattern with air bronchograms are classic for surfactant-deficient respiratory distress syndrome. TTN usually occurs in more mature infants with prominent vascular/interstitial markings and fluid in fissures. Meconium aspiration and lobar overinflation produce hyperinflation rather than low-volume granular lungs.

9. A. Sinus tachycardia usually varies with activity and retains a normal P-QRS relationship. SVT is often very regular with abrupt onset and offset and commonly exceeds 220 to 240 beats/min in infants. Complete heart block and ventricular tachycardia have different electrocardiographic relationships.

10. B. Laryngomalacia is the most common cause of congenital stridor and results from dynamic inspiratory collapse of supraglottic structures. Symptoms often vary with position and feeding. Choanal atresia, subglottic stenosis, and TE fistula produce different anatomic and symptom patterns.

11. C. Right-to-left shunting allows venous blood to bypass ventilated alveoli, limiting the improvement in arterial oxygen tension despite high inspired oxygen. Pure hypoventilation generally responds to increased inspired oxygen, and anemia lowers oxygen content more than PaO2. The echocardiographic shunt directly identifies the mechanism.

12. C. Open myelomeningocele requires protection from trauma, drying, and infection, usually with sterile moist nonadherent coverage and prone positioning while neurosurgical plans proceed. Associated hydrocephalus, Chiari II malformation, orthopedic problems, and neurogenic bladder require coordinated evaluation. Direct pressure and harsh antiseptics can injure neural tissue.

13. B. For a nonvigorous infant with meconium-stained fluid, effective ventilation should not be delayed for routine tracheal suctioning. Airway suctioning is reserved for suspected obstruction that prevents effective ventilation. The infant's apnea and bradycardia make prompt ventilation the priority.

14. A. Ethical patient care includes transparent disclosure of significant errors, including what happened, known or possible consequences, and steps taken to treat the patient and reduce recurrence. Lack of permanent harm does not erase the importance of honest communication. Documentation should remain accurate.

15. C. Ampicillin plus gentamicin has traditionally provided empiric coverage for GBS, E coli, Listeria, and other common early-onset organisms, though local epidemiology and clinical circumstances matter. Cultures should be obtained promptly when feasible, and therapy narrowed or stopped according to results and clinical course. The single-agent alternatives leave major pathogens uncovered.

16. C. Surfactant can rapidly improve lung compliance and oxygenation. If ventilator pressures or delivered volumes are not promptly adjusted, the now more compliant lung can be overdistended and exposed to volutrauma. Close monitoring and timely weaning are therefore important after treatment.

17. B. Conotruncal heart disease, hypocalcemia from parathyroid hypoplasia, and thymic hypoplasia are classic for 22q11.2 deletion syndrome. Paternal 15q11-q13 deletion causes Prader-Willi syndrome, beta-globin variants cause hemoglobinopathies, and FGFR3 variants cause achondroplasia.

18. A. Complete atrioventricular septal defect is strongly associated with trisomy 21 and includes a common AV valve plus primum atrial and inlet ventricular septal defects. The echocardiographic anatomy distinguishes it from isolated VSD, tetralogy of Fallot, and anomalous pulmonary venous return.

19. C. Neonatal lymphedema, webbed neck, and left-sided cardiac lesions such as coarctation are classic for Turner syndrome. Noonan syndrome can resemble Turner phenotypically but occurs in both sexes and has different genetic causes. Trisomy 21 and Beckwith-Wiedemann syndrome have distinct features.

20. D. PaCO2 is inversely related to effective alveolar ventilation. When severe respiratory acidosis is due to inadequate ventilation, increasing effective minute ventilation by adjusting rate or tidal delivery lowers PaCO2, while avoiding injurious volumes and pressures. Increasing oxygen alone does not correct hypercapnia.

21. B. Rapid transition to anabolic metabolism can drive phosphorus, potassium, and magnesium into cells, particularly in growth-restricted or nutritionally depleted infants. This refeeding-like physiology can be clinically important during aggressive parenteral nutrition. Management requires monitoring and appropriate nutrient/electrolyte adjustment rather than simply stopping nutrition.

22. D. At the limits of viability, ethically appropriate counseling acknowledges uncertainty, explains expected burdens and outcomes, and incorporates family goals within the range of medically reasonable care. Shared decision-making does not mean abandoning families to decide alone. Repeated conversations are often needed as pregnancy and clinical circumstances evolve.

23. D. Newborns depend heavily on brown-fat nonshivering thermogenesis, which consumes oxygen and glucose. Cold stress therefore can worsen hypoglycemia, hypoxemia, and metabolic acidosis, particularly in preterm infants with limited reserves. Preventing evaporative, conductive, convective, and radiant heat loss is central to stabilization.

24. C. To establish congenital CMV, viral detection should occur within the first 3 weeks of life; saliva PCR is sensitive and urine confirmation is commonly used because saliva can be contaminated by breast milk. Testing later cannot reliably distinguish congenital from postnatal acquisition. Maternal IgG transfer also limits interpretation of neonatal serology.

25. A. Meconium aspiration causes a mixture of airway obstruction, uneven air trapping, atelectasis, inflammation, surfactant dysfunction, and sometimes PPHN. This produces patchy infiltrates with hyperinflation. Pure surfactant deficiency generally causes low lung volumes and diffuse granular disease, not the classic meconium pattern.

26. A. Meconium ileus is strongly associated with cystic fibrosis and results from inspissated meconium obstructing the distal small intestine; a microcolon may be seen because the colon was unused in utero. Hirschsprung disease can cause delayed meconium passage but has a different anatomic mechanism and contrast pattern.

27. C. Standardized developmental tests such as the Bayley scales are commonly used in high-risk infant follow-up to quantify developmental performance across multiple domains. Scores require clinical context and are not equivalent to a definitive long-term diagnosis. They are different from resuscitation or gestational-age scoring systems.

28. A. In Pierre Robin sequence, micrognathia leads to glossoptosis and variable upper-airway obstruction. Prone or side positioning can improve airway patency in selected monitored infants, while

severe cases may require nasopharyngeal airway or surgical strategies. Feeding and aspiration risk must be assessed alongside breathing.

29. D. Pulmonary hemorrhage in very preterm infants is multifactorial and can be associated with a hemodynamically significant PDA and pulmonary overcirculation. The acute presentation requires stabilization and evaluation of contributing factors. Lesions that markedly reduce pulmonary blood flow do not fit the same pathophysiologic association.

30. C. Group B Streptococcus remains an important cause of early-onset neonatal sepsis, particularly with maternal colonization and inadequate intrapartum prophylaxis. Symptoms can include respiratory distress, shock, and bacteremia or meningitis. The alternative organisms are not typical causes of this exposure-linked early-onset syndrome.

31. D. A large VSD often becomes clinically apparent over the first weeks as pulmonary vascular resistance falls, leading to pulmonary overcirculation and heart failure symptoms. Small ASDs and a patent foramen ovale do not usually cause severe early heart failure. Benign peripheral pulmonary stenosis does not produce this congestive picture.

