COMPREHENSIVE MOCK EXAM 4

Supplemental to: Neonatology Board Review 2026-2027

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American Board of Pediatrics (ABP) Neonatal-Perinatal Medicine subspecialty certification examination - 200-question MEDREV supplemental simulation, A-D single-best-answer format, domain-weighted to the approved ABP content outline

Board-Review Questions

1. A former 24-week infant is discharged after a prolonged NICU course. Which follow-up model best addresses long-term risk?

A. Continue subspecialty respiratory follow-up and use primary care to initiate developmental referral if milestone concerns emerge later.

B. Use routine primary-care growth checks and defer formal developmental, hearing, and vision follow-up unless a specific symptom appears.

C. Multidisciplinary high-risk follow-up should integrate growth, nutrition, motor and cognitive development, hearing, vision, family needs, and primary care coordination.

D. Transfer long-term follow-up entirely to a neurology clinic because prematurity-related outcomes are primarily neurologic rather than multisystem in the situation described.

2. A newborn is delivered to a mother known to be HBsAg positive. Which intervention should occur within 12 hours of birth?

A. Give hepatitis B vaccine alone and defer immune globulin until the infant's HBsAg result is available at 6 months of age.

B. Give hepatitis B immune globulin alone because active vaccination is ineffective during the first month of life after maternal exposure.

C. Administer hepatitis B vaccine and hepatitis B immune globulin at separate injection sites, regardless of birth weight here.

D. Delay both vaccine and immune globulin until maternal HBV DNA is repeated because prophylaxis is unnecessary if the newborn appears well.

3. A hearing-screening test has a specificity of 98%. What does this indicate?

A. Among many positive screens, 98% are true positives regardless of the prevalence of hearing loss.

B. Among infants with the hearing disorder, about 98% will have a positive screening result.

C. The test detects a hearing disorder 98% earlier than clinical observation would detect the same condition.

D. Among infants without the target hearing disorder, about 98% will have a negative screening result.

4. A newborn has truncus arteriosus, hypocalcemic seizures, and a small thymic shadow. Which genetic diagnosis best explains the combination?

A. Trisomy 21 causing isolated atrioventricular septal disease with normal calcium regulation and normal thymic development.

B. 22q11.2 deletion syndrome causing conotruncal heart disease, hypoparathyroidism, and thymic hypoplasia given these findings.

C. Turner syndrome causing coarctation and congenital lymphedema without a characteristic association with thymic hypoplasia.

D. Prader-Willi syndrome causing neonatal hypotonia and poor feeding together with conotruncal malformations and hypocalcemia here.

5. Which infant should clearly be included in a retinopathy of prematurity screening program?

A. A 31-week infant weighing 1.7 kg with an uncomplicated course and no prolonged oxygen exposure because gestational age above 30 weeks still requires routine serial examinations.

B. A 33-week infant weighing 1.9 kg who needed brief delivery-room CPAP but has remained stable in room air because any respiratory support places the infant in the standard screening group.

C. A 35-week infant weighing 2.1 kg who received low-flow oxygen for transient tachypnea for one day because short-term oxygen exposure is the principal screening criterion.

D. An infant born at 27 weeks and 950 g because extreme prematurity and very low birth weight confer substantial ROP risk given the gestational and clinical findings described.

6. A healthy 2-day-old has scattered erythematous macules and papules with small central pustules on the trunk. The infant is afebrile and feeding well. Which diagnosis is most likely?

A. Congenital candidiasis, which can cause diffuse erythematous papules and pustules, often present at birth and sometimes followed by scale or desquamation.

B. Neonatal herpes simplex infection, which can produce grouped vesicles or erosions and may be accompanied by systemic or neurologic illness.

C. Transient neonatal pustular melanosis, which begins with superficial pustules that rupture and leave hyperpigmented macules with a collarette of scale.

D. Erythema toxicum neonatorum, a benign self-limited eruption that requires reassurance rather than antimicrobial therapy given these findings.

7. A newborn screen suggests congenital hypothyroidism. The infant looks well. What is the most important next step?

A. Repeat the newborn screen before obtaining serum thyroid tests because a clinically well infant is unlikely to need immediate confirmatory evaluation here.

B. Obtain confirmatory serum thyroid testing promptly and begin levothyroxine without unnecessary delay when primary congenital hypothyroidism is confirmed.

C. Begin antithyroid medication because an elevated screening TSH indicates neonatal thyrotoxicosis rather than thyroid hormone deficiency.

D. Repeat the newborn screen at one year and avoid serum testing because treatment before then interferes with normal thyroid maturation.

8. Two newborns have the same total serum bilirubin concentration, but one is 36 hours old at 35 weeks' gestation with hemolysis and the other is 5 days old at 40 weeks without risk factors. Why might their treatment thresholds differ?

A. Phototherapy and escalation thresholds depend on postnatal age, gestational age, and neurotoxicity risk factors rather than a single universal bilirubin value.

B. Use a common treatment threshold after the first day because neurotoxicity risk is determined mainly by the measured bilirubin concentration.

C. Base treatment primarily on birth weight because smaller infants tolerate lower bilirubin concentrations regardless of postnatal age or hemolysis status for this case.

D. Base treatment primarily on postnatal age because gestational age and hemolysis influence follow-up intensity but not phototherapy thresholds.

9. A therapy lowers the rate of severe BPD from 30% to 20%. What is the number needed to treat to prevent one additional case?

A. The relative risk reduction is 10%, so the number needed to treat is 1/10 = 0.1.

B. The absolute risk reduction is 50%, so the number needed to treat is 2.

C. The absolute risk reduction is 10 percentage points, so the number needed to treat is 1/0.10 = 10.

D. The event-rate ratio is 0.67, so the number needed to treat is 0.67 regardless of follow-up duration.

10. A ventilated preterm infant suddenly becomes bradycardic and severely hypoxemic. Breath sounds are absent on the right, the chest is asymmetric, and transillumination is strongly positive on that side. What is the immediate priority?

A. Wait for a formal radiograph before treatment because pleural decompression is contraindicated without imaging confirmation in an unstable neonate.

B. Decompress a suspected tension pneumothorax immediately, followed by definitive pleural drainage as needed when the findings are considered together.

C. Administer surfactant before any other intervention because abrupt unilateral loss of breath sounds is the usual presentation of recurrent RDS.

D. Increase peak inspiratory pressure because unilateral absent breath sounds indicate worsening atelectasis that requires more aggressive ventilation here.

11. A 1,000-g infant is tolerating full human-milk feeds but has suboptimal growth. Why is human-milk fortification commonly used in this setting?

A. Fortification increases protein, energy, calcium, phosphorus, and other nutrients to better meet the high growth needs of very preterm infants here.

B. Fortification mainly increases feeding volume so the infant receives more calories without materially changing the protein or mineral concentration of human milk.

C. Fortification is used chiefly to reduce feeding osmolality and thereby lower the risk of necrotizing enterocolitis in very-low-birth-weight infants.

D. Fortification replaces the need for separate micronutrient planning because standard fortifiers provide complete long-term nutrition at any feeding volume.

12. An infant of a diabetic mother becomes jittery at 18 hours of age. Glucose is normal, but total and ionized calcium are low. Which association is most relevant?

A. Maternal diabetes is associated mainly with late hypercalcemia from increased fetal parathyroid activity, so early low ionized calcium suggests an unrelated disorder.

B. Jitteriness with low calcium in an infant of a diabetic mother usually reflects primary neonatal hypoparathyroidism that is independent of magnesium status.

C. Infants of diabetic mothers are at increased risk of early neonatal hypocalcemia, partly related to altered parathyroid response and magnesium status.

D. Mild early hypocalcemia can occur during normal transition, so calcium replacement is unnecessary unless the glucose level is also low.

13. A newborn has petechiae, hepatosplenomegaly, microcephaly, and periventricular calcifications. Which test best confirms congenital CMV rather than postnatal acquisition?

A. Perform CMV PCR on saliva or urine within the first 3 weeks of life, with urine confirmation when a saliva screen is positive in this clinical context.

B. Obtain CMV IgM and IgG serology during the first three weeks because neonatal antibody patterns can distinguish congenital from postnatal infection.

C. Use a dried-blood-spot antibody assay after the newborn screen because serology is more specific than direct viral detection in urine or saliva.

D. Obtain a urine CMV PCR at 6 to 8 weeks because later testing provides a higher viral load and therefore distinguishes congenital infection more reliably.

14. In a randomized neonatal trial, some infants assigned to treatment A cross over and receive treatment B. What analysis best preserves the benefit of randomization for the primary comparison?

A. Analyze participants according to their original randomized assignment using the intention-to-treat principle in the neonatal setting described.

B. Analyze infants according to the treatment actually received because crossover makes original randomization less relevant to the estimate of treatment effect.

C. Restrict the primary analysis to infants who adhered fully to the assigned regimen because this best estimates efficacy without dilution from nonadherence.

D. Exclude crossover participants from the primary analysis because their treatment exposure cannot be assigned cleanly to either randomized group.

15. A newborn with a lethal or life-limiting condition is receiving comfort-focused care. Which statement best describes neonatal palliative care?

A. Reserve palliative-care consultation until the family has chosen to forgo disease-directed treatment, so the goals of intensive and comfort-focused care remain separate.

B. Use palliative care mainly for medication-based symptom control near the end of life, while psychosocial and bereavement support remain separate services.

C. Palliative care can be integrated from diagnosis and focuses on comfort, symptom relief, family support, communication, memory-making, and care aligned with goals.

D. Transfer an infant to a dedicated palliative unit before providing comfort-focused care because intensive-care monitoring is inconsistent with palliative goals.

16. A term newborn is lethargic and having seizures. Plasma glucose is 23 mg/dL. What is the immediate treatment priority?

A. Give enteral glucose immediately and reassess in 30 minutes because even neurologic symptoms can often be managed without intravenous therapy in a term infant.

B. Give buccal dextrose gel and a feed as the sole initial therapy because this approach is sufficient even with seizures and a plasma glucose in the low 20s.

C. Begin a continuous high-calorie enteral feed and avoid an IV glucose bolus because rapid correction can cause rebound neurologic injury.

D. Give prompt intravenous dextrose while confirming the value and evaluating why severe symptomatic hypoglycemia occurred for this infant.

17. A newborn screen is positive for phenylketonuria. Which pathophysiologic process causes neurologic injury if the disorder is untreated?

A. Phenylalanine hydroxylase deficiency causes accumulation of phenylalanine and insufficient tyrosine production, with toxic effects on the developing brain.

B. Galactose-1-phosphate uridyltransferase deficiency causes phenylalanine accumulation and can be treated by restricting dietary phenylalanine.

C. Biotinidase deficiency causes isolated tyrosine excess that secondarily blocks phenylalanine absorption from the intestine.

D. Medium-chain acyl-CoA dehydrogenase deficiency causes persistent hyperphenylalaninemia because fatty acids are the main substrate for phenylalanine hydroxylase.

18. An extremely preterm infant remains ventilator dependent with a high predicted risk of bronchopulmonary dysplasia. Which statement best reflects the role of postnatal systemic corticosteroids?

A. Use early inhaled corticosteroid therapy as the preferred strategy because airway delivery avoids systemic adverse effects while reliably preventing BPD.

B. Avoid systemic corticosteroids even when prolonged ventilation continues because extubation benefit has not been demonstrated in high-risk preterm infants.

C. Use a routine high-dose dexamethasone course early in life because greater anti-inflammatory potency improves BPD-free survival without added neurodevelopmental risk.

D. A carefully selected low-dose course may be considered when the benefits of facilitating extubation outweigh neurodevelopmental and other steroid risks here.

19. A 26-week infant remains ventilator dependent. Echocardiography shows a large patent ductus arteriosus with left atrial enlargement and diastolic flow reversal in the descending aorta. What do these findings suggest?

A. The PDA is likely small because descending-aortic diastolic flow reversal mainly reflects high systemic vascular resistance rather than ductal steal.

B. The findings are more consistent with pulmonary outflow obstruction because left atrial enlargement and diastolic runoff occur when pulmonary flow is restricted.

C. The PDA is unlikely to be clinically important because left atrial enlargement reflects reduced pulmonary venous return rather than pulmonary overcirculation.

D. The ductal shunt is hemodynamically important, with pulmonary overcirculation and systemic steal that may contribute to clinical instability in this case.

20. An extremely preterm infant is at risk for metabolic bone disease. Which nutrient pair is most directly important for skeletal mineralization?

A. Vitamin K and folate are the principal minerals incorporated into hydroxyapatite during neonatal bone formation.

B. Sodium and chloride alone determine bone mineral content because calcium and phosphorus are transferred mainly after birth.

C. Calcium and phosphorus intake must be adequate because most fetal mineral accretion normally occurs during the third trimester.

D. Iron and copper are the primary determinants of bone mineral density and can replace calcium-phosphorus supplementation in preterm infants.

21. A newborn has excessive oral secretions, coughs and becomes cyanotic with the first feed, and an orogastric tube cannot be passed into the stomach. Which anomaly is most likely?

A. Duodenal atresia, which typically prevents the tube from entering the stomach and causes copious oral secretions before feeding begins.

B. Esophageal atresia with or without a tracheoesophageal fistula, requiring airway and aspiration precautions plus surgical evaluation.

C. Pyloric stenosis, which prevents passage of an orogastric tube through the esophagus and causes choking with the very first feed.

D. Hirschsprung disease, which blocks esophageal tube passage because distal aganglionosis causes generalized upper gastrointestinal obstruction.

22. A 25-week infant on mechanical ventilation suddenly develops bloody endotracheal secretions, worsening oxygenation, diffuse lung opacities, and a falling hematocrit. Which diagnosis is most likely?

A. Uncomplicated apnea of prematurity, which produces respiratory pauses without diffuse alveolar bleeding or new radiographic infiltrates.

B. Congenital lobar overinflation, which causes focal hyperlucency rather than acute bilateral opacification with blood in the airway.

C. Transient tachypnea of the newborn, which causes retained lung fluid but does not typically produce acute bloody airway secretions and anemia.

D. Pulmonary hemorrhage, which can occur in extremely preterm infants and may be associated with hemodynamic instability and a large PDA.

23. A 25-week infant on prolonged parenteral nutrition develops rising direct bilirubin and hepatomegaly. Which complication should be suspected?

A. Gilbert syndrome, because it routinely produces progressive conjugated hyperbilirubinemia and hepatomegaly in extremely preterm infants.

B. Breast-milk jaundice, because human milk exposure is the main cause of direct hyperbilirubinemia in infants receiving minimal enteral feeding.

C. Parenteral-nutrition-associated cholestasis, especially in the setting of prolonged PN, prematurity, limited enteral feeding, and recurrent illness.

D. Physiologic unconjugated jaundice, because prolonged intravenous nutrition selectively raises direct bilirubin without reflecting hepatobiliary disease.

24. A stable ventilated preterm infant with evolving chronic lung disease has a modestly elevated PaCO2 but acceptable pH and oxygenation. Which ventilation principle may reduce lung injury?

A. Normalize PaCO2 to the low 20s with large tidal volumes because hypocapnia is the main protective strategy against bronchopulmonary dysplasia.

B. Permissive hypercapnia can be acceptable within a clinically safe pH range if it allows lower ventilator pressures and tidal volumes in context.

C. Increase oxygen concentration rather than adjust ventilation because carbon dioxide levels do not reflect alveolar ventilation in preterm infants.