32. A. Post-discharge nutrition for preterm infants should reflect gestational age, current growth, feeding type, mineral status, and medical complexity. Some infants need continued fortification or enriched formula while others can transition sooner. Adult cow's milk and free-water supplementation are not appropriate neonatal feeding strategies.

33. C. Human milk contains secretory IgA along with lactoferrin, oligosaccharides, leukocytes, and other bioactive factors that support mucosal defense. Secretory IgA can bind pathogens at epithelial surfaces without requiring systemic absorption. Maternal immune cells and complement do not permanently replace the infant's immune system.

34. D. Volutrauma, barotrauma, oxygen toxicity, and inflammation contribute to neonatal lung injury. Lung-protective strategies aim to provide adequate gas exchange with the lowest effective pressure, volume, and oxygen exposure. Severe hypocapnia and unnecessary hyperoxia carry neurologic and pulmonary risks rather than preventing BPD.

35. A. Twin anemia-polycythemia sequence results from chronic net transfusion across very small placental anastomoses, producing anemia in the donor and polycythemia in the recipient, sometimes without classic amniotic-fluid discordance. The striking paired hematologic difference is not physiologic anemia or a platelet-antibody disorder.

36. C. A term or near-term infant with evidence of significant intrapartum hypoxia-ischemia and moderate-to-severe encephalopathy should be evaluated promptly for therapeutic hypothermia, which is time sensitive and typically begun within 6 hours. MRI is useful for prognosis but should not delay eligible treatment. Hyperthermia is harmful, and corticosteroids are not standard neuroprotective therapy for HIE.

37. C. T-cell receptor excision circle screening identifies impaired production of naive T cells and can detect severe combined immunodeficiency before infections appear. Prompt immunologic evaluation and infection precautions are important because live vaccines and certain blood products may be hazardous. CGD affects phagocyte oxidative killing rather than T-cell output.

38. B. A significant preductal-postductal saturation difference in a hypoxemic term infant suggests right-to-left shunting across the ductus from elevated pulmonary vascular resistance, characteristic of PPHN. A VSD does not create this differential cyanosis pattern. Coarctation and SVT do not by themselves explain the classic saturation gradient.

39. A. Current neonatal resuscitation guidance supports starting term and late-preterm infants who need respiratory support with 21% oxygen and titrating to target saturations. Routine 100% oxygen can produce unnecessary hyperoxia. Pulse oximetry, not visual assessment of color alone, guides oxygen adjustment.

40. C. Omphalocele is a midline defect at the umbilical ring with herniated abdominal contents covered by a sac and is often associated with chromosomal or other major anomalies. Gastroschisis is usually right-sided and uncovered. The distinction affects delivery-room handling and associated-anomaly evaluation.

41. D. Trophic or minimal enteral feeds provide small amounts of substrate to stimulate gut maturation and motility while full nutritional support is developed. They are not intended to meet the entire caloric requirement initially and do not eliminate NEC risk. Prolonged bowel inactivity is not the physiologic goal.

42. A. For most term and preterm infants who do not need immediate resuscitation, current guidance supports deferred cord clamping for at least 60 seconds. Immediate clamping is not routinely required solely because of prematurity. Cord milking is not a universal substitute and has gestation-specific considerations.

43. D. Aminoglycosides are eliminated primarily by the kidneys, and glomerular filtration and tubular handling are immature in preterm neonates. Dosing therefore incorporates gestational/postnatal age, renal function, and therapeutic drug monitoring. Longer intervals help achieve efficacy while limiting accumulation and toxicity.

44. D. Inhaled nitric oxide selectively dilates ventilated pulmonary vascular beds and is a standard therapy for appropriate term or near-term infants with hypoxemic respiratory failure and PPHN. Carbon dioxide and prostaglandin inhibition do not provide selective pulmonary vasodilation. Systemic vasopressors may support blood pressure but do not substitute for targeted pulmonary vasodilation.

45. C. Pierre Robin sequence consists of micrognathia leading to glossoptosis and often a characteristic cleft palate, with variable airway and feeding difficulty. It can occur in isolation or as part of another

syndrome. Choanal atresia is a nasal obstruction, and macroglossia is the opposite tongue-mandible relationship.

46. D. Clenched overlapping fingers, rocker-bottom feet, severe growth restriction, and congenital heart disease are characteristic of trisomy 18. Trisomy 13 more often includes midline central nervous system and facial defects with polydactyly. The other syndromes have different defining neonatal phenotypes.

47. C. Organic acidemias commonly present with anion-gap metabolic acidosis, ketosis, hyperammonemia, and encephalopathy after feeds begin. Urea-cycle disorders generally lack a major metabolic acidosis early. The acid-base pattern is therefore a useful discriminator in a sick neonate with hyperammonemia.

48. A. BPD-associated pulmonary hypertension requires attention to the underlying lung disease, adequate oxygenation, nutrition, cardiac anatomy, and pulmonary vascular therapy when appropriate. Chronic hypoxemia worsens pulmonary vasoconstriction, and growth is beneficial rather than harmful. Management should be physiologically targeted, not based on automatic closure of every shunt.

49. A. Prader-Willi syndrome results from loss of paternal gene expression in 15q11-q13 and often presents in the newborn period with severe hypotonia and feeding difficulty. Angelman syndrome involves loss of maternal expression and usually has a different later neurodevelopmental phenotype. SMA is an important hypotonia differential but has a different molecular basis.

50. C. Serum creatinine crosses the placenta, so the newborn's early value initially reflects maternal concentration before declining according to the infant's renal function and gestation. Trends, urine output, and clinical context are more informative than a single day-1 value. Immediate labeling as intrinsic or obstructive renal failure is therefore inappropriate.

51. D. Urea-cycle defects often present after protein feeding with severe hyperammonemia, encephalopathy, and respiratory alkalosis from central stimulation. Organic acidemias more often produce anion-gap metabolic acidosis and ketosis. Severe hyperammonemia is a medical emergency requiring rapid reduction of nitrogen burden.

52. A. Choanal atresia is a congenital obstruction of the posterior nasal passage and may be unilateral or bilateral. Bilateral disease can be a neonatal emergency; unilateral disease may present more subtly with persistent unilateral obstruction or discharge. Passage of a nasal catheter helps localize the obstruction.

53. C. Conotruncal cardiac defects plus hypocalcemia strongly suggest 22q11.2 deletion syndrome, reflecting abnormal pharyngeal pouch and arch development. Williams syndrome is classically associated with supravalvar aortic stenosis. Prader-Willi and cri-du-chat have different genetic mechanisms and phenotypes.

54. A. A large PDA in a very preterm infant can produce pulmonary overcirculation and systemic diastolic runoff, causing bounding pulses, widened pulse pressure, and respiratory deterioration. Critical

pulmonary stenosis and tetralogy reduce pulmonary blood flow, while PPHN typically involves elevated pulmonary resistance and right-to-left shunting rather than this hyperdynamic runoff pattern.

55. D. Aminoglycoside pharmacokinetics vary substantially with gestational age, postnatal age, fluid status, and renal function. Appropriately timed peak/trough or model-based concentrations can guide dosing and reduce nephrotoxic or ototoxic accumulation. A random level without timing context is difficult to interpret.

56. B. A passed newborn hearing screen does not exclude delayed or progressive hearing loss in infants with important risk factors such as meningitis or certain ototoxic exposures. Ongoing developmental and audiologic surveillance is appropriate. Early detection supports timely amplification, communication intervention, and language development.

57. D. Preterm kidneys have limited tubular sodium reabsorptive capacity, and ongoing urinary sodium losses can contribute to hyponatremia and poor growth. This differs from SIADH, in which water retention is central, and from diabetes insipidus, which produces hypernatremia. Nutritional sodium needs may therefore exceed those of term infants.

58. C. Severe thrombocytopenia in an otherwise well newborn with a normal maternal platelet count and an affected sibling strongly suggests neonatal alloimmune thrombocytopenia. Maternal antibodies target a paternally inherited fetal platelet antigen and can cause antenatal intracranial hemorrhage. Maternal ITP usually involves maternal thrombocytopenia and tends to produce less severe neonatal disease.

59. B. Subgaleal hemorrhage occurs in the potential space beneath the galea and can spread extensively across the scalp, allowing life-threatening blood loss and shock. Cephalohematoma is subperiosteal and limited by sutures, while caput is edema in a more superficial tissue plane. Prompt recognition requires serial head measurements and hemodynamic monitoring.

60. C. Extremely preterm infants can develop nonoliguric hyperkalemia early after birth because of immature Na-K-ATPase activity, cellular potassium shifts, and limited renal excretion. Preserved urine output does not exclude dangerous hyperkalemia. ECG changes require urgent treatment while confirming a nonhemolyzed sample.

61. A. A prospective cohort study classifies participants by exposure before the outcome develops and follows them forward to measure incidence. This design can estimate risk and relative risk directly. Without randomization, however, differences between exposed and unexposed groups can confound associations.

62. B. Spinal muscular atrophy results from loss of SMN1 function and degeneration of anterior horn cells. Newborn screening permits presymptomatic diagnosis, which is important because disease-modifying therapy is most effective when started early. Duchenne, myotonic dystrophy, and Pompe disease have different molecular causes.

63. C. The classic waiter's-tip posture with preserved grasp indicates upper brachial plexus injury, usually involving C5-C6. Lower plexus lesions affect hand function more prominently. Many birth-related upper plexus injuries improve, but serial examination and early therapy are important.

64. A. Trisomy 13 is strongly associated with severe midline developmental anomalies, including holoprosencephaly, orofacial clefts, microphthalmia, and polydactyly. Trisomy 18 has a different limb pattern, while trisomy 21 and Turner syndrome do not typically produce this combination.

65. D. Obstructed TAPVR causes pulmonary venous hypertension, pulmonary edema, and severe cyanosis and is a surgical emergency. Unobstructed anomalous return may present less dramatically. Ductal runoff and pulmonary stenosis do not match the combination of pulmonary edema and anomalous pulmonary venous anatomy.

66. B. Congenital diaphragmatic hernia allows abdominal viscera into the thorax and is associated with pulmonary hypoplasia and PPHN. Bag-mask ventilation can distend intrathoracic bowel and worsen lung compression, so tracheal intubation and gastric decompression are preferred. The findings do not indicate a pleural air leak requiring thoracentesis.

67. A. Loop diuretics increase urinary calcium and electrolyte losses and can contribute to nephrocalcinosis, ototoxicity, and disturbances of sodium and potassium balance. They do not cause renal calcium retention or directly close the ductus through cyclooxygenase inhibition. Ongoing use therefore requires careful reassessment and monitoring.

68. D. ROP is a vasoproliferative retinal disorder of prematurity associated with incomplete vascularization and postnatal oxygen and growth factors. Screening is based on gestational age, birth weight, and clinical course. Timely treatment of severe disease can reduce retinal detachment and blindness.

69. A. Extremely preterm infants have compliant chest walls, immature respiratory muscles, and limited respiratory reserve, all of which increase work of breathing and predispose to fatigue. Their chest wall is not rigid, and respiratory musculature is not more mature than that of term infants. These developmental limitations complicate liberation from support.

70. A. Delayed meconium passage, distal obstruction, and explosive stool after rectal examination suggest Hirschsprung disease from absence of enteric ganglion cells. Diagnosis is confirmed by rectal biopsy. Meconium ileus, pyloric stenosis, and duodenal atresia have different locations and characteristic presentations.

71. C. Transient abnormal myelopoiesis is uniquely associated with trisomy 21 and GATA1-mutated megakaryoblastic proliferation. It may resolve spontaneously but can cause serious neonatal complications and is associated with later risk of myeloid leukemia of Down syndrome. Follow-up is therefore required even after apparent resolution.

72. C. Congenital rubella is classically associated with cataracts, congenital heart disease such as PDA or pulmonary artery stenosis, and sensorineural hearing impairment. The other congenital infections have different characteristic ocular, neurologic, or skeletal patterns.

73. B. A P value is a probability of observing data at least as extreme as those obtained, assuming the null hypothesis and statistical model are correct. It is not the probability that the null hypothesis is true and does not measure clinical importance. Effect size and confidence intervals provide additional information.

74. D. Magnesium crosses the placenta and, at elevated neonatal levels, can depress neuromuscular transmission and respiratory drive. The typical picture is hypotonia, weak reflexes, and respiratory depression. The other proposed mechanisms do not fit the pharmacology or characteristic examination findings.

75. C. Human milk alone does not reliably provide enough protein, calcium, phosphorus, and other nutrients for rapid growth in very preterm infants. Human milk fortification helps meet these needs while preserving the benefits of human milk. Dilution or dextrose-only therapy would worsen nutrient deficits.

76. A. When pulmonary blood flow is ductal dependent, prostaglandin E1 maintains ductal patency and can be lifesaving. Indomethacin would promote closure and worsen cyanosis. Diuresis and adenosine do not provide an alternative source of pulmonary blood flow.

77. C. Invasive candidiasis in extremely preterm infants can disseminate widely, including to the CNS, kidneys, eyes, heart, and other organs. A positive blood culture should be taken seriously and prompts systemic antifungal therapy plus evaluation for dissemination and source control. It is not appropriately dismissed as routine contamination.

78. A. Pulmonary interstitial emphysema occurs when ventilator-associated alveolar rupture allows gas to dissect along the pulmonary interstitium. Radiographs show characteristic linear or cystic lucencies. Pleural fluid, hemorrhage, and simple atelectasis do not create this branching interstitial air pattern.

79. D. Positive predictive value rises as disease prevalence increases because a positive result is more likely to represent true disease in a higher-risk population. Sensitivity and specificity are intrinsic test characteristics under the same operating conditions. Likelihood ratios are also not directly changed by prevalence.