D. Use repeated manual hyperventilation because intermittent large breaths reduce volutrauma more effectively than consistent lung-protective settings.

25. A ventilated 26-week infant has PaCO2 values persistently in the low 20s mm Hg because of aggressive ventilation. Why is this undesirable?

A. Marked hypocapnia can reduce cerebral blood flow and is associated with brain injury, so ventilation should avoid unnecessary overventilation.

B. Marked hypocapnia increases cerebral blood flow and is routinely targeted to prevent periventricular leukomalacia in extremely preterm infants.

C. Hypocapnia prevents ventilator-induced lung injury clinically, so values below 25 mm Hg are generally preferred in evolving BPD.

D. Low PaCO2 has no meaningful physiologic effect in premature infants because cerebral vessels do not respond to carbon dioxide after birth.

26. A 6-week-old infant has tachypnea, diaphoresis with feeds, poor weight gain, and a harsh holosystolic murmur. Echocardiography shows a large ventricular septal defect. Why did symptoms become more prominent several weeks after birth?

A. Closure of the foramen ovale redirected venous blood across the VSD from right to left and produced isolated pulmonary edema.

B. The VSD enlarged rapidly after birth because muscular ventricular septum normally dissolves during the first two months of life.

C. Rising pulmonary vascular resistance increased the left-to-right shunt, causing pulmonary overcirculation primarily after the neonatal period.

D. Falling pulmonary vascular resistance increased the left-to-right shunt and pulmonary blood flow, producing congestive symptoms.

27. A 900-g preterm infant is clinically stable and the mother is expressing milk. Which enteral feeding source is preferred when available?

A. Pasteurized donor human milk should replace maternal milk because milk-bank processing standardizes nutritional content and makes it superior even when maternal milk is available.

B. Preterm formula should be the first choice because its higher protein and mineral content outweighs the protective advantages of the mother's own milk.

C. Extensively hydrolyzed formula should be preferred because reducing intact bovine protein is the most effective strategy for preventing feeding intolerance and NEC.

D. The mother's own milk is preferred because it is associated with important nutritional, immunologic, and gastrointestinal benefits for very-low-birth-weight infants.

28. A 3-week-old exclusively breast-fed infant who did not receive vitamin K prophylaxis presents with intracranial hemorrhage. Platelet count is normal and PT is markedly prolonged. Which diagnosis is most likely?

A. Hemolytic disease of the newborn because unconjugated bilirubin elevation directly prolongs PT and causes isolated intracranial bleeding.

B. Vitamin K deficiency bleeding because low vitamin-K-dependent clotting factor activity can cause severe bleeding despite a normal platelet count.

C. Hemophilia A is most likely because factor VIII deficiency can cause intracranial hemorrhage while leaving the platelet count normal.

D. Severe liver dysfunction is most likely because impaired hepatic factor synthesis can prolong PT and cause intracranial bleeding in a young infant.

29. A preterm infant with a peripherally inserted central catheter develops unilateral arm swelling. Doppler ultrasonography confirms a catheter-associated venous thrombosis. Which principle should guide management?

A. Remove the catheter immediately and observe without anticoagulation because source removal is sufficient for most symptomatic neonatal upper-extremity thromboses.

B. Leave the catheter in place if it functions and monitor the clot without anticoagulation because propagation risk is low in line-associated neonatal thrombosis.

C. Assess whether the catheter is still needed and manage anticoagulation based on clot extent, symptoms, bleeding risk, and neonatal thrombosis guidance for this case.

D. Use thrombolytic therapy to restore venous patency promptly because symptomatic catheter-associated thrombosis is an indication for fibrinolysis before anticoagulation.

30. A newborn has abdominal distention and fails to pass meconium. Contrast enema shows a microcolon with inspissated pellets in the terminal ileum. Which inherited disorder is strongly associated with this presentation?

A. Cystic fibrosis, in which thick intestinal secretions can cause meconium ileus in the newborn period in this clinical context.

B. Prader-Willi syndrome, in which hypotonia commonly causes a microcolon filled with inspissated meconium from CFTR dysfunction.

C. Turner syndrome, which commonly produces distal intestinal obstruction from thick mucus despite normal exocrine pancreatic function.

D. Noonan syndrome, which is strongly associated with meconium ileus because RAS-pathway activation reduces pancreatic chloride secretion.

31. A patient at 27 weeks is expected to deliver imminently because of severe preeclampsia. Which maternal medication is used specifically for fetal neuroprotection when very preterm birth is anticipated?

A. Administer maternal phenobarbital because suppressing fetal cortical activity prevents germinal-matrix hemorrhage after birth here.

B. Administer high-dose vitamin K because fetal coagulation enhancement is the principal strategy for preventing cerebral palsy.

C. Administer maternal furosemide because lowering fetal intravascular volume reduces pressure-related white-matter injury.

D. Administer magnesium sulfate before delivery because exposure shortly before very preterm birth reduces the risk of cerebral palsy.

32. A 12-day-old infant presents with fever, lethargy, and bulging fontanelle. Bacterial meningitis is suspected. Which management principle is most appropriate?

A. Obtain neuroimaging before antimicrobial therapy because a bulging fontanelle raises concern for increased pressure and lumbar puncture or treatment should wait for imaging.

B. Begin narrow therapy directed at group B Streptococcus after cultures because broad empiric neonatal coverage is unnecessary once meningitis is clinically suspected.

C. Obtain cultures promptly and start age-appropriate parenteral empiric antibiotics without delaying therapy for nonessential diagnostic procedures here.

D. Perform lumbar puncture and await initial CSF cell count and Gram stain before starting antibiotics so treatment can be targeted from the first dose.

33. A term newborn has severe respiratory distress and a scaphoid abdomen. Prenatal imaging showed a left-sided congenital diaphragmatic hernia. What is the preferred initial respiratory approach?

A. Intubate for controlled ventilation and decompress the stomach with an orogastric tube while avoiding routine bag-mask ventilation.

B. Perform immediate surgical repair in the delivery room before stabilizing oxygenation, ventilation, and pulmonary hypertension here.

C. Begin nasal CPAP as the definitive initial treatment because spontaneous breathing prevents bowel distention in the thorax.

D. Use vigorous bag-mask ventilation to recruit the left lung rapidly before inserting a gastric tube or securing the airway.

34. A newborn with Down syndrome has leukocytosis with circulating blasts that resemble megakaryoblasts. The infant is clinically stable, and the blast count begins to fall spontaneously. Which diagnosis is most likely?

A. Chronic myeloid leukemia, which commonly presents at birth in Down syndrome with a transient GATA1-mutated megakaryoblastic proliferation.

B. Physiologic neonatal leukocytosis, because circulating blasts in a newborn with trisomy 21 generally does not represent a clonal hematologic process.

C. Acute lymphoblastic leukemia, which is defined by spontaneous disappearance of megakaryoblasts during the first weeks of life in most infants with Down syndrome here.

D. Transient abnormal myelopoiesis associated with trisomy 21, which can resolve spontaneously but requires monitoring for complications and later myeloid leukemia risk.

35. A newborn has a dense unilateral congenital cataract that obscures the visual axis. What is the most important management principle?

A. Use topical antibiotics as first-line therapy because congenital cataracts are usually bacterial infections of the lens that clear with treatment.

B. Patch the affected eye continuously because preventing light entry into the cataractous eye stimulates normal visual cortical development.

C. Prompt ophthalmologic evaluation and timely treatment are required because prolonged visual deprivation can cause irreversible amblyopia here.

D. Observe until school age because monocular visual pathways remain fully plastic throughout childhood and early treatment offers no advantage.

36. A newborn has a midline abdominal wall defect at the umbilical ring containing liver and bowel covered by a membrane. Which associated issue deserves particular attention?

A. The lesion is gastroschisis, which is usually covered by a membrane and has a stronger association with aneuploidy than omphalocele.

B. The finding is physiologic midgut herniation, which normally persists after term birth and needs no congenital anomaly assessment.

C. Omphalocele is associated with chromosomal, cardiac, and other structural anomalies, so a broad congenital-anomaly evaluation is needed.

D. The defect represents isolated umbilical hernia, which commonly contains liver and requires emergency operative closure in the delivery room.

37. A stable 2-week-old has a regular narrow-complex tachycardia at 260 beats/min. Vagal maneuvers do not terminate the rhythm and blood pressure is preserved. What is the next treatment?

A. Give rapid intravenous adenosine while monitoring the rhythm because reentrant supraventricular tachycardia is likely in this case.

B. Administer indomethacin because closure of a patent ductus is the standard treatment for regular narrow-complex neonatal tachycardia.

C. Give intravenous atropine because increasing atrioventricular nodal conduction terminates most neonatal reentrant tachycardias.

D. Begin chest compressions because any neonatal heart rate above 250 beats/min should be treated as pulseless arrest despite preserved perfusion.

38. A newborn has a webbed neck, low-set ears, pectus deformity, pulmonary valve stenosis, and normal chromosomes on routine karyotype. Which syndrome is most likely?

A. Turner syndrome, which requires monosomy X and therefore remains the best diagnosis despite a normal chromosomal analysis here.

B. Trisomy 18, which is defined by severe growth restriction, overlapping fingers, and a characteristic autosomal aneuploidy.

C. Beckwith-Wiedemann syndrome, which causes macrosomia and macroglossia rather than pulmonary stenosis and a webbed neck.

D. Noonan syndrome, a RAS-pathway disorder that can resemble Turner syndrome but occurs with a normal sex-chromosome complement.

39. A term infant with culture-positive sepsis develops acute diffuse bilateral lung disease, severe hypoxemia, and no primary cardiac explanation for pulmonary edema. Which concept best describes the respiratory process?

A. Neonatal acute respiratory distress can result from inflammatory lung injury with impaired gas exchange in severe systemic illness in this setting.

B. Transient tachypnea is the likely diagnosis because any neonatal pulmonary edema reflects delayed fetal lung-fluid absorption rather than inflammation.

C. Congenital diaphragmatic hernia is the defining cause of diffuse inflammatory lung injury when echocardiography excludes congenital heart disease.

D. Respiratory distress syndrome from developmental surfactant deficiency is certain because term infants with sepsis cannot develop secondary surfactant dysfunction.

40. A term infant with severe hyperbilirubinemia becomes lethargic, develops a high-pitched cry, poor feeding, and abnormal extensor tone. Which neurologic complication is most concerning?

A. Benign neonatal sleep myoclonus, which typically presents with lethargy, abnormal tone, and a high-pitched cry during severe jaundice.

B. Periventricular leukomalacia, which is caused directly by unconjugated bilirubin deposition in periventricular white matter of term infants.

C. Spinal muscular atrophy, which can emerge abruptly during hyperbilirubinemia and produce extensor hypertonia rather than progressive weakness in this case.

D. Acute bilirubin encephalopathy from bilirubin neurotoxicity, which can progress to permanent kernicterus spectrum disorder without urgent treatment.

41. A former preterm infant had bilateral cystic periventricular leukomalacia. Which later motor outcome is particularly associated with this pattern of white-matter injury?

A. Progressive muscular dystrophy because preterm white-matter injury triggers ongoing degeneration of skeletal-muscle dystrophin.

B. Pure cerebellar ataxia without tone abnormality because periventricular white-matter injury spares corticospinal pathways.

C. Spastic cerebral palsy, often with greater lower-extremity involvement when periventricular corticospinal pathways are injured.

D. Isolated flaccid paraplegia from anterior horn cell loss because cystic PVL primarily destroys lower motor neurons in the spinal cord.

42. A term infant has severe but potentially reversible hypoxic respiratory failure from meconium aspiration and PPHN despite optimal ventilation, surfactant when indicated, inhaled nitric oxide, and cardiovascular support. What rescue therapy may be considered at an experienced center?

A. Routine bilateral lung transplantation is the next-line neonatal treatment because reversible respiratory failure rarely improves after maximal ventilation here.

B. Extracorporeal membrane oxygenation can provide temporary gas exchange and circulatory support while the underlying lung and pulmonary vascular disease recovers.

C. Chronic home oxygen should be started immediately because severe neonatal hypoxemia is best managed by avoiding further intensive care interventions.

D. High-dose indomethacin should be used as rescue therapy because ductal constriction directly substitutes for extracorporeal oxygenation.

43. A 700-g infant with a central venous catheter and prolonged broad-spectrum antibiotics develops thrombocytopenia, glucose instability, and persistent positive blood cultures for Candida albicans. Which management principle is most appropriate?

A. Treat with systemic antifungal therapy and remove the central venous catheter when feasible because invasive candidiasis can disseminate widely in extremely preterm infants.

B. Treat systemically but retain the central catheter if it remains functional because line removal does not materially affect clearance of neonatal candidemia.

C. Stop antifungal therapy after bloodstream cultures clear because a negative repeat culture is sufficient evidence that disseminated infection has been eradicated in this case.

D. Use enteral fluconazole alone for a clinically stable infant because systemic candidemia without shock can be managed without parenteral therapy.

44. A stable preterm infant has occasional small gastric residuals but a soft abdomen, normal stooling, no bloody stool, and no systemic instability. Which interpretation is most appropriate?

A. Gastric residuals alone are nonspecific; feeding decisions should integrate the full abdominal examination and systemic clinical picture for this case.

B. Pause feeds and obtain abdominal radiography whenever a gastric residual is present because residual volume is an early marker of evolving NEC.

C. Reduce feed volume and change to continuous infusion because intermittent residuals mainly indicate delayed gastric emptying that predicts intestinal intolerance.

D. Switch from human milk to a hydrolyzed formula because recurrent residuals are most often caused by milk-protein intolerance in preterm infants.

45. A term infant after severe brain injury develops rising serum sodium, high serum osmolality, very high urine output, and urine that remains markedly dilute. Which diagnosis is most likely?

A. SIADH from excessive antidiuretic hormone release, which produces water retention, hyponatremia, and concentrated urine.

B. Renal vein thrombosis, which produces hematuria and renal enlargement rather than profound polyuria with dilute urine.

C. Central diabetes insipidus from deficient antidiuretic hormone release, producing free-water loss and inappropriately dilute urine.

D. Acute tubular injury, which universally causes severe oliguria and therefore cannot produce any increase in urine output during recovery.

46. A newborn has severe cyanosis at rest that improves when crying. A catheter cannot be passed through either nostril into the pharynx. Which diagnosis is most likely?

A. Laryngomalacia, which causes inspiratory stridor that improves with feeding and prevents passage of nasal catheters bilaterally.

B. Pierre Robin sequence, which causes posterior tongue displacement but characteristically obstructs both nasal choanae on catheter testing.

C. Bilateral choanal atresia causing critical nasal airway obstruction in an obligate or preferential nasal-breathing newborn here.

D. Tracheoesophageal fistula, which causes nasal obstruction relieved by crying and prevents passage of catheters through the posterior nares.

47. A severely growth-restricted preterm infant starts aggressive parenteral nutrition and develops hypophosphatemia with hypokalemia. Which concept best explains this pattern?

A. Physiologic postnatal diuresis causes isolated phosphate loss but cannot affect potassium during nutritional advancement.

B. A refeeding-like intracellular shift can occur when anabolic nutrition is introduced to a chronically undernourished fetus with limited mineral stores.

C. Human-milk fortification causes immediate extracellular release of phosphate and potassium, making low serum levels evidence of laboratory error for this case.

D. Surfactant therapy directly binds circulating phosphate and potassium, so the abnormalities indicate an expected pulmonary medication effect.

48. A 27-week infant develops progressive normocytic anemia over several weeks without evidence of hemolysis or blood loss. Reticulocyte response is inappropriately low. Which mechanism contributes most to anemia of prematurity?

A. Chronic low-grade immune hemolysis from maternal red-cell antibodies causes the typical low-reticulocyte anemia that develops in most very preterm infants.

B. Excessive erythropoietin production suppresses marrow red-cell output through negative feedback, producing the characteristic low reticulocyte count.

C. Premature infants have abnormally long red-cell survival, so physiologic accumulation of old erythrocytes suppresses reticulocyte production.

D. An immature erythropoietin response to falling hemoglobin, combined with rapid growth and phlebotomy losses, limits compensatory erythropoiesis.

49. A preterm infant is delivered after maternal fever, uterine tenderness, fetal tachycardia, and foul-smelling amniotic fluid. Which neonatal problem should be anticipated most directly from this intrapartum history?

A. Late-onset candidemia is specifically predicted because maternal fever primarily transmits Candida through breast milk after birth.

B. Early-onset bacterial infection risk is increased, so the infant requires prompt clinical assessment and sepsis risk management.

C. Neonatal tetanus is the expected infection because foul-smelling amniotic fluid indicates contamination with Clostridium tetani.

D. Congenital cytomegalovirus is the main concern because intrapartum uterine tenderness is characteristic of primary maternal CMV infection.

50. A 30-week infant develops respiratory failure and hypotension within 3 hours of birth after prolonged rupture of membranes and maternal fever. Which empiric antibiotic regimen is commonly used while cultures are pending?

A. Use ampicillin plus a third-generation cephalosporin routinely because broader gram-negative coverage is preferred over an aminoglycoside for uncomplicated early-onset sepsis.

B. Ampicillin plus gentamicin provides standard initial coverage for common early-onset neonatal bacterial pathogens in this clinical setting in the situation described.

C. Use vancomycin plus gentamicin because resistant coagulase-negative staphylococci are the dominant pathogens in sepsis presenting during the first hours after birth.

D. Use ampicillin alone initially because maternal fever and prolonged rupture primarily increase the risk of group B streptococcal infection rather than gram-negative sepsis.

51. A 27-week infant has substantially lower serum IgG than a term infant. Which explanation is most accurate?

A. Most maternal IgG transfer occurs late in gestation, so very preterm delivery interrupts placental acquisition of passive antibody.

B. Preterm infants rapidly catabolize self-produced IgG because neonatal B cells produce higher IgG concentrations than adult B cells.

C. Maternal IgM crosses the placenta during the first trimester and normally replaces IgG as the major neonatal systemic immunoglobulin.

D. IgG does not cross the placenta, so term and preterm infants obtain equivalent passive systemic antibody primarily after breast-feeding begins.

52. An observational study finds that infants receiving prolonged mechanical ventilation have higher mortality. Sicker infants were also much more likely to receive prolonged ventilation. What epidemiologic problem is most important?

A. Confounding by illness severity may create or exaggerate the association because severity influences both the exposure and the outcome.

B. Spectrum bias is present because disease severity can affect primarily diagnostic-test performance, not associations between treatments and outcomes.

C. Randomization bias is present because observational studies randomly assign the sickest infants to longer ventilation.

D. Lead-time bias is the main issue because earlier diagnosis of respiratory failure typically lengthens measured survival without changing mortality.

53. A 29-week infant treated with surfactant has improving oxygen need, good spontaneous respiratory effort, and low ventilator settings. Which next step best supports lung-protective care?

A. Continue invasive ventilation for a fixed seven-day course because extubation after surfactant increases the risk of recurrent RDS.

B. Increase ventilator pressures before extubation because several hours of larger tidal volumes strengthen respiratory muscles in premature infants.

C. Discontinue distending pressure after extubation because CPAP delays alveolar fluid absorption and promotes recurrent surfactant deficiency.

D. Consider early extubation to noninvasive support such as CPAP when the infant meets readiness criteria in the neonatal setting described.

54. A newborn has multiple fractures of different ages, blue-gray sclerae, and generalized osteopenia. Which diagnosis is most likely?

A. Nutritional rickets from postnatal vitamin D deficiency, which commonly causes multiple prenatal fractures in a term newborn.

B. Osteogenesis imperfecta from abnormal type I collagen synthesis or structure, with severity varying by genotype in this newborn.

C. Hypophosphatasia is excluded by neonatal fractures because it causes primarily delayed tooth eruption without skeletal mineralization abnormalities.

D. Achondroplasia from FGFR3 activation, which causes rhizomelic shortening but not diffuse bone fragility and multiple fractures at birth.

55. A newborn's heart rate remains 50 beats/min after 30 seconds of effective positive-pressure ventilation with visible chest movement. What is the next resuscitation step?

A. Stop ventilation and give epinephrine immediately because compressions reduce pulmonary blood flow during neonatal transition.

B. Use a 15:2 compression-to-ventilation ratio because neonatal resuscitation follows the standard two-rescuer pediatric CPR sequence for this infant.

C. Begin coordinated chest compressions with ventilation, preferably after securing an alternate airway, using a 3:1 compression-to-ventilation ratio.

D. Continue ventilation alone for another five minutes because compressions are reserved for a heart rate below 30 beats/min.

56. Investigators compare two feeding strategies in preterm infants and want the strongest design for estimating a causal treatment effect while minimizing confounding. Which design is preferred when ethical and feasible?

A. A case series because describing consecutive infants receiving one feeding strategy provides an unbiased estimate of comparative treatment effect.

B. A randomized controlled trial because random allocation tends to balance measured and unmeasured prognostic factors between treatment groups.

C. A cross-sectional survey because measuring exposure and outcome at one time point establishes treatment temporality and controls confounding.

D. An ecological study because comparing hospital-level feeding rates eliminates individual-level confounding through aggregation.

57. A cyanotic newborn has pulmonary atresia with an intact ventricular septum and minimal antegrade pulmonary blood flow. Which initial therapy can preserve pulmonary perfusion?

A. Use inhaled nitric oxide to lower pulmonary vascular resistance and increase pulmonary blood flow without maintaining the ductus arteriosus.

B. Infuse prostaglandin E1 to maintain ductal blood flow to the pulmonary arteries while anatomy and intervention are defined in this setting.

C. Start a beta blocker to reduce infundibular spasm because fixed valvar atresia is a dynamic outflow obstruction similar to a tet spell.

D. Use a beta blocker to reduce right-ventricular oxygen demand because fixed pulmonary atresia can be stabilized by slowing heart rate until the valve opens.

58. A medication error results in a clinically important but treatable complication in a preterm infant. What is the appropriate professional response to the family?

A. Disclose the error honestly, explain known effects and next steps, apologize as appropriate, and activate institutional safety and reporting processes.

B. Withhold the information if the infant is expected to recover because disclosure creates unnecessary distress and does not improve patient safety.

C. Alter the medical record to remove the erroneous order before discussing the complication so the family receives a simplified explanation.

D. Delay disclosure until the formal root-cause analysis is complete so the family receives a single finalized explanation rather than preliminary information.

59. A vigorous 30-week infant is born and does not need immediate resuscitation. Which cord-management approach is preferred when maternal and fetal conditions permit?

A. Delay umbilical cord clamping for at least about 60 seconds to support placental transfusion in a stable preterm infant.

B. Clamp the cord immediately because preterm infants are harmed by any placental transfusion after the first breath.

C. Wait at least 10 minutes before clamping because shorter delays do not provide hematologic benefit to preterm infants.

D. Perform intact-cord milking repeatedly in infants below 28 weeks because this has replaced delayed cord clamping in extreme prematurity.

60. A 24-week infant on minimal feeds suddenly develops abdominal distention and free intraperitoneal air but has little bowel-wall thickening and no pneumatosis. Which diagnosis is most likely?

A. Meconium ileus, which causes fetal intestinal perforation primarily in term infants and cannot present as a focal postnatal perforation given these findings.

B. Necrotizing enterocolitis, which typically produces extensive pneumatosis and portal venous gas before any perforation can occur.

C. Hirschsprung enterocolitis, which is confined to term infants and is the usual cause of isolated free air in a 24-week neonate.

D. Spontaneous intestinal perforation, which often occurs in extremely preterm infants as a focal perforation without the diffuse bowel disease typical of NEC.

61. A preterm infant with a large intraventricular hemorrhage develops progressive ventricular enlargement on serial ultrasound and a rapidly increasing head circumference. What complication is developing?

A. Posthemorrhagic ventricular dilatation from impaired cerebrospinal fluid circulation after intraventricular bleeding for this infant.

B. Benign familial macrocephaly because rapid ventricular enlargement after a major IVH is unrelated to cerebrospinal fluid obstruction.

C. Craniosynostosis because premature suture fusion causes increasing ventricular size and rapid head growth after intraventricular hemorrhage.

D. Subgaleal hemorrhage because extracranial blood accumulation produces progressive enlargement of the cerebral ventricles on ultrasound.

62. A newborn has cleft lip and palate, postaxial polydactyly, microphthalmia, and a holoprosencephaly pattern on neuroimaging. Which diagnosis is most likely?

A. Trisomy 13, which commonly includes midline craniofacial abnormalities, polydactyly, ocular anomalies, and severe brain malformations.

B. Trisomy 18, which is more characteristically associated with overlapping fingers and rocker-bottom feet than postaxial polydactyly and holoprosencephaly.

C. Turner syndrome, which causes monosomy X and is not typically associated with holoprosencephaly or polydactyly.

D. Prader-Willi syndrome, which causes neonatal hypotonia and feeding difficulty but not major midline malformations and polydactyly.

63. A newborn has uncovered bowel loops protruding through a right-sided paraumbilical abdominal wall defect. What is the immediate management priority?

A. Search first for chromosomal anomalies because gastroschisis is enclosed in a sac and is more strongly linked to aneuploidy than omphalocele in context.

B. Begin oral feeds immediately because exposed bowel requires enteral nutrition to prevent ischemia before surgical reduction.

C. Protect the exposed bowel in a sterile moisture-preserving covering, decompress the stomach, prevent heat and fluid loss, and obtain surgical care.

D. Apply dry gauze tightly around the bowel because evaporative cooling reduces intestinal edema and improves spontaneous reduction.

64. A newborn has bilateral cataracts, a continuous murmur from a patent ductus arteriosus, and sensorineural hearing loss. Which congenital infection is most likely?

A. Congenital syphilis, which primarily causes cataracts and isolated PDA without rash, hepatosplenomegaly, or skeletal findings.

B. Congenital rubella syndrome, which classically includes cataracts, cardiac defects such as PDA, and sensorineural hearing impairment.

C. Neonatal HSV, which classically causes PDA and cataracts as a congenital malformation complex rather than vesicular or CNS disease.

D. Congenital toxoplasmosis, which more typically causes hydrocephalus, intracranial calcifications, and chorioretinitis rather than PDA with cataracts.

65. A 27-week infant receiving feeds develops abdominal distention, bloody stool, temperature instability, and pneumatosis intestinalis on radiography. What is the immediate nutritional management?

A. Increase enteral feeding volume because pneumatosis reflects hunger-related bowel gas and resolves faster with continuous milk infusion.

B. Continue the same feeds and add thickener because bloody stool in a preterm infant usually indicates uncomplicated gastroesophageal reflux in this clinical context.

C. Stop enteral feeds and provide bowel rest with gastric decompression while treating suspected necrotizing enterocolitis and supporting nutrition parenterally.

D. Switch immediately to whole cow's milk because higher mineral content promotes healing of injured premature intestinal mucosa.

66. An infant of a diabetic mother develops gross hematuria, thrombocytopenia, and a palpable flank mass. Doppler ultrasonography shows an enlarged kidney with abnormal venous flow. What is the most likely diagnosis?

A. Nephrotic syndrome, which primarily causes heavy proteinuria and edema rather than acute hematuria with an enlarged thrombosed kidney.

B. Posterior urethral valves, which cause bilateral urinary tract obstruction but do not characteristically cause a unilateral enlarged kidney with thrombocytopenia.

C. Renal vein thrombosis, for which dehydration, polycythemia, sepsis, and maternal diabetes are recognized risk factors for this case.

D. Renal tubular acidosis, which produces growth failure and metabolic acidosis rather than a flank mass and venous-flow abnormality.

67. A fetus develops hydrops with cardiomegaly and a markedly elevated middle cerebral artery peak systolic velocity in a pregnancy complicated by maternal red-cell alloimmunization. What is the most likely fetal problem?

A. Congenital nephrotic syndrome is causing isolated protein loss, and the elevated cerebral artery velocity is unrelated to anemia.

B. Fetal polycythemia is increasing blood viscosity and cardiac afterload, so partial exchange transfusion should be performed before birth.

C. Isolated fetal thrombocytopenia is producing hydrops through capillary bleeding despite a normal red-cell mass.

D. Severe fetal anemia is causing high-output cardiac failure and hydrops, and intrauterine transfusion may be required.

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

A. Respiratory distress syndrome from surfactant deficiency causing low compliance and diffuse alveolar collapse for the presentation described.

B. Congenital lobar overinflation from focal airway obstruction, which produces unilateral hyperlucency rather than diffuse low-volume disease.

C. Meconium aspiration syndrome from intrauterine aspiration, which more often causes coarse patchy infiltrates and hyperinflation in a term infant.

D. Transient tachypnea of the newborn from delayed lung-fluid clearance, which usually produces hyperinflation and prominent perihilar markings.

69. A 39-week infant has severe perinatal acidemia, required prolonged resuscitation, and at 2 hours of age has moderate encephalopathy with abnormal tone and depressed consciousness. Which therapy should be considered urgently if eligibility criteria are met?

A. Maintain mild hyperthermia for 24 hours because increased cerebral metabolism accelerates recovery after hypoxic-ischemic injury.

B. Use prophylactic high-dose corticosteroids because reducing systemic inflammation is the established neuroprotective therapy for term HIE.

C. Initiate whole-body or selective-head therapeutic hypothermia within the first 6 hours and maintain target cooling for about 72 hours.

D. Delay neuroprotective treatment until seizures occur because encephalopathy without seizures does not benefit from early intervention.

70. A term infant with hypoxic-ischemic encephalopathy has repetitive lip smacking and eye deviation, but many electrographic seizures have no visible clinical correlate. What monitoring method is most useful?

A. Pulse oximetry is the preferred seizure monitor because electrographic seizures usually produce a stereotyped oxygen desaturation pattern.

B. Continuous or prolonged EEG monitoring can detect subclinical neonatal seizures and help assess treatment response given these findings here.

C. Use serial brief bedside EEG recordings because intermittent studies capture enough of the seizure burden to guide therapy in most encephalopathic neonates.

D. A normal bedside neurologic examination excludes seizures because neonatal seizures typically produce obvious generalized tonic-clonic movements.

71. A term newborn with hypoplastic left heart syndrome becomes gray and acidotic as the ductus begins to close. Which medication is essential during initial stabilization?

A. Adenosine should be given continuously because systemic hypoperfusion in hypoplastic left heart syndrome is caused by occult SVT.

B. Digoxin should be used as sole therapy because increasing left ventricular contractility restores adequate aortic output despite hypoplasia.

C. Indomethacin should be given to accelerate ductal closure and force the left ventricle to support systemic circulation.