80. A. Rapid early bilirubin rise with anemia, reticulocytosis, and a positive direct antiglobulin test indicates immune hemolysis. This can result from Rh or ABO antibodies depending on maternal-infant blood groups and antibody characteristics. Physiologic and breast milk jaundice do not produce immune coating of erythrocytes.

81. C. Extremely preterm infants benefit from coordinated high-risk follow-up that tracks motor, cognitive, language, sensory, feeding, and family outcomes. Early identification of delay allows timely

intervention during a period of high neuroplasticity. Follow-up should complement, not replace, primary care.

82. D. Essential fatty acids are required for growth, skin integrity, and cell membrane function. Prolonged fat-free nutrition can produce dermatitis, alopecia, thrombocytopenia, and growth failure. Modern neonatal parenteral nutrition therefore includes appropriately dosed lipid unless contraindicated.

83. A. A ductal-dependent systemic lesion such as critical coarctation, interrupted aortic arch, or severe left-sided obstruction can present with shock as the ductus closes. Prostaglandin E1 can restore ductal flow while definitive diagnosis and stabilization proceed. Therapies that close the ductus can be dangerous in this setting.

84. D. Sensitivity is the proportion of truly affected patients who test positive: 95 true positives divided by 100 affected infants, or 95%. Specificity would use the unaffected group. Sensitivity is independent of disease prevalence in the sampled population.

85. C. TTN reflects delayed clearance of fetal lung fluid and is associated with cesarean delivery without labor. The typical radiograph shows hyperinflation, streaky perihilar markings, and fissural fluid, with improvement over the next day or two. RDS, PIE, and diaphragmatic hernia have different radiographic and clinical patterns.

86. B. E coli is a major cause of early-onset sepsis and meningitis, particularly among preterm and very-low-birth-weight infants. Empiric treatment strategies for symptomatic early-onset sepsis therefore include coverage for common maternal-genital-tract organisms. The listed alternatives are not typical dominant pathogens in this setting.

87. D. The absolute risk reduction is 20% minus 10% = 10%, or 0.10. Number needed to treat is 1 divided by absolute risk reduction, so NNT = 10. Relative risk reduction alone cannot be used to calculate NNT without the baseline event rate.

88. C. Maternal hypertension and placental insufficiency are associated with transient neonatal neutropenia, especially in preterm or growth-restricted infants. Clinical context and severity guide evaluation for infection or marrow disease. SCID primarily affects lymphocytes, and vitamin K deficiency is a coagulation disorder.

89. B. Early congenital syphilis can cause rash involving palms and soles, hepatosplenomegaly, cytopenias, rhinitis, and characteristic long-bone abnormalities. Maternal treatment history and infant evaluation determine management. The alternate congenital infections have different hallmark findings.

90. D. Apical displacement of the tricuspid valve and atrialization of the right ventricle define Ebstein anomaly. Maternal lithium exposure has a historical association with this lesion, although absolute risk is low. The echocardiographic anatomy is distinct from AV septal defect, tetralogy, and aortic stenosis.

91. A. Phenobarbital remains a commonly recommended first-line medication for acute neonatal seizures after reversible metabolic causes are corrected, although choice may vary with etiology and evolving evidence. Continuous EEG is important because clinical observation underestimates seizure burden. Absence-seizure therapies do not match typical neonatal seizure physiology.

92. A. Loop diuretics increase urinary calcium excretion and are a recognized risk factor for nephrocalcinosis in preterm infants. The ultrasound appearance can be medullary echogenicity. Furosemide does not cause congenital obstructive lesions or inherited cystic kidney disease.

93. D. VACTERL association is recognized when several component anomalies cluster: vertebral, anal, cardiac, tracheoesophageal, renal, and limb defects. The infant described displays nearly the full association. CHARGE, Beckwith-Wiedemann, and Noonan syndromes have different defining phenotypes.

94. D. Maternal intraamniotic infection and prolonged rupture of membranes substantially increase exposure to ascending bacterial pathogens and the risk of early-onset sepsis. The comparison conditions do not create the same infectious risk. A symptomatic newborn requires prompt clinical assessment and empiric management rather than reassurance based on gestational age alone.

95. B. Maternal IgG is actively transported across the placenta, with transfer increasing markedly during the third trimester. Very preterm infants therefore miss much of this passive antibody acquisition and have lower IgG concentrations. IgM does not efficiently cross the placenta.

96. C. The classic triad of congenital toxoplasmosis includes hydrocephalus, intracranial calcifications, and chorioretinitis. CMV more often causes microcephaly and periventricular calcifications. Rubella and syphilis have distinct multisystem manifestations.

97. B. Neonatal HSV can occur even when maternal infection was unrecognized and may present as skin-eye-mouth disease, encephalitis, or disseminated infection. Vesicles plus seizures warrant immediate IV acyclovir while diagnostic studies are obtained. Delaying systemic therapy can increase morbidity and mortality.

98. B. TSH-receptor-stimulating immunoglobulins can persist in a mother with a history of Graves disease even when she is euthyroid and can cross the placenta to stimulate the fetal thyroid. Maternal thyroid hormone transfer does not usually produce this sustained biochemical pattern. Elevated neonatal TSH would be expected with primary hypothyroidism, not suppressed TSH with high free thyroxine.

99. C. A congenital pulmonary airway malformation is a developmental cystic lung lesion supplied by the pulmonary circulation and may cause respiratory distress after birth. Pulmonary sequestration usually has systemic arterial supply. PIE is acquired after ventilation, while TTN does not produce a congenital cystic mass.

100. B. Growth assessment in preterm infants should integrate longitudinal anthropometrics and actual energy/protein/mineral intake. Weight alone can be strongly influenced by fluid status. Length and head

circumference provide additional information about lean tissue and brain growth and help identify disproportionate growth failure.

101. D. Marked polycythemia increases blood viscosity and can impair tissue perfusion, contributing to neurologic, gastrointestinal, cardiorespiratory, and metabolic symptoms. Increased red-cell turnover also raises bilirubin load. The physiology is the opposite of anemia and does not reduce bilirubin production.

102. C. Chronic opioid exposure produces fetal neuroadaptation; after delivery, the drug supply falls and withdrawal can cause autonomic, gastrointestinal, and neurologic hyperactivity. Persistent intoxication would more often cause sedation and respiratory depression rather than hypertonic withdrawal. The syndrome is not explained by serotonin toxicity or an autoimmune neuropathy.

103. C. High-stakes decisions require accurate, confidential communication through a qualified medical interpreter when language barriers exist. Clinicians should address the parents directly and check understanding. Children or untrained relatives should not carry the burden of interpreting complex medical information when professional services are available.

104. D. A substantial direct bilirubin elevation is pathologic and indicates cholestasis until proven otherwise. Evaluation should consider biliary atresia, infection, metabolic disease, endocrine disorders, and parenteral nutrition exposure, among other causes. Physiologic and breast milk jaundice are primarily unconjugated.