D. Prostaglandin E1 should be infused to maintain ductal systemic blood flow until staged cardiac intervention can be performed.

72. A cyanotic newborn has d-transposition of the great arteries and a restrictive atrial communication. Despite prostaglandin E1, oxygenation remains critically low. What intervention most directly improves mixing?

A. Close the ductus arteriosus pharmacologically because ductal mixing worsens systemic oxygenation in transposition.

B. Increase pulmonary blood flow with inhaled nitric oxide because better oxygenation of pulmonary venous blood will correct systemic hypoxemia despite restricted atrial mixing.

C. Perform balloon atrial septostomy to enlarge the interatrial communication and improve mixing between parallel circulations.

D. Place a pulmonary artery band because reducing pulmonary blood flow is the immediate method for improving systemic saturation.

73. A large-for-gestational-age infant requires a very high glucose infusion rate to maintain normal plasma glucose. Insulin is inappropriately detectable during hypoglycemia, beta-hydroxybutyrate is suppressed, and free fatty acids are low. What is the most likely mechanism?

A. Isolated glycogen storage disease typically produces measurable insulin during hypoglycemia and suppresses both ketones and free fatty acids.

B. Growth hormone excess is increasing ketone production and lipolysis, creating hypoglycemia despite abundant alternative cerebral fuels.

C. Cortisol excess is blocking gluconeogenesis and producing a classic hypoketotic state with inappropriately high insulin concentrations.

D. Hyperinsulinism is suppressing hepatic glucose output, lipolysis, and ketogenesis while increasing peripheral glucose utilization here.

74. An infant with symptomatic congenital CMV passes the newborn hearing screen. What follow-up principle is most appropriate?

A. Repeat hearing testing after speech or language delay becomes apparent because a normal newborn screen makes clinically important later loss unlikely.

B. Schedule tympanometry during routine visits because CMV-related hearing change is usually conductive and can be followed through middle-ear function.

C. Repeat a single diagnostic hearing test near one year of age because delayed CMV hearing loss is usually stable once detected after infancy.

D. Continue serial audiologic surveillance because CMV-associated sensorineural hearing loss can be delayed in onset or progressive for this case.

75. A preterm infant develops respiratory distress, temperature instability, and increasing oxygen need several hours after birth. Chest radiography shows diffuse infiltrates rather than the classic low-volume granular pattern of RDS. Which diagnosis should remain high on the differential?

A. Congenital or early-onset pneumonia, particularly when maternal infection risk factors or neonatal systemic signs are present in this setting.

B. Pulmonary interstitial emphysema, because interstitial air collections commonly develop before any exposure to positive-pressure ventilation.

C. Congenital diaphragmatic hernia, because diffuse bilateral infiltrates are the expected radiographic hallmark even with a normal abdominal contour.

D. Isolated transient tachypnea, because temperature instability and progressive oxygen need strongly favor benign delayed lung-fluid clearance.

76. A 40-week newborn requires positive-pressure ventilation at birth. There is no known cyanotic heart disease. What oxygen concentration is a reasonable starting point while pulse oximetry guides subsequent titration?

A. Start with 10% oxygen because transient neonatal cyanosis reflects fetal hemoglobin and does not require normal inspired oxygen levels.

B. Start with 21% oxygen and adjust according to the infant's response and age-specific preductal oxygen saturation targets.

C. Start with 100% oxygen in many term infants because room air is insufficient for transition during any assisted ventilation.

D. Start with 60% oxygen because fixed intermediate oxygen minimizes both hypoxemia and oxidative stress without titration.

77. A newborn has severe hypotonia, facial weakness, respiratory failure, and bilateral clubfeet. The mother has grip myotonia and difficulty releasing her hand after a handshake. Which diagnosis is most likely?

A. Prader-Willi syndrome because maternal myotonia predicts paternal chromosome 15 deletion and congenital respiratory failure.

B. Congenital myotonic dystrophy due to maternal transmission of an expanded DMPK repeat, often with severe neonatal respiratory and feeding weakness.

C. Spinal muscular atrophy because maternal grip myotonia is a characteristic carrier manifestation of biallelic SMN1 deletion.

D. Neonatal myasthenia because maternal myotonic dystrophy antibodies cross the placenta and cause temporary acetylcholine receptor blockade for this case.

78. A 10-day-old has vesicular skin lesions, lethargy, elevated transaminases, and seizures. Neonatal herpes simplex virus infection is suspected. What is the immediate treatment?

A. Treat vesicular lesions with topical acyclovir while awaiting blood and CSF testing because systemic therapy is reserved for proven disseminated or CNS disease.

B. Obtain surface cultures and PCR studies first, then start IV acyclovir after virologic confirmation if neurologic symptoms persist.

C. Start intravenous acyclovir promptly while obtaining diagnostic specimens because delayed therapy worsens outcomes in disseminated or CNS HSV disease.

D. Start oral acyclovir because enteral therapy achieves adequate CNS exposure in a hemodynamically stable neonate with suspected HSV.

79. A spontaneously breathing 29-week infant has mild-to-moderate RDS in the delivery room and does not require immediate intubation. Which initial support strategy is generally preferred?

A. Withhold distending pressure because CPAP increases alveolar collapse and should be avoided until after surfactant is given.

B. Use 100% oxygen by free flow without positive pressure because oxygen concentration alone corrects the low functional residual capacity of RDS.

C. Begin nasal CPAP and use selective surfactant if respiratory distress or oxygen need progresses in the neonatal setting described.

D. Intubate preterm infants prophylactically and provide prolonged mandatory ventilation regardless of initial respiratory effort.

80. A preterm infant receiving gentamicin develops rising creatinine and decreasing urine output. What medication principle is most important?

A. Reassess renal function and drug exposure, then adjust or hold nephrotoxic medication and dosing intervals according to pharmacokinetics and clinical need.

B. Add indomethacin to improve renal perfusion because cyclooxygenase inhibition reliably reverses aminoglycoside-associated kidney injury given these findings.

C. Increase gentamicin frequency because reduced urine output lowers renal elimination and therefore decreases drug exposure between doses.

D. Continue the same regimen without monitoring because neonatal aminoglycoside clearance is independent of kidney function.

81. A term infant of a mother with poorly controlled diabetes has respiratory distress and a systolic murmur. Echocardiography shows marked interventricular septal hypertrophy with dynamic left ventricular outflow obstruction. What is the most likely diagnosis?

A. Dilated cardiomyopathy from chronic fetal insulin deficiency causing a thin ventricular septum and severe systolic dilation.

B. Endocardial fibroelastosis caused by maternal diabetes, which characteristically produces isolated right ventricular hypertrophy.

C. Anomalous left coronary artery from the pulmonary artery, which typically presents at birth with septal hypertrophy from fetal hyperinsulinemia.

D. Transient hypertrophic cardiomyopathy associated with fetal hyperinsulinemia in an infant of a diabetic mother for this case.

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

A. 22q11.2 deletion syndrome, which is more strongly associated with conotruncal cardiac lesions, hypocalcemia, and thymic hypoplasia.

B. Trisomy 21, which is strongly associated with hypotonia and atrioventricular septal defects in the neonatal period for this infant.

C. Trisomy 18, which more often features clenched hands with overlapping fingers, severe growth restriction, and rocker-bottom feet.

D. Turner syndrome, which typically affects phenotypic females and is associated with lymphedema and left-sided obstructive heart disease.

83. A newborn has severe cyanosis and pulmonary edema. Echocardiography shows all pulmonary veins joining a confluence that drains below the diaphragm with marked venous obstruction. What is the priority?

A. Treat with diuretics alone for several weeks because anomalous pulmonary venous drainage usually resolves as pulmonary resistance falls in this newborn.

B. Urgent surgical correction is required because obstructed total anomalous pulmonary venous return causes life-threatening pulmonary venous hypertension.

C. Perform balloon pulmonary valvuloplasty because the primary lesion is obstruction at the pulmonary valve rather than the venous pathway.

D. Begin indomethacin and wait for ductal closure because reducing pulmonary blood flow reliably relieves the venous obstruction.

84. A healthy term newborn takes the first breaths and pulmonary blood flow rises rapidly. Which physiologic change is primarily responsible for this transition?

A. The first breaths increase alveolar pressure enough to compress pulmonary vessels and decrease flow through the lungs.

B. Loss of placental circulation lowers systemic vascular resistance, maintaining the fetal right-to-left ductal shunt after birth.

C. Lung expansion and increased oxygen tension lower pulmonary vascular resistance, promoting increased pulmonary blood flow.

D. Closure of the ductus venosus directly raises pulmonary vascular resistance and diverts blood away from the lungs.

85. A very preterm infant receiving positive-pressure ventilation develops worsening oxygenation. Chest radiography shows multiple linear and cystic lucencies extending through the lung interstitium. What is the most likely diagnosis?

A. Congenital pulmonary airway malformation, which generally presents as a fixed developmental cystic lesion rather than new ventilator-associated interstitial air.

B. Transient tachypnea caused by retained fetal lung fluid, which produces perihilar streaking without interstitial air collections.

C. Pulmonary edema from a PDA, which produces diffuse hazy opacities and vascular congestion rather than branching air lucencies.

D. Pulmonary interstitial emphysema caused by air dissecting from overdistended alveoli into the pulmonary interstitium in this case.

86. A screening test for neonatal sepsis has a sensitivity of 95%. What does this statistic mean?

A. The test reduces the risk of sepsis by 95% when used as part of a screening program.

B. Among infants who truly have sepsis, about 95% will have a positive test result here.

C. Among infants who do not have sepsis, about 95% will have a negative test result.

D. Among infants with a positive test, about 95% truly have sepsis regardless of disease prevalence.

87. Prenatal ultrasonography showed persistent hydronephrosis. The newborn is well and voiding normally. What is an appropriate postnatal management principle?

A. Start dialysis prophylactically because antenatal collecting-system dilation predicts progressive renal failure in most otherwise well newborns in this setting.

B. Arrange postnatal renal and bladder ultrasonography at an appropriate interval and tailor further studies to the severity and pattern of urinary tract dilation.

C. Perform immediate nephrectomy because persistent prenatal hydronephrosis usually represents a nonfunctioning kidney that cannot recover.

D. Obtain a voiding cystourethrogram before renal ultrasonography because prenatal hydronephrosis most often reflects vesicoureteral reflux.

88. A newborn has annular erythematous skin lesions and persistent complete heart block. The mother is asymptomatic but has anti-Ro/SSA antibodies. What is the unifying diagnosis?

A. Neonatal lupus caused by transplacental maternal autoantibodies, with transient cutaneous disease but potentially permanent cardiac conduction injury.

B. Severe combined immunodeficiency because absent T cells cause photosensitive annular rash and antibody-mediated complete heart block.

C. Congenital rubella because anti-Ro/SSA antibodies are viral antibodies that directly indicate maternal rubella infection during pregnancy for this infant.

D. Systemic juvenile idiopathic arthritis because maternal autoantibodies trigger neonatal arthritis and reversible complete heart block.

89. A nonvigorous term infant is born through meconium-stained fluid and is apneic. What is the best immediate airway approach?

A. Begin standard resuscitation with ventilation when indicated; perform suctioning only if secretions appear to obstruct effective ventilation.

B. Perform routine tracheal intubation and suction before any ventilation because meconium exposure alone requires airway clearance in this newborn.

C. Delay positive-pressure ventilation until the trachea has been repeatedly suctioned even when the heart rate is falling.

D. Avoid any airway intervention because meconium-stained fluid is not associated with airway obstruction or respiratory compromise.

90. A preterm infant coughs and desaturates repeatedly during oral feeds but remains stable between feeds. Which evaluation is most appropriate if the problem persists despite bedside feeding support?

A. Add acid-suppressive medication as the sole evaluation because coughing during feeds is diagnostic of gastroesophageal reflux disease.

B. Assess suck-swallow-breathe coordination and consider an instrumental swallowing study when aspiration is suspected in context.

C. Increase nipple flow rate substantially because faster milk delivery improves airway protection by shortening feeding time.

D. Stop oral-motor therapy because neonatal dysphagia cannot improve with maturation or individualized feeding strategies.

91. A 25-week infant has repeated cold stress despite adequate calories. Which response to cold is limited in extremely preterm infants compared with term newborns?

A. Shivering thermogenesis is the dominant neonatal heat source and is exaggerated in extreme prematurity, causing routine hyperthermia.

B. Evaporative heat loss is minimal in extremely preterm infants because their skin is thick and highly keratinized at birth.

C. Nonshivering thermogenesis is limited because preterm infants have less brown adipose tissue and a high surface-area-to-mass ratio.

D. Peripheral vasodilation is the main heat-conserving response in premature infants and becomes stronger as environmental temperature falls.

92. A 2-day-old develops sudden bilious vomiting and abdominal tenderness. The infant had previously fed well. What diagnosis must be excluded urgently?

A. Malrotation with midgut volvulus because vascular compromise can rapidly cause intestinal ischemia and necrosis given these findings.

B. Hypertrophic pyloric stenosis because gastric outlet obstruction commonly causes bilious emesis during the first 48 hours of life.

C. Physiologic gastroesophageal reflux because bilious vomiting is expected during normal intestinal adaptation in the newborn period.

D. Meconium plug syndrome because bilious emesis in newborns reflects transient distal colonic obstruction that can be observed safely here.

93. A term infant born to a mother with poorly controlled diabetes becomes jittery at 1 hour of life. Bedside glucose is low. Which mechanism most directly explains this infant's hypoglycemia?

A. Placental insulin crosses into the fetus and remains active for several hours after delivery, driving neonatal glucose uptake.

B. Maternal glucagon crosses the placenta in high concentrations and prevents hepatic glycogenolysis during the first postnatal hours.

C. Reduced fetal insulin secretion persists after birth because maternal hyperglycemia suppresses pancreatic beta-cell activity.

D. Persistent fetal hyperinsulinemia continues after the placental maternal glucose supply is abruptly interrupted at birth here.

94. A 10-day-old infant has vomiting, weight loss, dehydration, hyponatremia, hyperkalemia, and shock. Examination shows atypical genitalia. Which diagnosis is most likely?

A. Salt-wasting congenital adrenal hyperplasia, most commonly 21-hydroxylase deficiency with cortisol and aldosterone deficiency here.

B. Central diabetes insipidus, which produces hypernatremia and dilute urine rather than salt-wasting hyponatremia with hyperkalemia here.

C. Congenital hypothyroidism, which usually causes severe hyperkalemic dehydration and adrenal crisis during the second week of life.

D. Hyperinsulinemic hypoglycemia, which directly causes mineralocorticoid deficiency and virilization of a genetic female newborn.

95. In a cohort study, NEC occurs in 12% of formula-fed infants and 6% of human-milk-fed infants. What is the relative risk of NEC in the formula-fed group compared with the human-milk-fed group?

A. The relative risk is 1 divided by the 6% absolute difference, which equals about 16.7.

B. The relative risk is the absolute difference, 12% minus 6%, which equals 6.0.

C. The relative risk is 12% divided by 6%, which equals 2.0 in this clinical context.

D. The relative risk is 6% divided by 12%, which equals 0.5 for formula feeding compared with human milk.

96. An Rh-negative mother is highly alloimmunized and delivers a pale infant with anemia, jaundice, hepatosplenomegaly, and a positive direct antiglobulin test. What is the underlying mechanism?