105. A. Bronchopulmonary dysplasia is chronic lung disease of prematurity resulting from immature lung development plus inflammatory, oxygen, and ventilator injury. Persistent respiratory support near 36 weeks postmenstrual age is a common clinical context. TTN resolves rapidly, and the course described is not explained by one acute infectious episode.

106. D. Fetal renin-angiotensin blockade can impair renal perfusion and development, leading to oligohydramnios and the downstream sequence of pulmonary and skeletal consequences. Polyhydramnios is the opposite fluid abnormality. Ductal constriction is classically associated with prostaglandin inhibition rather than ACE inhibition.

107. A. Listeria can cross the placenta or be acquired around delivery and may cause fetal loss, preterm birth, sepsis, pneumonia, meningitis, and granulomatosis infantiseptica. Maternal foodborne illness can be a clue. The alternative bacteria do not characteristically produce this congenital granulomatous sepsis pattern.

108. A. Blood and inflammatory debris after severe IVH can obstruct CSF pathways or impair resorption, leading to progressive ventricular dilation. Serial head circumference and ultrasound trends guide timing of neurosurgical consultation and temporizing interventions. The imaging finding cannot be explained by benign macrocephaly or a scalp hemorrhage.

109. C. Protein is essential for lean tissue, organ, and linear growth, but it must be delivered with enough nonprotein energy to support anabolism. Weight gain driven mainly by fluid or fat does not

ensure appropriate length and head growth. Routine protein restriction can worsen growth failure in preterm infants.

110. A. Neonatal seizures are frequently electrographic-only or have subtle clinical manifestations, and many nonepileptic movements resemble seizures. Continuous video EEG provides the most reliable correlation between behavior and cortical electrical activity. It also helps quantify seizure burden and response to therapy.

111. C. ECMO requires systemic anticoagulation, making extreme prematurity and severe intracranial bleeding risk major limitations. Term or near-term infants with reversible severe respiratory failure, including PPHN or meconium aspiration, may be candidates when conventional and rescue therapies fail. Selected CDH patients may also receive ECMO depending on center practice and overall prognosis.

112. A. A neonatal UTI can be associated with structural urinary tract abnormalities, and renal/bladder ultrasonography is commonly used as an initial imaging study. Additional studies depend on ultrasound findings, organism, recurrence, and local practice. Antibiotic treatment should not be delayed solely to complete imaging.

113. D. Risk in the exposed group is 0.30 and risk in the unexposed group is 0.15. Relative risk is 0.30 divided by 0.15 = 2.0, meaning the exposed group has twice the observed risk. The calculation does not by itself establish causation.

114. B. Ventilation is the priority in neonatal resuscitation because inadequate lung inflation is the usual cause of persistent bradycardia at birth. An apneic infant with a heart rate below 100 beats/min requires positive-pressure ventilation. Chest compressions and epinephrine are reserved for persistent severe bradycardia after effective ventilation has been established.

115. D. Hemolytic disease from Rh alloimmunization is mediated by maternal IgG that crosses the placenta and targets fetal erythrocyte antigens. IgM does not efficiently cross the placenta, and the direction of clinically important antibody transfer is maternal to fetal. The positive direct antiglobulin test supports antibody-coated neonatal erythrocytes.

116. A. Cephalohematoma is a subperiosteal blood collection, so it is bounded by cranial sutures and may become evident hours after delivery. It can contribute to jaundice as blood is resorbed. Subgaleal hemorrhage crosses sutures and carries much greater acute blood-loss risk.

117. C. Contemporary preterm respiratory care favors effective noninvasive support and, when indicated, surfactant delivery with the least invasive method feasible. Thin-catheter surfactant techniques can reduce exposure to mechanical ventilation in spontaneously breathing infants. Routine prolonged ventilation or paralysis is not required simply to administer surfactant.

118. D. Transient neonatal pustular melanosis is benign and may be present at birth; fragile pustules evolve into hyperpigmented macules with a collarette of scale. Unlike erythema toxicum, lesions lack a

prominent erythematous base and may involve palms and soles. Infection should be considered when lesions or systemic findings are atypical.

119. B. Classic 21-hydroxylase deficiency causes cortisol and aldosterone deficiency with salt wasting, hyperkalemia, dehydration, and possible hypoglycemia. Affected males may lack obvious genital ambiguity and present in adrenal crisis. The alternative disorders predict opposite electrolyte patterns or lack the acute shock physiology.

120. D. A regular narrow-complex tachycardia near 250 beats/min in a neonate is typical of SVT. If the infant is stable and vagal maneuvers fail, rapid IV adenosine is appropriate because it transiently blocks AV nodal conduction. Cardioversion is used for hemodynamic instability, while atropine and prostaglandin do not treat reentrant SVT.

121. A. Very preterm infants miss much of third-trimester calcium and phosphorus accretion and are at risk for metabolic bone disease, especially with inadequate mineral intake or prolonged parenteral nutrition. Elevated alkaline phosphatase and low phosphorus support the diagnosis. Achondroplasia and osteopetrosis have different pathophysiology and imaging patterns.

122. B. For a relative risk, the null value is 1.0. Because the 95% confidence interval includes 1.0, the study does not demonstrate a statistically significant difference at the conventional two-sided 0.05 level. Lack of significance is not proof of equivalence or no effect.

123. D. Renal vein thrombosis classically presents with hematuria, thrombocytopenia, and an enlarged kidney, especially in sick, dehydrated, or catheterized neonates. Obstructive uropathy and nephrocalcinosis do not typically cause this triad. Doppler ultrasonography is useful for evaluation.

124. A. Neurodevelopmental outcome is influenced by medical risk and by access to coordinated developmental, sensory, nutritional, and psychosocial services. Care coordination can reduce fragmentation and help families engage with early intervention and specialty care. Family-centered support is part of high-quality follow-up, not separate from it.

125. D. Jitteriness is typically stimulus-sensitive and suppressible with gentle restraint and lacks the ocular or autonomic features that can accompany seizures. It may be benign or associated with hypoglycemia, hypocalcemia, drug withdrawal, or other conditions. Suspicious events that are not clearly benign should be evaluated, often with EEG.

126. C. Corrected age subtracts the degree of prematurity from chronologic age and is useful when interpreting early growth and developmental milestones in infants born preterm. Its relevance decreases as children get older. It does not replace chronologic age for immunizations or all other medical decisions.

127. B. DIC is a systemic consumptive coagulopathy associated with severe sepsis, asphyxia, shock, and other major insults. The combination of thrombocytopenia, prolonged clotting times, low fibrinogen,

and increased fibrin degradation strongly supports DIC. Hemophilia and alloimmune thrombocytopenia produce more selective laboratory abnormalities.

128. A. Chest compressions are indicated when the heart rate remains below 60 beats/min despite effective ventilation. Compressions are coordinated with ventilation rather than replacing it because ventilation remains central to neonatal resuscitation. Sodium bicarbonate is not the first-line intervention for this scenario.