A. Maternal IgM antibodies crossed the placenta and produced intravascular hemolysis because IgM is the predominant transplacental immunoglobulin.

B. Fetal autoantibodies against maternal erythrocytes caused maternal anemia and secondarily reduced fetal red-cell production.

C. Placental transfer of maternal complement proteins alone lysed fetal erythrocytes without any antigen-specific maternal antibody.

D. Maternal IgG antibodies crossed the placenta and destroyed fetal erythrocytes bearing the targeted red-cell antigen in context.

97. A 30-week infant is gaining weight but head circumference and length are crossing downward percentiles. How should growth be assessed?

A. Assess primarily birth-weight recovery because growth velocity after the birth weight is regained has no relation to nutritional adequacy.

B. Evaluate weight, length, and head circumference serially rather than using weight gain alone as proof of adequate nutrition in this setting.

C. Judge nutritional adequacy mainly by daily weight velocity because length and head circumference change too slowly to guide NICU nutrition decisions.

D. Use weight-for-age percentile alone because proportionality among weight, length, and head circumference is not needed for preterm growth assessment.

98. A 3-day-old becomes lethargic and poorly perfused as the ductus arteriosus closes. Femoral pulses are weak, upper-extremity blood pressure is higher than lower-extremity pressure, and metabolic acidosis is present. What should be done immediately?

A. Start prostaglandin E1 to restore ductal patency while urgent evaluation and definitive management of critical coarctation proceed.

B. Begin inhaled nitric oxide as the primary therapy because the pulse discrepancy indicates isolated pulmonary hypertension.

C. Give indomethacin to close any residual ductal flow because the ductus is worsening lower-body systemic perfusion.

D. Restrict fluids and observe because spontaneous ductal closure generally improves systemic blood flow in critical coarctation here.

99. A 1,100-g infant needs enteral feeding, but the mother's own milk supply is temporarily unavailable. Which alternative is generally preferred over preterm formula when an appropriate milk bank source is available?

A. Use preterm formula because its higher protein and mineral content makes it preferable to donor human milk when maternal milk is temporarily unavailable.

B. Pasteurized donor human milk can be used because it lowers exposure to bovine formula and is associated with a lower risk of NEC in very preterm infants.

C. Use standard term formula because donor milk is lower in protein and energy and therefore should be reserved for infants with cow-milk allergy.

D. Use extensively hydrolyzed preterm formula because hydrolysis provides the same NEC protection as donor human milk while improving nutrient density.

100. A 30-week infant loses 9% of birth weight over the first several days while urine output increases and serum sodium remains normal. The infant is otherwise stable. What is the best interpretation?

A. Any postnatal weight loss in a preterm infant suggests inadequate nutrition and requires immediate replacement of the lost weight with isotonic fluid boluses.

B. The finding indicates diabetes insipidus because physiologic neonatal transition should produce weight gain from extracellular fluid retention.

C. This can represent expected contraction of extracellular water during postnatal diuresis rather than pathologic dehydration.

D. The infant has renal failure because normal neonatal kidneys do not increase urine output during the first week after birth.

101. A newborn has an imperforate anus, a ventricular septal defect, a tracheoesophageal fistula, unilateral renal agenesis, and radial limb abnormality. Which diagnostic framework best fits?

A. Turner syndrome, which explains the renal anomaly and cardiac defect but not the characteristic tracheoesophageal and limb findings.

B. VACTERL association, defined by a nonrandom combination of vertebral, anal, cardiac, tracheoesophageal, renal, and limb anomalies.

C. CHARGE syndrome, which requires choanal atresia and coloboma as the primary explanation for this classic vertebral-anal-TE-limb pattern.

D. Prader-Willi syndrome, which produces neonatal hypotonia and feeding difficulty but not this cluster of structural malformations.

102. A 4-day-old develops a rapid bilirubin rise after an intercurrent illness. The infant has ancestry associated with G6PD deficiency and no obvious blood-group incompatibility. Why is G6PD deficiency important?

A. G6PD deficiency causes isolated conjugated hyperbilirubinemia by blocking bile-acid transport without affecting red-cell survival.

B. Oxidative stress can trigger abrupt hemolysis and severe unconjugated hyperbilirubinemia, sometimes with little warning from the early bilirubin trajectory.

C. The disorder protects neonatal erythrocytes from oxidative injury, so severe bilirubin elevation makes G6PD deficiency unlikely.

D. G6PD deficiency causes bilirubin elevation primarily after six months of age because fetal hemoglobin clinically prevents neonatal hemolysis in this setting.

103. A preterm infant receiving caffeine for apnea has stable clinical response and no signs of toxicity. Which pharmacokinetic feature helps explain once-daily maintenance dosing?

A. Caffeine is stored persistently in adipose tissue and released at a fixed daily rate independent of gestational age or renal function.

B. Caffeine is eliminated within minutes by mature neonatal hepatic enzymes, so once-daily dosing works primarily because the drug irreversibly binds respiratory receptors.

C. Caffeine undergoes complete first-pass destruction after enteral administration, so maintenance dosing depends on accumulation of its inactive metabolites.

D. Caffeine has a prolonged half-life in preterm infants because hepatic metabolism and renal elimination are immature in this clinical context.

104. A term newborn has diffuse petechiae and an intracranial hemorrhage. Platelet count is 12,000/microliter, but the mother has a normal platelet count and is healthy. Which diagnosis is most likely?

A. Hemophilia A, which causes severe thrombocytopenia from factor VIII deficiency and commonly produces diffuse petechiae at birth.

B. Fetal and neonatal alloimmune thrombocytopenia from maternal antibodies directed against a fetal platelet antigen inherited from the father.

C. Maternal immune thrombocytopenia, which usually produces severe maternal thrombocytopenia and neonatal platelet antigen incompatibility with the father.

D. Vitamin K deficiency bleeding, which causes isolated profound thrombocytopenia while coagulation studies remain normal.

105. A ventilated preterm infant with meningitis develops hyponatremia, low serum osmolality, inappropriately concentrated urine, and continued urine sodium excretion. Which disorder is most likely?

A. Syndrome of inappropriate antidiuretic hormone secretion causing water retention despite hypo-osmolality.

B. Physiologic postnatal diuresis causing appropriately dilute urine in response to low serum osmolality.

C. Central diabetes insipidus causing excessive free-water loss with dilute urine and rising serum sodium.

D. Primary hyperaldosteronism causing sodium retention and hypernatremia with suppressed urinary sodium excretion.

106. A growth-restricted infant is born after a pregnancy complicated by severe placental insufficiency. The hematocrit is 68%. Which prenatal process most likely contributed to this finding?

A. Reduced placental blood flow suppressed fetal erythropoietin, so the high hematocrit must represent postnatal hemoconcentration alone.

B. Maternal iron deficiency caused placental transfer of excess erythrocytes into the fetus, raising the neonatal hematocrit.

C. Placental insufficiency caused chronic fetal hyperoxia, which directly increased marrow erythropoiesis despite suppressed erythropoietin.

D. Chronic fetal hypoxemia increased erythropoietin production and stimulated red-cell mass before delivery given these findings.

107. A preterm infant receives minimal enteral feeds while most calories are parenteral. What is one physiologic purpose of small enteral volumes when clinically appropriate?

A. They sterilize the intestinal lumen because human milk suppresses bacterial colonization during the first weeks of life.

B. They replace the need for parenteral amino acids immediately because even tiny milk volumes provide complete protein needs in extreme prematurity for this case.

C. They provide luminal stimulation that supports gastrointestinal maturation while full nutritional requirements are still met largely by parenteral nutrition.

D. They eliminate the risk of feeding intolerance because trophic volumes strongly predict normal intestinal motility in preterm infants.

108. A newborn's serum creatinine is elevated on the first day of life, but urine output is acceptable and the value falls over the next several days. Which explanation is most appropriate?

A. A falling creatinine after birth indicates worsening renal function because healthy neonatal kidneys should maintain the maternal level for several weeks here.

B. The elevated value suggests congenital renal failure because maternal creatinine does not cross the placenta during pregnancy.

C. Creatinine is produced primarily by the placenta before birth, so neonatal serum levels are unrelated to either maternal or infant renal function.

D. Early neonatal creatinine partly reflects maternal creatinine transferred across the placenta, so serial trends are more informative than a single birth value.

109. A term newborn has marked hypotonia, a weak cry, poor suck, and little spontaneous movement. Several months later, the major concern would be development of hyperphagia and obesity. Which diagnosis is most likely?

A. Achondroplasia, which causes disproportionate short stature but not the characteristic sequence of neonatal hypotonia followed by hyperphagia.

B. Turner syndrome, which causes lymphedema and short stature but is not associated with profound neonatal hypotonia and later hyperphagia.

C. Beckwith-Wiedemann syndrome, which presents with neonatal overgrowth and macroglossia followed by progressive severe hyperphagia in infancy.

D. Prader-Willi syndrome, which often presents with severe neonatal hypotonia and feeding difficulty before later hyperphagia emerges here.

110. A term newborn develops recurrent focal clonic movements of the right arm at 12 hours of age. MRI shows an acute left middle cerebral artery infarction. What is the most likely diagnosis?

A. Perinatal arterial ischemic stroke, which often presents in the neonatal period with focal seizures despite otherwise subtle examination findings.

B. Periventricular leukomalacia, which produces acute focal clonic seizures from a unilateral cortical arterial occlusion in the first day of life here.

C. Benign neonatal sleep myoclonus, which occurs primarily during sleep and is accompanied by a focal arterial infarct on MRI.

D. Germinal-matrix hemorrhage, which is the typical cause of unilateral cortical middle cerebral artery infarction in healthy term newborns.

111. A term newborn becomes visibly jaundiced at 10 hours of life. Which interpretation is most appropriate?

A. This is classic physiologic jaundice because normal bilirubin peaks during the first 12 hours and then falls steadily through day 3.

B. Breast-milk jaundice is the most likely cause because it characteristically begins before the first feeding and peaks during the first day in this clinical context.

C. Jaundice in the first 24 hours is pathologic until proven otherwise and warrants prompt bilirubin measurement and evaluation for hemolysis or other disease.

D. Repeat a visual jaundice assessment after the next feed and defer serum testing unless the infant becomes more yellow or develops anemia.

112. A preterm infant has persistent non-anion-gap metabolic acidosis, poor growth, and inappropriately high urinary bicarbonate losses while serum bicarbonate is low. Which renal process is most likely?

A. Renal vein thrombosis selectively blocks distal hydrogen secretion and therefore typically presents as isolated hyperkalemic acidosis without hematuria.

B. Central diabetes insipidus causes bicarbonate wasting by suppressing antidiuretic hormone, making metabolic acidosis its defining feature.

C. Respiratory alkalosis is the likely cause because renal bicarbonate loss raises serum bicarbonate and produces a high anion gap.

D. Proximal renal tubular bicarbonate wasting can produce a normal-anion-gap metabolic acidosis in an immature or diseased kidney here.

113. A septic preterm infant has cool extremities, prolonged capillary refill, oliguria, and rising lactate. Which interpretation best fits these findings?

A. They are expected findings of normal preterm transition because capillary refill and urine output do not reflect neonatal perfusion.

B. They indicate inadequate systemic perfusion and evolving shock even if the blood pressure has not yet fallen below the gestational-age reference range.

C. They indicate isolated pulmonary overcirculation because systemic shock in neonates cannot occur until severe hypotension is documented.

D. They suggest cardiogenic shock from congenital heart disease because distributive and hypovolemic shock do not elevate lactate in newborns for this infant.

114. A healthy newborn has repetitive jerking of both arms only during sleep. The movements stop immediately when the infant awakens, and EEG during events shows no seizure activity. What is the most likely diagnosis?

A. Jitteriness from hypoglycemia, which occurs exclusively during sleep and is not influenced by stimulation or restraint of the limb.

B. Neonatal epilepsy, because any repetitive neonatal movement during sleep represents electrographic seizures even when EEG remains normal.

C. Hyperekplexia, which is defined by rhythmic sleep-primarily jerking that stops with arousal and generally does not includes exaggerated startle or stiffness.

D. Benign neonatal sleep myoclonus, a nonepileptic condition in which movements occur during sleep and cease with arousal in this newborn.

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

A. Noonan syndrome, which more commonly features pulmonary stenosis and a webbed neck than macroglossia and neonatal hyperinsulinism.

B. Trisomy 18, which is associated with prenatal growth restriction and micrognathia rather than macrosomia and omphalocele.

C. Beckwith-Wiedemann syndrome, an overgrowth disorder associated with macroglossia, abdominal wall defects, hyperinsulinemic hypoglycemia, and embryonal tumor risk.

D. Prader-Willi syndrome, which typically causes severe neonatal hypotonia and poor feeding rather than prenatal overgrowth and macroglossia in the situation described.

116. A rapidly growing preterm infant is receiving primarily human milk. Which micronutrient commonly requires supplementation to reduce the risk of iron deficiency after the early neonatal period?

A. Copper should replace iron because human milk contains excessive iron that causes oxidative injury in premature infants.

B. Folate supplementation should replace iron because rapid erythropoiesis in preterm infants is limited primarily by folate rather than by iron stores.

C. Vitamin E supplementation should replace iron because antioxidant support prevents depletion of neonatal iron during rapid red-cell production.

D. Iron supplementation is commonly needed because preterm infants have smaller iron stores and rapid postnatal erythropoietic and growth demands.

117. An infant born after severe maternal preeclampsia has thrombocytopenia on the first day of life but is otherwise clinically stable. Which mechanism is most consistent with this association?

A. Maternal hypertension causes transplacental passage of platelet-specific alloantibodies in affected pregnancies here.

B. Preeclampsia routinely produces neonatal disseminated intravascular coagulation even when the infant is well appearing.

C. Placental insufficiency can impair fetal megakaryopoiesis and is associated with early neonatal thrombocytopenia in context.

D. Maternal antihypertensive therapy destroys neonatal platelets by direct immune-mediated toxicity after placental transfer here.

118. A former 28-week infant is 8 months old chronologically and is being assessed for early developmental milestones. Which age is most appropriate for interpreting developmental expectations during the first two years?

A. Use chronological age without correction because developmental norms are interpreted from time since birth even in former preterm infants.

B. Use corrected age, subtracting the degree of prematurity from chronological age, while also tracking the child's individual trajectory here.

C. Use gestational age at birth as a fixed developmental reference because milestone timing should remain anchored to the original gestation.

D. Use postmenstrual age rather than corrected age because the sum of gestational and postnatal age is the preferred milestone scale after discharge.

119. A newborn has coloboma, a complex congenital heart defect, choanal atresia, growth restriction, genital hypoplasia, and abnormal external ears. Which syndrome is most likely?

A. VACTERL association, which centers on vertebral, anal, cardiac, tracheoesophageal, renal, and limb anomalies rather than coloboma and choanal atresia for this case.

B. Noonan syndrome, which can include pulmonary stenosis and webbed neck but does not characteristically combine coloboma with choanal atresia.

C. Beckwith-Wiedemann syndrome, which features overgrowth, macroglossia, abdominal wall defects, and neonatal hypoglycemia rather than choanal atresia.

D. CHARGE syndrome, a pattern that includes coloboma, heart disease, choanal atresia, growth and developmental abnormalities, genital anomalies, and ear findings.