129. B. Peripheral facial nerve palsy can follow birth trauma and affects the upper and lower face, including eye closure and forehead movement. Congenital absence or hypoplasia of the depressor anguli oris causes asymmetric crying but does not impair eyelid closure. Eye protection is important when closure is incomplete.

130. D. Pneumatosis intestinalis with systemic or abdominal signs is highly concerning for NEC. Enteral feeds are stopped while the infant receives bowel rest, decompression, antimicrobial therapy, and supportive care as indicated. Continuing or increasing feeds in active NEC can worsen bowel injury.

131. B. Perinatal hypoxia-ischemia can injure the myocardium and cause transient ventricular systolic dysfunction, hypotension, and biochemical evidence of myocardial injury. The global pattern after asphyxia differs from diabetic hypertrophic cardiomyopathy. A small restrictive PDA would not explain this degree of myocardial dysfunction.

132. C. Preterm infants have lower iron stores and rapid postnatal growth, and many require supplemental iron depending on feeding type, transfusion history, and local protocols. Folate and other micronutrients remain important as well. The alternatives incorrectly assume excess stores or toxicity rather than common nutritional vulnerability.

133. B. Intention-to-treat analysis preserves participants in their original randomized groups regardless of protocol adherence or crossover. This maintains the benefits of randomization and generally gives an unbiased estimate of the effect of assignment to the intervention. Per-protocol analysis instead focuses on those who adhered to the assigned strategy.

134. A. Infants of diabetic mothers are at increased risk of early hypocalcemia, and low magnesium can blunt parathyroid hormone secretion and action. Neuromuscular irritability may result. Hyperparathyroidism and vitamin D excess would cause hypercalcemia rather than the observed findings.

135. D. Caffeine has a prolonged half-life in preterm neonates because metabolic pathways and renal elimination are immature. This permits relatively infrequent maintenance dosing compared with older patients. Pharmacokinetics change with maturation, so dosing and toxicity assessment should reflect age and clinical status.

136. B. Comfort-focused care is active medical care directed at symptom relief, dignity, bonding, and family support. Analgesia, treatment of dyspnea, warmth, feeding or mouth care as appropriate, and

psychosocial or spiritual support may all be important. Palliative care can coexist with selected disease-directed treatments when they align with the family's goals.

137. C. The acronym CHARGE describes coloboma, heart disease, choanal atresia, growth/developmental issues, genital anomalies, and ear abnormalities; CHD7 variants are common. VACTERL lacks the defining coloboma and ear pattern. Pierre Robin is a sequence of mandibular and airway findings rather than this multisystem syndrome.

138. B. Gastroschisis bowel is often inflamed and dysmotile, so parenteral nutrition is commonly required while decompression and staged or primary closure are managed. Enteral feeds are introduced when bowel function returns and then advanced as tolerated. Immediate full feeds or prolonged complete starvation are inappropriate.

139. A. Late vitamin K deficiency bleeding can occur in infants who did not receive prophylaxis, particularly with exclusive breastfeeding or cholestatic disease, and intracranial hemorrhage is a feared presentation. Vitamin K-dependent factor deficiency prominently prolongs PT. Hemophilia and platelet disorders have different laboratory patterns.

140. C. Specificity is the proportion of disease-free patients who test negative: 880 divided by 900, approximately 97.8%. The false-positive rate is the complement, about 2.2%. Predictive values additionally depend on disease prevalence.

141. C. SIADH produces water retention, low serum osmolality, and urine that remains inappropriately concentrated despite hyponatremia. Central diabetes insipidus causes dilute polyuria and hypernatremia. The laboratory pattern therefore reflects excess antidiuretic effect rather than free-water loss.

142. B. Antenatal corticosteroids accelerate fetal organ maturation and reduce major preterm morbidities, including respiratory distress syndrome and intraventricular hemorrhage. Diuresis does not mature the fetal lung, indomethacin is a tocolytic rather than a surfactant-maturing therapy, and erythropoietin is not used for this purpose.

143. C. A case-control study begins with outcome status - cases with disease and controls without disease - and then assesses prior exposures. It is efficient for uncommon outcomes and commonly estimates odds ratios. Because exposure is not randomized, confounding and bias require careful consideration.

144. A. Noninvasive positive pressure can insufflate the gastrointestinal tract, producing abdominal distention that splints the diaphragm. Orogastric venting can reduce this mechanical burden while respiratory support continues. Symmetric breath sounds and lack of an air leak argue against emergency thoracentesis.

145. B. Marked weight loss, low urine output, poor milk transfer, and hypernatremia point to inadequate fluid intake with free-water deficit. Salt-wasting adrenal disease generally causes hyponatremia, while

SIADH causes dilutional hyponatremia. Management requires careful rehydration and feeding support because overly rapid sodium correction can be dangerous.

146. B. In very preterm infants, fragile germinal-matrix vessels are prone to hemorrhage. Blood confined to the germinal matrix is classified as grade I, while ventricular extension and ventricular dilation define higher grades. PVL is ischemic white-matter injury rather than germinal-matrix bleeding.

147. A. Maternal hyperglycemia drives fetal pancreatic beta-cell hyperplasia and hyperinsulinemia. After birth, placental glucose delivery stops abruptly while insulin remains high, producing early neonatal hypoglycemia. Maternal insulin does not cross the placenta in clinically important amounts, and infants of diabetic mothers generally have increased rather than reduced glycogen and fat stores.

148. C. Epidermolysis bullosa comprises inherited disorders of structural proteins that produce blistering after minor mechanical trauma. The neonatal priority is gentle handling, wound care, infection prevention, and subtype evaluation because severity varies widely. Benign neonatal rashes do not cause friction-induced erosions.

149. D. Erythema toxicum is a common benign neonatal eruption that often appears during the first several days as erythematous macules, papules, and pustules, typically sparing palms and soles. A well infant with the classic distribution needs reassurance rather than antimicrobial therapy. Vesicular HSV, candidiasis, and toxin-mediated staphylococcal disease require different clinical contexts and often more intensive evaluation.

150. D. Birth-related phrenic nerve injury can paralyze one hemidiaphragm, causing elevation and paradoxical movement with respiratory distress. TTN and RDS affect lung mechanics diffusely and do not cause isolated diaphragmatic paralysis. Choanal atresia is an upper-airway obstruction without this imaging finding.

151. B. Leukocyte adhesion deficiency prevents effective neutrophil extravasation and classically causes delayed umbilical cord separation, recurrent infection, and little pus despite leukocytosis. Antibody deficiencies do not produce this characteristic early adhesion phenotype. Complement defects have different infection patterns.

152. D. Hyperglycemia in extremely preterm infants reflects limited insulin response, stress, medications, and sometimes excessive glucose delivery. The glucose infusion rate should be reviewed first while sepsis and other contributors are assessed. Insulin may be used selectively, but aggressive boluses without correcting excessive substrate can cause dangerous hypoglycemia.

153. A. Pneumatosis intestinalis in a preterm infant with systemic and gastrointestinal signs is highly characteristic of NEC. Management includes bowel rest, decompression, antibiotics, and supportive care, with surgical evaluation for perforation or necrosis. Hirschsprung disease and pyloric stenosis have different timing and imaging findings.

154. D. Hypernatremia with high serum osmolality and inappropriately dilute polyuria indicates deficient antidiuretic hormone action, which can follow severe central nervous system injury. SIADH produces the opposite pattern. Acute kidney injury can be nonoliguric, but it does not typically produce this classic water-diuresis physiology.

155. C. The combination of characteristic craniofacial findings, neonatal hypotonia, and complete AV septal defect strongly suggests trisomy 21. Turner syndrome more often involves lymphedema and left-sided obstructive heart disease. Trisomy 18 and 22q11.2 deletion have different characteristic anomaly patterns.

156. D. Omenn syndrome is a form of SCID with oligoclonal activated T cells and can present with erythroderma, diarrhea, lymphadenopathy, eosinophilia, and elevated IgE. This severe inflammatory phenotype is not a normal variation of neonatal immunity. Early specialist evaluation and definitive immune therapy are required.

157. A. In transposition, survival depends on mixing between parallel systemic and pulmonary circulations. Prostaglandin maintains ductal patency, but a restrictive atrial septum may require urgent balloon atrial septostomy to create effective mixing. Closing the ductus or delaying treatment can worsen profound hypoxemia.

158. A. Congenital hypothyroidism can be clinically subtle at birth but untreated hormone deficiency can impair neurodevelopment. Confirmatory testing and prompt levothyroxine treatment are essential, and imaging should not delay therapy. Antithyroid medication would worsen hypothyroidism.

159. B. Severe ROP is treated by reducing the angiogenic stimulus from avascular retina, traditionally with laser photocoagulation and, in selected cases, intravitreal anti-VEGF therapy. Choice and follow-up depend on zone, stage, plus disease, systemic considerations, and local expertise. ROP is not an infectious retinitis.

160. C. A soft systolic murmur radiating to the axillae and back in an otherwise well neonate is typical of physiologic peripheral pulmonary artery stenosis and often resolves with growth. Critical coarctation, large VSD, and pulmonary atresia produce additional hemodynamic or oxygenation abnormalities.

161. A. Macroglossia, omphalocele, overgrowth, and neonatal hypoglycemia are classic for Beckwith-Wiedemann syndrome. The disorder is related to abnormal imprinting in the 11p15 region and carries tumor-surveillance implications. The alternative syndromes do not produce this characteristic overgrowth phenotype.

162. D. Cystic periventricular leukomalacia injures white matter near the lateral ventricles, including descending motor pathways to the legs, and is associated with spastic diplegia. The injury also increases risk of cognitive and visual-perceptual problems. SMA and muscular dystrophy are genetic neuromuscular disorders with different pathology.

163. A. Maple syrup urine disease results from impaired metabolism of branched-chain amino acids and can cause neonatal encephalopathy with a characteristic odor. Rapid recognition and treatment are important to limit neurotoxicity. The other disorders have different metabolites, triggers, and early presentations.

164. C. Chromosomal microarray is widely used as a first-tier test for infants with multiple congenital anomalies or unexplained developmental disorders because it detects clinically important copy-number variants. Targeted testing is added when a specific syndrome is suspected. Hemoglobin electrophoresis and metabolic assays answer different clinical questions.

165. B. Aplasia cutis congenita is a congenital absence of skin, most often on the scalp, ranging from small superficial defects to deeper lesions. Scalp hemorrhages and edema present as swellings rather than a true absence of skin. Larger or atypical lesions may prompt assessment for associated skull or syndromic abnormalities.

166. B. Visible jaundice during the first 24 hours is not considered routine physiologic jaundice and should prompt measurement and assessment for hemolysis, infection, bruising, or other causes. Treatment decisions depend on bilirubin level, age in hours, gestational age, and neurotoxicity risk factors. Breastfeeding should not be stopped solely because jaundice appears early.

167. A. Perinatal arterial ischemic stroke often presents with focal neonatal seizures and a focal arterial-territory lesion on MRI. Many cases occur without an obvious obstetric sentinel event. Long-term follow-up is important because hemiplegic cerebral palsy, epilepsy, and developmental effects may emerge later.

168. B. Prolonged parenteral nutrition, lack of enteral stimulation, inflammation, and intestinal failure can contribute to cholestasis. A rising direct bilirubin is not physiologic jaundice or breast milk jaundice, which are predominantly unconjugated. Evaluation should also exclude infection, anatomic obstruction, and metabolic liver disease.

169. C. Neonatal hypotension is physiologically heterogeneous; blood pressure alone does not identify the mechanism. Targeted echocardiography and clinical perfusion data can help distinguish preload limitation, ventricular dysfunction, and vascular tone abnormalities. Reflexive fluid loading or ductal treatment without supportive physiology can be harmful.

170. B. Maternal anti-Ro/SSA and anti-La/SSB antibodies can cross the placenta and injure the fetal atrioventricular conduction system, causing irreversible congenital heart block. Accessory-pathway SVT, diabetic hypertrophic cardiomyopathy, and medication-related ductal constriction have different mechanisms and are not linked to anti-Ro antibodies.

171. C. Donor human milk is commonly used for very preterm infants when mother's own milk is insufficient, particularly when reducing NEC risk is a priority. Because pasteurization and donor milk

composition may reduce some nutrient density, fortification is usually needed. Cow's milk and water are not appropriate neonatal substitutes.

172. A. Leukocoria can reflect congenital cataract, retinoblastoma, retinal detachment, or other serious ocular disease and warrants prompt evaluation. A normal red reflex should be symmetric. Early treatment of visually significant cataract is particularly important to prevent deprivation amblyopia.

173. B. Bilious emesis in a newborn is an emergency because malrotation with midgut volvulus can rapidly compromise intestinal blood flow. Shock and bloody stool increase concern for ischemia. Reflux and pyloric stenosis are typically nonbilious and do not explain the acute ischemic picture.

174. C. Classic galactosemia can present after lactose exposure with liver dysfunction, hypoglycemia, feeding intolerance, and a striking association with E coli sepsis. Immediate removal of galactose/lactose is required while confirmatory testing proceeds. PKU and MCAD deficiency do not produce this characteristic milk-triggered hepatic-sepsis syndrome.

175. D. Persistent shock after standard resuscitation in the setting of obvious blood loss should prompt rapid volume replacement. Isotonic crystalloid can be used, while uncrossmatched O-negative or immediately available compatible blood is preferred when hemorrhage is substantial. Volume restriction or diuresis would worsen hypovolemia.

176. D. Osteogenesis imperfecta results from quantitative or qualitative abnormalities of type I collagen and can present with fractures, osteopenia, and blue sclerae. Achondroplasia changes endochondral bone growth rather than causing generalized bone fragility. SMA is neuromuscular, and hypophosphatasia has a different biochemical mechanism.

177. B. Anemia of prematurity reflects a blunted erythropoietin response to falling oxygen tension, shortened red-cell survival, rapid growth, and frequent laboratory blood loss. Reticulocyte production is often inappropriately low for the degree of anemia. Hemolysis would generally increase reticulocytes unless marrow response is impaired.