120. Why are extended dosing intervals often used for gentamicin in premature infants?

A. Gentamicin has time-dependent killing that requires a constant low serum concentration, making frequent small doses safer than interval-based dosing.

B. Gentamicin is cleared primarily by the liver, and premature hepatic metabolism is faster than adult metabolism, requiring longer intervals to prevent low peaks here.

C. The drug accumulates primarily in red blood cells, so dosing interval is determined by hematocrit rather than kidney function or gestational age.

D. Renal clearance is reduced and variable, so longer intervals can allow adequate drug elimination while preserving concentration-dependent antibacterial activity.

121. The same neonatal screening test is used in a high-risk NICU population and in a low-risk well-baby nursery. Sensitivity and specificity are unchanged. Which measure is most likely to be higher in the high-risk population?

A. Positive predictive value generally rises as disease prevalence increases, assuming test characteristics otherwise remain similar.

B. Negative predictive value must typically rise with prevalence because a negative result becomes more reassuring when disease is common.

C. Specificity must fall because prevalence directly changes the fraction of unaffected infants who test negative.

D. Sensitivity must rise because prevalence directly changes the fraction of affected infants who test positive.

122. A newborn has weak femoral pulses but normal brachial pulses. Which bedside finding would further support coarctation of the aorta?

A. Equally strong pulses in all four extremities during shock because coarctation does not affect pulse amplitude in the neonatal period.

B. A fixed wide pulse pressure in the legs with bounding femoral pulses because distal runoff is the defining physiology of coarctation.

C. A higher systolic blood pressure in the upper extremities than in the lower extremities, especially as the ductus constricts.

D. A lower right-arm pressure than right-leg pressure because coarctation selectively reduces perfusion of the upper body.

123. A newborn has congenital heart disease, renal anomalies, abnormal ears, and limb asymmetry without a recognizable single syndrome on examination. Which genetic test is often an appropriate first-line study for multiple unexplained congenital anomalies?

A. A standard metabolic panel replaces genomic testing because structural anomalies are rarely caused by chromosomal copy-number changes.

B. Chromosomal microarray can detect clinically significant copy-number gains and losses not visible on a routine karyotype for this infant.

C. Obtain a conventional karyotype as the sole initial test because it detects most clinically significant submicroscopic copy-number changes causing multisystem anomalies.

D. Use targeted sequencing for the most prominent organ abnormality first because genome-wide testing has low yield when more than one system is involved.

124. A 5-week-old has progressive projectile nonbilious vomiting, remains hungry after emesis, and has a hypochloremic metabolic alkalosis. Which diagnosis is most likely?

A. Malrotation with volvulus, which usually causes bilious vomiting and acute abdominal compromise rather than progressive nonbilious emesis.

B. Hirschsprung disease, which presents primarily with distal bowel obstruction, delayed meconium passage, and abdominal distention.

C. Hypertrophic pyloric stenosis causing gastric outlet obstruction proximal to the duodenum when the findings are considered together.

D. Necrotizing enterocolitis, which causes systemic illness, bloody stool, and pneumatosis rather than isolated projectile nonbilious vomiting.

125. A previously well infant becomes lethargic and hypoglycemic during a viral illness after an overnight fast. Ketones are unexpectedly low and liver enzymes are mildly elevated. Which disorder is most likely?

A. Medium-chain acyl-CoA dehydrogenase deficiency causing impaired fatty-acid oxidation and hypoketotic hypoglycemia during fasting stress.

B. Congenital hypothyroidism, which characteristically produces severe hypoketotic hypoglycemia primarily during short fasting periods.

C. Classic phenylketonuria, which causes fasting-induced hypoglycemia because phenylalanine cannot be converted directly into ketone bodies.

D. Galactosemia, which is triggered by absence of milk rather than milk exposure and therefore becomes most severe during an overnight fast.

126. A term infant had difficult delivery and develops firm violaceous subcutaneous nodules over the back and shoulders. Two weeks later the infant is irritable and has poor feeding. Which metabolic complication should be checked?

A. Severe hypophosphatemia is the defining metabolic complication because fat necrosis consumes circulating phosphate without affecting calcium.

B. Hypernatremia is expected because subcutaneous fat necrosis directly causes central diabetes insipidus through hypothalamic inflammation.

C. Hypercalcemia can follow subcutaneous fat necrosis and may emerge days to weeks after the skin lesions appear in the situation described.

D. Persistent hypoglycemia is the characteristic delayed complication because necrotic adipocytes release excess insulin into the bloodstream.

127. A 30-week infant has recurrent apnea with bradycardia and desaturation after infection, anemia, hypoglycemia, and seizures have been excluded. Which medication is first-line for apnea of prematurity?

A. Caffeine citrate because methylxanthine therapy stimulates respiratory drive and reduces clinically important apnea in preterm infants.

B. Morphine because lowering respiratory effort prevents unstable breathing and decreases the frequency of central apnea episodes in this case.

C. Furosemide because pulmonary diuresis is the primary treatment for central apnea when chest radiography is normal.

D. Propranolol because reducing sympathetic tone improves immature medullary respiratory rhythm generation in premature infants.

128. A 24-week infant has a sudden fall in hematocrit and increased apnea on day 3. Cranial ultrasonography shows blood originating in the caudothalamic groove and extending into the ventricles. What is the most likely diagnosis?

A. Germinal-matrix intraventricular hemorrhage, a complication of extreme prematurity related to fragile periventricular vasculature and unstable cerebral blood flow.

B. Periventricular leukomalacia, which is primarily a white-matter ischemic injury and does not present as acute intraventricular blood on ultrasound.

C. Congenital CMV, which causes periventricular calcifications that are sonographically indistinguishable from acute intraventricular hemorrhage.

D. Arterial ischemic stroke, which most commonly begins in the caudothalamic germinal matrix and produces bilateral ventricular blood in extremely preterm infants here.

129. A newborn has hepatosplenomegaly, snuffles, rash, and long-bone abnormalities. Maternal syphilis was untreated. Which neonatal serologic approach is appropriate?

A. Use primarily a neonatal treponemal IgG test because passively transferred maternal treponemal antibody disappears within hours of birth here.

B. Do not test the infant because congenital syphilis can be diagnosed primarily by maternal serology and neonatal examination has no role.

C. Obtain a quantitative nontreponemal test such as RPR or VDRL on the infant's serum and compare it with the maternal titer using the same test.

D. Use umbilical cord blood as the preferred specimen because it cannot be contaminated with maternal blood or affected by Wharton jelly.

130. A critically ill septic neonate develops oozing from venipuncture sites, thrombocytopenia, prolonged PT and aPTT, low fibrinogen, and elevated D-dimer. What is the most likely diagnosis?

A. Hemophilia A, which causes isolated aPTT prolongation rather than thrombocytopenia, low fibrinogen, and elevated fibrin degradation products.

B. Fetal and neonatal alloimmune thrombocytopenia, which causes isolated platelet destruction with normal coagulation factors and fibrinogen.

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

D. Vitamin K deficiency alone, which typically lowers fibrinogen and causes marked thrombocytopenia through systemic microvascular clot formation.

131. A term newborn is profoundly pale and tachycardic at birth with hemoglobin 6 g/dL, minimal jaundice, and a negative direct antiglobulin test. A maternal Kleihauer-Betke-type assessment suggests a large fetal-to-maternal bleed. Which diagnosis best fits?

A. Vitamin K deficiency bleeding, which causes prolonged coagulation studies but does not explain severe anemia present immediately at birth without overt bleeding.

B. ABO hemolytic disease, which causes antibody-mediated red-cell destruction and usually produces a positive direct antiglobulin test with hyperbilirubinemia.

C. Polycythemia from placental transfusion, which increases hemoglobin and produces hyperviscosity rather than profound anemia and pallor.

D. Massive fetomaternal hemorrhage causing acute or chronic fetal blood loss and severe neonatal anemia given the clinical findings described.

132. Monochorionic diamniotic twins show oligohydramnios and growth restriction in one sac, while the co-twin has polyhydramnios and cardiomegaly. Which diagnosis best explains this pattern?

A. Twin anemia-polycythemia sequence caused by a large acute placental hemorrhage that usually produces identical amniotic-fluid volumes.

B. Dichorionic selective growth restriction caused by two independent placentas with no shared vascular connections between the fetuses.

C. Maternal diabetes causing symmetric fetal overgrowth in one twin and isolated renal agenesis with oligohydramnios in the other.

D. Twin-twin transfusion syndrome from unbalanced placental vascular connections, producing a donor and a volume-overloaded recipient.

133. A 3-week-old infant is jaundiced with pale stools and dark urine. Direct bilirubin is elevated. What is the most important interpretation?

A. This is typical breast-milk jaundice because breast milk commonly causes pale stools and isolated elevation of direct bilirubin.

B. Conjugated hyperbilirubinemia is never physiologic and requires prompt evaluation for cholestasis, including biliary obstruction and hepatocellular disease.

C. Manage as prolonged breast-milk jaundice and repeat fractionated bilirubin in several weeks because pale stools can occur with benign unconjugated hyperbilirubinemia.

D. Use the total bilirubin phototherapy threshold to determine urgency because a low total bilirubin makes clinically important cholestasis unlikely.

134. A fetus has severe prolonged oligohydramnios after very early preterm premature rupture of membranes. Which neonatal respiratory problem is the most important anticipated consequence?

A. Surfactant excess is expected because prolonged membrane rupture accelerates type II pneumocyte secretion beyond normal levels.

B. Congenital lobar overinflation is expected because reduced amniotic fluid causes focal bronchial cartilage weakness.

C. Pulmonary hypoplasia can result because prolonged severe oligohydramnios interferes with normal lung growth and expansion.

D. Transient tachypnea is expected because oligohydramnios causes excessive fetal lung fluid that remains after birth.

135. A very preterm infant becomes excessively sedated and hypoventilates after repeated morphine doses. Which pharmacokinetic principle increases this risk?

A. Morphine cannot cross the neonatal blood-brain barrier, so respiratory depression implies an unrelated pulmonary disorder rather than opioid effect.

B. Immature hepatic metabolism and renal clearance can prolong opioid and metabolite exposure, so dosing must be individualized and closely monitored.

C. Renal function has no effect on morphine metabolites, making gestational age and kidney injury irrelevant to neonatal opioid toxicity.

D. Preterm infants clear morphine much faster than adults, so sedation after repeated dosing indicates opioid withdrawal rather than drug accumulation.

136. A term small-for-gestational-age infant is plethoric, lethargic, hypoglycemic, and has a venous hematocrit of 72%. Which intervention may be appropriate if symptoms are attributed to hyperviscosity?

A. Partial exchange transfusion with an appropriate replacement fluid may be considered for symptomatic severe neonatal polycythemia for this infant.

B. Packed red-cell transfusion is indicated because increasing hematocrit improves microvascular flow and corrects hyperviscosity-related hypoglycemia.

C. Platelet transfusion is the definitive therapy because neonatal hyperviscosity is caused primarily by thrombocytopenia rather than red-cell mass.

D. Provide IV hydration and repeat the hematocrit after several hours without exchange because symptoms of hyperviscosity usually resolve with volume expansion alone.

137. A 28-week infant with a long-standing central line develops temperature instability and apnea at 4 weeks of age. Blood cultures repeatedly grow coagulase-negative staphylococci. Which interpretation is most appropriate?

A. The finding suggests bacterial meningitis because coagulase-negative staphylococci cause CSF infection in most neonates with a positive blood culture.

B. Coagulase-negative staphylococci can cause true late-onset bloodstream infection in preterm infants with intravascular devices, especially when cultures and symptoms are consistent.

C. The organism is typically a contaminant because coagulase-negative staphylococci cannot cause invasive disease in neonates with central venous catheters in the neonatal setting described.

D. The infection must have been acquired transplacentally because late-onset sepsis cannot arise from skin flora after several weeks in the NICU.

138. A newborn develops jaundice, hepatomegaly, vomiting, poor feeding, and Escherichia coli sepsis after milk feeds are started. Which metabolic disorder should be suspected?

A. Maple syrup urine disease, which is characterized by galactose accumulation, cataracts, and gram-negative sepsis after milk exposure.

B. MCAD deficiency, which usually presents with hepatomegaly and E. coli sepsis primarily while the infant is continuously fed and generally does not fasting.

C. Classic galactosemia from galactose-1-phosphate uridyltransferase deficiency, requiring prompt removal of galactose and lactose from the diet.

D. Phenylketonuria, which typically causes acute hepatic failure and E. coli sepsis immediately after lactose-containing feeds.

139. A hemodynamically stable 29-week infant has no signs of intestinal disease. Which feeding strategy is generally reasonable rather than keeping the infant completely NPO for a prolonged period?

A. Rely on parenteral nutrition until the infant reaches a more mature postmenstrual age because early enteral exposure increases intestinal oxygen demand.

B. Begin a hydrolyzed preterm formula rather than human milk because peptide-based feeds are better tolerated by the immature intestine during initial advancement.

C. Begin small enteral feeds and advance as tolerated while monitoring the infant's overall clinical status and abdominal findings in this case.

D. Withhold enteral nutrition until 34 weeks postmenstrual age because premature intestine cannot safely process milk before that point.

140. A term newborn has marked abdominal distention and has not passed meconium by 48 hours. Rectal examination produces an explosive release of stool and gas. Which diagnosis is most likely?

A. Necrotizing enterocolitis, which usually presents at birth in healthy term infants with an empty rectum and explosive stool after examination.

B. Hirschsprung disease from absence of enteric ganglion cells in the distal bowel, causing functional obstruction for this case.

C. Gastroschisis, which causes delayed meconium passage because the bowel is enclosed within a membrane at the umbilical ring.

D. Pyloric stenosis from hypertrophy of the pyloric muscle, which causes distal colonic obstruction and failure to pass meconium at birth.

141. Why are very preterm infants particularly susceptible to invasive bacterial and fungal infection even when no specific primary immunodeficiency is present?

A. Adaptive immunity is fully mature at 24 weeks, but infection risk rises primarily because incubators expose infants to more environmental oxygen.

B. Preterm infants have excessive complement activation that destroys antibodies and makes encapsulated bacteria resistant to opsonization.

C. Multiple innate and adaptive defenses are immature, including skin and mucosal barriers, neutrophil reserve and function, complement activity, and passive antibody levels.

D. Prematurity causes complete absence of leukocytes until term-equivalent age, so infection risk is determined primarily by transient neonatal leukopenia in this setting here.

142. A 38-week infant delivered by elective cesarean without labor develops tachypnea soon after birth. Oxygen need is mild, and radiography shows hyperinflation, perihilar streaking, and fluid in the fissures. What is the most likely diagnosis?

A. Respiratory distress syndrome caused by severe surfactant deficiency, which usually produces low lung volumes and diffuse granular opacities.

B. Pulmonary hypoplasia from oligohydramnios, which typically presents with hyperinflated lungs and rapid spontaneous resolution.

C. Congenital pneumonia with septic shock, which is expected to cause primarily fissural fluid without systemic signs.

D. Transient tachypnea of the newborn caused by delayed clearance of fetal lung fluid in the neonatal setting described.

143. A newborn has hydrocephalus, diffuse intracranial calcifications, and chorioretinitis. Which congenital infection classically causes this triad?

A. Congenital CMV, which is more classically associated with periventricular calcifications and sensorineural hearing loss than the classic toxoplasmosis triad.

B. Perinatal hepatitis B, which is usually asymptomatic in the newborn and does not characteristically cause intracranial calcifications.

C. Congenital toxoplasmosis, which can produce chorioretinitis, hydrocephalus, and intracranial calcifications for the presentation described.

D. Congenital syphilis, which commonly causes snuffles, hepatosplenomegaly, rash, and bone lesions rather than hydrocephalus with chorioretinitis.

144. A clinically stable 25-week infant has a patent ductus arteriosus detected on screening echocardiography at 5 days of age but no clear signs of hemodynamic compromise. Which approach is most consistent with current evidence?

A. Arrange immediate surgical ligation because early procedural closure prevents bronchopulmonary dysplasia in otherwise stable preterm infants.

B. Use expectant clinical and echocardiographic assessment rather than routine prophylactic or early drug closure solely because the PDA is present.

C. Start prostaglandin E1 because maintaining a larger PDA decreases pulmonary blood flow and protects the immature lungs from overcirculation.

D. Give ibuprofen to extremely preterm infants with a PDA before two weeks because universal early closure improves survival and neurodevelopment here.

145. A term infant with prenatal opioid exposure is irritable, tremulous, difficult to console, and has loose stools but remains able to feed and sleep with support. What is the most appropriate initial management principle?

A. Begin scheduled naloxone because opioid antagonism is the preferred first-line treatment for withdrawal in a spontaneously breathing newborn in this case.

B. Separate the infant from the parent and provide a brightly stimulating nursery environment to reduce reinforcement of withdrawal behaviors.

C. Start phenobarbital immediately in opioid-exposed newborns because autonomic symptoms predict neonatal seizures without treatment.

D. Optimize nonpharmacologic care such as rooming-in, low stimulation, swaddling, feeding support, and functional assessment before medication when safe.

146. A newborn with a history of prolonged severe oligohydramnios has small lungs, severe hypoxemia, and difficult ventilation immediately after birth. Which underlying abnormality is most likely?

A. Pulmonary hypoplasia with reduced airway, alveolar, and pulmonary vascular development for this case.

B. Isolated delayed lung-fluid clearance with normal lung size and rapid resolution over the first day.

C. Congenital lobar overinflation involving a single lobe with compression of otherwise normally developed lungs.

D. Excessive surfactant production with abnormally high lung compliance and alveolar overexpansion.

147. A 26-week infant develops abdominal distention, bloody stools, metabolic acidosis, and pneumatosis intestinalis. Which diagnosis is most likely?

A. Pyloric stenosis, which produces nonbilious projectile vomiting and hypochloremic alkalosis rather than bloody stool and intestinal pneumatosis.

B. Hirschsprung disease, which causes distal functional obstruction but does not characteristically produce pneumatosis with systemic deterioration in a very preterm infant.

C. Necrotizing enterocolitis, an inflammatory-ischemic intestinal disease of prematurity with pneumatosis as a characteristic radiographic finding.

D. Spontaneous intestinal perforation, which typically produces an isolated perforation without the diffuse intestinal inflammation and pneumatosis described here.

148. A male newborn has a poor urinary stream, a distended bladder, bilateral hydronephrosis, and rising creatinine. Which diagnosis is most likely?

A. Ureteropelvic junction obstruction, which usually causes isolated upper-tract dilation without a distended bladder or weak urinary stream.

B. Multicystic dysplastic kidney, which usually produces a nonfunctioning unilateral cystic kidney rather than bilateral hydronephrosis from bladder outlet obstruction.

C. Posterior urethral valves causing congenital bladder outlet obstruction and potentially severe obstructive nephropathy.

D. Renal vein thrombosis, which produces hematuria and renal enlargement rather than lower urinary tract obstruction.

149. Parents with limited English proficiency must consent to a high-risk neonatal procedure. A bilingual adolescent sibling offers to interpret. What is the best approach?

A. Use a qualified medical interpreter and communicate directly with the parents so informed consent is accurate, private, and family centered.

B. Use the adolescent sibling because family members are more accurate than professional interpreters for complex medical risk discussions.

C. Use the bilingual sibling for the consent discussion because family interpreters can preserve nuance and reduce delays when the procedure is high risk.

D. Provide written translated material and obtain the parent's signature without a live interpreter because the document itself supplies the necessary informed-consent content.

150. A former extremely preterm toddler has a developmental assessment score within the average range at 18 months corrected age. How should this result be interpreted?

A. It strongly predicts normal long-term neurodevelopment because standardized toddler testing has perfect predictive accuracy for school-age function.

B. It suggests the child no longer needs hearing, vision, growth, or developmental surveillance after the two-year visit.

C. It is reassuring for current performance but does not guarantee normal later executive, language, motor, behavioral, or school-age outcomes.

D. It is uninterpretable because standardized developmental testing cannot be used in children born preterm under any circumstances.

151. A 6-week-old infant has fever and meningitis caused by group B Streptococcus. The mother received appropriate intrapartum GBS prophylaxis. Which statement is most accurate?

A. Intrapartum antibiotic prophylaxis reduces early-onset GBS disease but does not reliably prevent late-onset GBS infection for this infant.

B. Appropriate intrapartum prophylaxis eliminates both early- and late-onset GBS disease, so the culture result must represent contamination.

C. Maternal prophylaxis prevents meningitis but not bacteremia, so a CSF-positive late-onset infection is incompatible with true GBS disease.

D. Late-onset GBS occurs primarily in infants whose mothers had negative prenatal cultures and generally does not in infants exposed to intrapartum antibiotics.

152. A pregnant patient at 29 weeks has preterm labor and a high likelihood of delivery within the next several days. Which antenatal intervention most directly reduces neonatal respiratory distress syndrome and other complications of prematurity?

A. Schedule elective cesarean delivery immediately because avoiding labor is more effective than antenatal medication for lung maturation.

B. Give maternal loop diuretics to reduce fetal lung fluid and thereby prevent surfactant deficiency after premature birth.

C. Administer a course of antenatal corticosteroids because fetal lung maturation can be accelerated when preterm birth is imminent.

D. Begin maternal indomethacin primarily to stimulate fetal surfactant production, even if uterine contractions have already stopped.

153. A preterm infant with a tension pneumothorax improves immediately after emergency needle decompression but still has a large persistent pleural air collection and ongoing ventilator requirement. What is the next step?

A. Place a chest tube or other definitive pleural drain to maintain evacuation of the ongoing air leak for the presentation described.

B. Give surfactant into the pleural cavity because exogenous phospholipid closes the visceral pleural defect after needle decompression.

C. Administer inhaled nitric oxide as definitive pleural drainage because pulmonary vasodilation removes air from the pleural space.

D. Remove respiratory support immediately because decompression persistently seals the alveolar leak and prevents recurrence.

154. A term infant with meconium aspiration has severe hypoxemia. The right-hand oxygen saturation is 94%, while a foot saturation is 82%. Echocardiography shows right-to-left shunting across the ductus arteriosus. What is the most likely diagnosis?

A. Uncomplicated patent ductus arteriosus with a left-to-right shunt causing systemic desaturation primarily below the diaphragm.

B. Transposition of the great arteries with complete mixing, which typically produces a large persistent preductal-postductal saturation gradient.

C. Isolated ventricular septal defect causing preferential desaturation of postductal blood while preductal saturation remains normal.

D. Persistent pulmonary hypertension of the newborn causing elevated pulmonary vascular resistance and extrapulmonary right-to-left shunting.

155. A newborn has rhizomelic limb shortening, macrocephaly with frontal bossing, and a relatively narrow foramen magnum. Which molecular mechanism most commonly causes this condition?

A. A loss-of-function CFTR variant that disrupts chloride transport and causes shortened long bones from meconium obstruction.

B. A 22q11.2 deletion that impairs pharyngeal development and directly causes rhizomelic dwarfism with frontal bossing.

C. An activating FGFR3 variant that impairs endochondral bone growth in achondroplasia given the clinical findings described.

D. A COL1A1 or COL1A2 variant that reduces collagen integrity and produces the typical stable skull-base anatomy of achondroplasia.

156. A newborn of a mother with myasthenia gravis has a weak cry, poor suck, facial weakness, and shallow respirations shortly after birth. Which mechanism best explains this neonatal presentation?

A. Maternal IgG against acetylcholine-receptor targets crossed the placenta and transiently impaired neuromuscular transmission in the newborn in context.

B. A de novo SMN1 disorder caused acute loss of anterior horn cells specifically because maternal myasthenia increased fetal motor-neuron vulnerability.

C. Maternal acetylcholinesterase inhibitors crossed the placenta and permanently destroyed fetal motor end plates, producing irreversible congenital weakness.

D. A fetal botulinum infection blocked acetylcholine release in utero, a common consequence of maternal autoimmune neuromuscular disease during pregnancy.

157. A term infant with severe parenchymal lung disease has persistent pulmonary hypertension and an oxygenation index that remains high despite adequate lung recruitment and ventilation. Which targeted therapy may improve oxygenation?

A. Routine inhaled nitric oxide is recommended for extremely preterm infants with respiratory distress, regardless of evidence of pulmonary hypertension here here.

B. Inhaled nitric oxide can selectively lower pulmonary vascular resistance in appropriate term or near-term infants with hypoxic respiratory failure and PPHN.

C. Indomethacin is the preferred pulmonary vasodilator because cyclooxygenase inhibition selectively relaxes the neonatal pulmonary circulation.

D. Systemic phenylephrine should be used alone because increasing systemic pressure eliminates the need to treat elevated pulmonary vascular resistance.

158. A 25-week infant cannot yet receive enough enteral nutrition to meet metabolic needs. Which parenteral nutrition principle is most appropriate?

A. Use primarily intravenous saline because premature infants meet their energy needs through hepatic glycogen stores during the first weeks.

B. Provide adequate amino acids, glucose, lipids, electrolytes, vitamins, and trace elements while advancing enteral feeds as tolerated here.

C. Provide dextrose and lipids initially while delaying amino acids until enteral feeds are established because early protein delivery increases azotemia without improving growth.

D. Provide glucose and amino acids but defer lipids for the first several weeks because neonatal fat oxidation is too immature for parenteral lipid use.

159. Which immunologic component of human milk contributes most directly to mucosal defense in the neonatal gastrointestinal tract?

A. Secretory IgA coats mucosal surfaces and helps limit pathogen adherence and invasion without provoking intense inflammation in context.

B. Maternal IgM crosses the intestinal epithelium intact and becomes the main long-term serum immunoglobulin of the breast-fed infant.

C. Complement C9 in milk enters the neonatal bloodstream in high concentration and persistently replaces immature endogenous complement production.

D. Maternal neutrophils from milk engraft in neonatal marrow and provide the infant's circulating granulocyte population for the first year.

160. A 1,200-g infant on fortified human milk has poor linear growth despite adequate measured calories. Which nutritional issue deserves particular attention?

A. Carbohydrate should be eliminated because glucose is the principal cause of poor length gain in otherwise stable preterm infants.

B. Protein should be reduced because linear growth failure in preterm infants usually reflects excessive amino acid exposure rather than deficiency here.

C. Fortification should be stopped because human milk protein becomes toxic once an infant reaches full enteral feeding volumes.

D. Protein intake may be inadequate even when total calories appear sufficient, so nutrient composition should be reviewed rather than calories alone.

161. A newborn screen shows absent T-cell receptor excision circles. The infant appears well. Which condition must be urgently evaluated?

A. Severe combined immunodeficiency because very low or absent T-cell output can be detected before serious infections develop.

B. Hereditary spherocytosis because T-cell receptor excision circles reflect red-cell membrane turnover rather than lymphocyte development.

C. Selective IgA deficiency because absent T-cell receptor excision circles specifically identify failure of secretory IgA production.

D. Chronic granulomatous disease because T-cell receptor excision circles directly measure neutrophil oxidative burst activity.

162. A newborn's heart rate remains below 60 beats/min despite effective ventilation and 60 seconds of coordinated chest compressions. Umbilical venous access is available. Which epinephrine strategy is preferred?

A. Give intravascular epinephrine 0.1 to 0.3 mg/kg because neonatal dosing is tenfold higher than pediatric advanced life support dosing.

B. Withhold epinephrine until 10 minutes of compressions have elapsed because drug therapy before that point worsens neurologic outcome.

C. Give subcutaneous epinephrine 0.01 mg/kg because slower absorption produces a safer and more sustained effect during neonatal arrest.

D. Give intravascular epinephrine 0.01 to 0.03 mg/kg and repeat every 3 to 5 minutes if the heart rate remains below 60 in this setting.

163. A very preterm infant in the NICU passes an otoacoustic-emission screen. Why might an automated auditory brainstem response still be important in high-risk NICU screening programs?

A. Otoacoustic emissions detect most forms of neural hearing disorder, so auditory brainstem testing has no additional value in NICU infants.

B. Auditory brainstem response measures primarily ear-canal patency and therefore cannot identify sensorineural or neural hearing impairment.

C. Auditory brainstem response is used primarily to diagnose middle-ear fluid, whereas otoacoustic emissions directly measure cortical auditory processing for this infant.

D. Auditory brainstem response can detect neural pathway dysfunction such as auditory neuropathy that may be missed by tests of cochlear outer-hair-cell function alone.

164. A newborn has a left lower-lobe lung mass that does not communicate normally with the tracheobronchial tree and receives arterial blood from the descending aorta. Which diagnosis fits best?

A. Pulmonary sequestration, defined by nonfunctioning lung tissue with anomalous systemic arterial supply and abnormal airway connection.

B. Congenital pulmonary airway malformation, which by definition typically receives its primary arterial supply directly from the descending aorta.

C. Transient tachypnea, which produces a discrete noncommunicating lung mass with a systemic feeding vessel visible on imaging.

D. Pulmonary interstitial emphysema, which is a prenatal structural lesion supplied by systemic arteries rather than an acquired air-leak syndrome.

165. A NICU wants to reduce unplanned extubations. The team tests a new securement checklist in one pod for two weeks, reviews results, modifies the process, and repeats the cycle. Which quality-improvement method is being used?

A. A meta-analysis, because repeated local cycles statistically pool published studies to estimate a single treatment effect.

B. A case-control study, because the team is selecting infants by outcome and retrospectively comparing prior securement exposures.

C. A Plan-Do-Study-Act cycle, using small iterative tests of change to learn and refine a local process in this clinical context.

D. A randomized efficacy trial, because local workflow changes become blinded clinical experiments once outcomes are measured.

166. A trial reports a risk ratio of 0.82 with a 95% confidence interval of 0.68 to 0.99 for a binary adverse outcome. Which interpretation is most appropriate?

A. The treatment reduces risk by exactly 18% in future populations because the point estimate has no sampling uncertainty once the interval is reported.

B. The interval excludes the null value of 1.0, so the result is statistically compatible with a modest reduction at the conventional 0.05 level.

C. The confidence interval suggests the study is free of bias and confounding because values in the interval are below the null.

D. The result is not statistically significant because confidence intervals must exclude zero rather than one for a risk ratio.

167. An infant is born pale and hypotensive after a large placental abruption. Cord gases show severe metabolic acidosis and the hemoglobin is low. What maternal-fetal event best unifies these findings?

A. Chronic maternal iron deficiency alone produces abrupt neonatal hypotension and severe cord acidosis at the moment of delivery.

B. Placental separation improves fetal oxygen delivery transiently, so severe acidosis should prompt evaluation primarily for an inborn metabolic disease.