178. D. Bilateral choanal atresia can cause severe cyclical cyanosis in a newborn because nasal breathing predominates; symptoms often improve with crying as the mouth opens. Inability to pass a catheter through the nares supports the diagnosis. Laryngomalacia and vocal cord paralysis cause stridor, not posterior nasal obstruction.

179. B. Hemophilia A is an X-linked factor VIII deficiency that can present with procedural bleeding and an isolated prolonged aPTT. Family history through the maternal line can provide an important clue. Platelet disorders and DIC produce different laboratory patterns, while vitamin K deficiency typically prolongs PT first.

180. A. Placental insufficiency can produce chronic fetal hypoxemia, which raises fetal erythropoietin and increases erythropoiesis. This is a classic setting for neonatal polycythemia in a growth-restricted

infant. Maternal red cells do not normally provide the newborn's erythrocyte mass, and hypoxemia stimulates rather than suppresses fetal erythropoietin.

181. B. Monochorionic placentas contain vascular anastomoses that can permit net blood transfer from a donor to a recipient twin. This produces anemia and growth restriction in the donor and polycythemia or volume loading in the recipient. The pattern is not explained by equal blood loss or maternal red-cell transfer.

182. C. Achondroplasia is usually caused by an activating FGFR3 variant and produces disproportionate short stature with rhizomelic limb shortening and characteristic craniofacial features. Osteogenesis imperfecta causes bone fragility, while dystrophinopathy and Pompe disease cause neuromuscular weakness rather than this skeletal pattern.

183. B. Human milk is preferred for very preterm infants and is associated with lower NEC risk than formula. Donor milk is commonly used when sufficient maternal milk is unavailable, with fortification added to meet preterm nutrient needs. Prolonged avoidance of enteral feeding carries its own nutritional and gastrointestinal disadvantages.

184. C. G6PD deficiency can cause episodic oxidative hemolysis and severe neonatal hyperbilirubinemia, often with a negative direct antiglobulin test. Risk varies by ancestry and specific variant. Biliary atresia and Gilbert syndrome do not cause oxidant-triggered hemolysis.

185. C. Abrupt deterioration with unilateral absent breath sounds, hyperexpansion, and mediastinal shift strongly suggests tension pneumothorax. Hemodynamic compromise requires immediate decompression rather than waiting for confirmatory imaging. Increasing airway pressure can worsen the air leak.

186. D. Gray baby syndrome historically resulted from chloramphenicol accumulation in neonates because hepatic glucuronidation and renal excretion were immature. Toxic concentrations can cause gastrointestinal symptoms, gray discoloration, and cardiovascular collapse. The syndrome illustrates why neonatal drug dosing cannot simply be scaled from older children.

187. D. Profound congenital complete heart block with low output may require pacing, particularly when the ventricular rate is very slow and the infant is symptomatic. Adenosine treats certain tachyarrhythmias, not complete block. Ductal manipulation and pulmonary vasodilation do not restore atrioventricular conduction.

188. C. Late-pregnancy SSRI exposure can be followed by a self-limited poor neonatal adaptation syndrome with jitteriness, respiratory symptoms, and feeding difficulty. Opioid withdrawal has a different exposure history and characteristic course, while congenital myasthenia requires maternal antibodies. Infection must still be considered clinically, but SSRI exposure itself does not cause bacterial sepsis.

189. C. Infants requiring advanced resuscitation need monitored post-resuscitation care, including early and serial glucose assessment. Both hypoglycemia and hyperglycemia can occur in critically ill

newborns and may affect neurologic risk. Temperature and cardiorespiratory status also require ongoing monitoring rather than being ignored after initial stabilization.

190. B. A persistent hyperchloremic, non-anion-gap acidosis with inappropriate urinary acidification suggests renal tubular acidosis or renal immaturity-related bicarbonate handling problems. Urea-cycle defects usually cause respiratory alkalosis and hyperammonemia. Lactic acidosis increases the anion gap rather than producing this pattern.

191. A. After secondary causes are excluded, recurrent apnea in a preterm infant is usually apnea of prematurity related to immature respiratory control. Caffeine is first-line pharmacologic therapy because it stimulates respiratory drive and reduces clinically important apnea. Sedation and paralysis worsen respiratory control, and PDA is not diagnosed solely from apnea.

192. D. A failed newborn hearing screen does not by itself establish permanent hearing loss, but it requires prompt follow-up so diagnosis and intervention are not delayed. The pathway may include rescreening and diagnostic ABR depending on local protocol and risk factors. Early hearing access is important for language development.

193. B. Hyperinsulinism is characterized by inappropriate insulin action during hypoglycemia with suppressed ketones and free fatty acids and a high glucose requirement. Insulin prevents access to alternative fuels, increasing neurologic risk. Fatty-acid oxidation disorders also cause hypoketotic hypoglycemia but do not explain clearly detectable inappropriate insulin in this setting.

194. A. Early abnormal motor signs in a high-risk infant can predict cerebral palsy and justify prompt standardized assessment and early intervention. Modern follow-up aims to identify high-risk cerebral palsy before a late definitive label when possible. Waiting for missed milestones can delay beneficial therapy and family support.

195. B. Once a preterm infant is clinically ready, progressive enteral feeding supports intestinal maturation, growth, and earlier discontinuation of central-line-dependent parenteral nutrition. No feeding strategy eliminates NEC, and parenteral nutrition has infectious, metabolic, and hepatobiliary risks. Advancement should still be individualized to clinical tolerance.

196. C. PPHN improves when pulmonary vascular resistance falls, allowing more blood to traverse the pulmonary circulation and reducing right-to-left shunting. Increasing pulmonary resistance or reducing lung aeration worsens the fetal-pattern circulation. Ductal constriction is not a universal remedy and may be harmful depending on ventricular physiology.

197. C. A sentinel hypoxic event, severe acidosis, need for resuscitation, and evolving encephalopathy with seizures are characteristic of HIE. The diagnosis and severity guide consideration of therapeutic hypothermia and neurologic monitoring. Benign sleep myoclonus does not cause global encephalopathy.

198. D. A coiled tube in the proximal esophageal pouch confirms esophageal atresia. Gas in the abdomen indicates a distal fistulous connection from the trachea to the distal esophagus, the most common configuration. Pure atresia generally produces a gasless abdomen.

199. B. Unintentional hypothermia is associated with increased metabolic demand and adverse outcomes in preterm infants. Controlled rewarming with close physiologic monitoring is appropriate while causes and complications such as hypoglycemia are addressed. Newborns do not rely effectively on shivering, and overheating is also harmful.

200. B. Maternal varicella shortly before or just after delivery can cause severe neonatal disease because fetal exposure occurs before the mother has generated and transferred sufficient protective antibody. Timing of exposure guides use of postexposure prophylaxis and antiviral therapy. A live varicella vaccine is not a birth prophylaxis for neonates.