C. Acute fetal blood loss with impaired placental gas exchange can produce both anemia and perinatal asphyxia after major abruption.

D. Abruption causes isolated neonatal polycythemia because placental separation forces maternal erythrocytes into the fetal circulation.

168. A preterm infant has a symptomatic hemodynamically significant PDA and no contraindication to medical therapy. Which medication is an accepted pharmacologic option for ductal closure?

A. Prostaglandin E1 is used to close the ductus by stimulating smooth-muscle constriction in preterm infants with pulmonary overcirculation.

B. Adenosine closes the ductus by transiently blocking atrioventricular conduction and reducing left-to-right shunt volume.

C. Inhaled nitric oxide closes the ductus by selectively reducing pulmonary vascular resistance and eliminating prostaglandin signaling.

D. Ibuprofen can inhibit prostaglandin synthesis and is commonly used for pharmacologic PDA closure in selected preterm infants.

169. A mechanically ventilated extremely preterm infant has repeatedly large delivered tidal volumes despite modest pressure settings. Which ventilator strategy best limits volutrauma when feasible?

A. Use volume-targeted ventilation to keep delivered tidal volume within a set range while pressures adjust to changing lung compliance here.

B. Maintain severe hypocapnia because aggressive ventilation and very low carbon dioxide are protective against both BPD and brain injury.

C. Use fixed high inspiratory pressures without monitoring exhaled tidal volume because pressure alone determines ventilator-induced lung injury.

D. Increase tidal volume deliberately because larger breaths prevent bronchopulmonary dysplasia by eliminating atelectasis during ventilation.

170. A growth-restricted newborn has a prominent occiput, micrognathia, clenched hands with overlapping fingers, and rocker-bottom feet. Which diagnosis is most likely?

A. Trisomy 21, which usually causes this exact combination of overlapping fingers, rocker-bottom feet, and a prominent occiput.

B. Trisomy 18, a chromosomal disorder associated with severe growth restriction, characteristic hand positioning, and multiple congenital anomalies.

C. Noonan syndrome, which typically presents with severe prenatal growth restriction and clenched hands from fixed arthrogryposis.

D. Beckwith-Wiedemann syndrome, which is characterized by macroglossia and overgrowth rather than severe growth restriction and micrognathia for this case.

171. A newborn has widespread papules and nodules, hepatosplenomegaly, respiratory distress, and maternal febrile illness around delivery. Placental pathology shows microabscesses, and cultures grow Listeria monocytogenes. Which syndrome is being described?

A. Neonatal tetanus, which presents with rigidity and spasms rather than a congenital disseminated granulomatous or papular eruption.

B. Congenital CMV, which produces viral inclusions and periventricular calcifications rather than positive bacterial cultures for Listeria.

C. Late-onset GBS meningitis, which usually presents at several weeks of age and does not produce placental microabscesses from maternal listeriosis.

D. Early-onset congenital listeriosis with disseminated infection, sometimes termed granulomatosis infantiseptica in this clinical context.

172. A term newborn is apneic after birth with a heart rate of 110 beats/min. Initial warming, positioning, and stimulation have been completed. What should be done next?

A. Give intravenous epinephrine before ventilation because medication should precede airway support in an apneic newborn with preserved heart rate in this setting.

B. Begin chest compressions because any apnea at birth indicates inadequate cardiac output even when the heart rate is above 100.

C. Begin positive-pressure ventilation because apnea or ineffective breathing is the primary indication for assisted ventilation despite a heart rate above 100.

D. Observe for spontaneous recovery for several minutes because a heart rate above 100 excludes the need for respiratory intervention.

173. A newborn with tetralogy of Fallot develops an abrupt episode of deep cyanosis and irritability during crying. Which immediate maneuver can help reduce right-to-left shunting while additional treatment is arranged?

A. Give indomethacin to close the ductus because ductal flow is the usual cause of hypercyanotic spells in tetralogy of Fallot.

B. Place the infant in a knee-chest position to increase systemic vascular resistance and reduce right-to-left ventricular shunting.

C. Hyperventilate with very low carbon dioxide because respiratory alkalosis reliably worsens pulmonary vascular resistance and relieves the spell.

D. Lower systemic vascular resistance with a potent vasodilator because reducing left-sided pressure diverts more blood into the pulmonary artery.

174. A 25-week infant is 38 weeks postmenstrual age and still requires supplemental oxygen after a prolonged course of mechanical ventilation and oxygen exposure. Which diagnosis best describes the chronic lung disorder?

A. Bronchopulmonary dysplasia, a chronic lung disease of prematurity reflecting arrested lung development and postnatal injury in the clinical situation described here.

B. Persistent respiratory distress syndrome is the best diagnosis because primary surfactant deficiency commonly explains oxygen dependence through term-equivalent age.

C. Postinfectious bronchiolitis is the best diagnosis because chronic oxygen need in an extremely preterm infant usually reflects recurrent viral lower-airway injury.

D. Primary ciliary dyskinesia is the best diagnosis because prolonged neonatal ventilation is a typical cause of acquired ciliary dysfunction and chronic oxygen need.

175. A newborn has a sharply demarcated absence of skin on the scalp with an otherwise stable examination. What is the most likely diagnosis?

A. Neonatal lupus, which typically causes a solitary full-thickness scalp defect present at birth without annular photosensitive lesions.

B. Erythema toxicum, which is defined by a congenital focal absence of scalp skin rather than transient papules and pustules.

C. Staphylococcal scalded skin syndrome, which is present as a fixed isolated scalp defect at birth and remains localized without tenderness in this case.

D. Aplasia cutis congenita, which most often involves the scalp and may range from a superficial defect to a lesion over deeper structural abnormalities.

176. A group O mother delivers a group A term infant who develops early jaundice and mild anemia. The direct antiglobulin test is positive. Which diagnosis is most likely?

A. Rh hemolytic disease because ABO incompatibility cannot cause neonatal hemolysis when the mother has blood group O.

B. G6PD deficiency is proven because immune antibody binding to neonatal erythrocytes is the mechanism of oxidative hemolysis in G6PD deficiency.

C. Hereditary spherocytosis is proven because a positive direct antiglobulin test establishes an intrinsic red-cell membrane defect.

D. ABO hemolytic disease from maternal IgG anti-A or anti-B antibodies crossing the placenta and hemolyzing fetal red cells.

177. A newborn develops widespread superficial blisters and erosions after minimal friction. Similar problems affected several relatives. Which diagnosis should be considered?

A. Epidermolysis bullosa, an inherited group of skin-fragility disorders in which minor mechanical trauma produces blistering.

B. Subcutaneous fat necrosis, which presents as fragile superficial bullae rather than firm subcutaneous plaques and nodules.

C. Erythema toxicum, which produces friction-induced large bullae and chronic skin fragility across multiple generations.

D. Miliaria, which causes deep erosions and mucosal blistering because sweat-duct obstruction disrupts the dermal-epidermal junction.

178. A newborn with dehydration and sepsis develops seizures. MRI venography shows thrombosis of the superior sagittal sinus. Which diagnosis is present?

A. Periventricular leukomalacia, which is caused by venous sinus occlusion and is diagnosed by absence of flow in the superior sagittal sinus.

B. Cerebral sinovenous thrombosis, a neonatal stroke mechanism associated with systemic illness, dehydration, and prothrombotic conditions here.

C. Perinatal arterial ischemic stroke is the most likely diagnosis because seizures plus a major vascular abnormality usually indicate an arterial rather than venous occlusion.

D. Hypoxic-ischemic encephalopathy is the most likely diagnosis because systemic sepsis and dehydration cause diffuse cerebral injury rather than venous sinus thrombosis.

179. Prenatal ultrasonography shows a multicystic intrathoracic lung lesion supplied by the pulmonary circulation. After birth, the infant has mild respiratory distress. Which diagnosis is most likely?

A. Bronchopulmonary dysplasia, which is a focal congenital cystic lesion usually detected on prenatal ultrasonography in a term fetus.

B. Congenital pulmonary airway malformation, a developmental cystic lung lesion that can vary widely in size and postnatal symptoms here.

C. Pulmonary sequestration, which is defined by a normal bronchial connection and blood supply exclusively from the pulmonary artery.

D. Pulmonary interstitial emphysema, which is a congenital cystic malformation present before exposure to positive-pressure ventilation here.

180. During resuscitation of a very preterm infant, the axillary temperature is 35.6 C shortly after admission to the NICU. Which temperature goal best reflects recommended neonatal stabilization?

A. Use warming measures to maintain a temperature approximately 36.5 to 37.5 C while avoiding both hypothermia and hyperthermia.

B. Allow temperatures near 35 C because mild spontaneous hypothermia is neuroprotective for preterm infants after delivery.

C. Target 38.5 to 39 C because preterm infants require intentional hyperthermia to compensate for evaporative heat loss.

D. Do not actively manage temperature until respiratory status is stable because thermal control has little effect on neonatal morbidity.

181. Why is careful oxygen targeting important in an extremely preterm infant from an ophthalmologic perspective?

A. Hyperoxia prevents ROP by persistently dilating immature retinal vessels, so higher oxygen targets are preferred when the retina is incompletely vascularized.

B. Excessive and fluctuating oxygen exposure can disrupt normal retinal vascular development and contribute to retinopathy of prematurity.

C. ROP is caused primarily by low oxygen exposure, so episodes of hyperoxia have no role in retinal vascular injury.

D. Retinal vascularization is complete by 24 weeks, making postnatal oxygen exposure irrelevant to ophthalmologic outcomes in extremely preterm infants.

182. A newborn has an open lumbosacral myelomeningocele. What is the immediate care priority before definitive neurosurgical closure?

A. Place the infant supine directly on the lesion because pressure reduces cerebrospinal fluid leakage and promotes spontaneous skin closure in the neonatal setting described.

B. Protect the neural tissue with a sterile moist nonadherent dressing, position to avoid pressure on the sac, and evaluate for associated hydrocephalus and other anomalies.

C. Delay evaluation for hydrocephalus because ventricular enlargement cannot occur until after the spinal defect is surgically closed.

D. Begin vigorous range-of-motion exercises over the exposed placode because early friction prevents tethering and infection.

183. A thriving preterm infant has frequent small-volume regurgitation but no apnea temporally linked to feeds, no hematemesis, and normal growth. What is the best general interpretation?

A. Uncomplicated gastroesophageal reflux is common in infants and should not automatically be labeled the cause of apnea or treated with acid suppression.

B. The infant has pyloric stenosis because any postprandial regurgitation in a preterm infant represents progressive gastric outlet obstruction.

C. Acid reflux is the proven cause of most apnea of prematurity, so acid-suppressive medication should be started whenever regurgitation occurs.

D. The infant has NEC because physiologic reflux does not occur before term-equivalent age and small-volume emesis indicates intestinal ischemia in context.

184. A newborn with congenital complete heart block has a ventricular rate of 45 beats/min, poor perfusion, and signs of low cardiac output. Which management principle is most appropriate?

A. Adenosine should be given repeatedly because atrioventricular nodal blockade can restore normal conduction in immune-mediated complete block here.

B. Observation alone is preferred because congenital complete heart block is typically well tolerated when the heart is structurally normal.

C. Urgent cardiology evaluation for pacing is warranted because profound bradycardia with low cardiac output may require permanent pacemaker therapy.

D. Indomethacin should be started because ductal closure reliably raises the ventricular rate in congenital complete heart block.

185. A newborn with 22q11.2 deletion has a very small thymus and profound T-cell lymphopenia. Which precaution is most important while immune function is being defined?

A. Give live vaccines early because thymic hypoplasia improves vaccine replication and produces stronger protective immunity.

B. Transfuse unscreened family-member blood because related donors eliminate the risk of transfusion-associated graft-versus-host disease.

C. Avoid inactivated vaccines because nonlive vaccines are uniquely dangerous in T-cell lymphopenia.

D. Avoid unselected live vaccines and use appropriately screened or treated blood products when severe T-cell immunodeficiency is present.

186. A 26-week infant has persistent hyponatremia despite an appropriate total fluid intake. Urine sodium remains high and there is no edema or evidence of SIADH. What physiologic feature of prematurity can contribute?

A. The neonatal renin-angiotensin-aldosterone system is clinically inactive, so sodium balance cannot be influenced by mineralocorticoid signaling.

B. Immature renal tubules have limited sodium reabsorptive capacity, so preterm infants can have substantial urinary sodium losses during growth.

C. Low glomerular filtration eliminates urinary electrolyte loss, making hyponatremia in prematurity almost typically a laboratory artifact.

D. Preterm kidneys retain sodium more avidly than adult kidneys, so persistent urinary sodium loss is incompatible with renal immaturity.

187. A 28-week infant with RDS is on CPAP but now requires increasing oxygen and has substantial work of breathing. Which treatment addresses the underlying pathophysiology most directly?

A. Start inhaled nitric oxide routinely because selective pulmonary vasodilation corrects the primary surfactant deficiency of uncomplicated RDS.

B. Use prolonged hyperventilation to lower carbon dioxide because respiratory alkalosis stimulates endogenous surfactant production within minutes.

C. Administer a loop diuretic because fluid removal replaces deficient surfactant and immediately restores alveolar surface tension.

D. Administer exogenous surfactant using an appropriate minimally invasive or intubation-based technique while continuing lung-protective support.

188. After a vacuum-assisted delivery, a newborn becomes pale and tachycardic. The scalp swelling is fluctuant, crosses suture lines, and expands over several hours. What is the most dangerous diagnosis?

A. Cephalohematoma, which is confined by periosteal attachments and typically crosses multiple suture lines while causing rapid massive blood loss.

B. Aplasia cutis, which presents as fluctuant scalp swelling after vacuum extraction rather than a congenital absence of skin.

C. Caput succedaneum, which usually expands progressively after birth and can contain enough blood to cause severe anemia and shock.

D. Subgaleal hemorrhage, which can accommodate a large blood volume and cause life-threatening hypovolemic shock for this infant.

189. A 2-week-old has symmetric proximal weakness, absent deep tendon reflexes, tongue fasciculations, and preserved sensation. Which diagnosis is most likely?

A. Neonatal stroke, which typically causes symmetric proximal weakness and generalized areflexia rather than focal neurologic findings.

B. Congenital myotonic dystrophy, which characteristically causes brisk reflexes and preserved respiratory strength in a severely hypotonic infant.

C. Spinal muscular atrophy from loss of anterior horn motor neurons, most commonly related to biallelic SMN1 dysfunction in this setting.

D. Hypoxic-ischemic encephalopathy, which causes isolated lower motor neuron weakness with tongue fasciculations and absent reflexes but normal tone centrally.

190. A former 27-week infant later develops spastic diplegia. Earlier neuroimaging showed bilateral cystic injury in the periventricular white matter. Which neonatal brain lesion best explains this outcome?

A. Isolated choroid plexus cysts, which are postnatal destructive white-matter lesions that predict bilateral lower-extremity spasticity.

B. Periventricular leukomalacia, a white-matter injury of prematurity associated with later motor impairment and cerebral palsy.

C. Subgaleal hemorrhage, which causes extracranial blood loss but not selective bilateral periventricular white-matter necrosis.

D. Benign enlargement of the subarachnoid spaces, which commonly causes spastic diplegia after preterm birth despite normal white matter.

191. A very preterm infant has abnormal general movements and persistent asymmetry at 4 months corrected age. What is the best next step?

A. Repeat the developmental examination in six months before referral because motor asymmetry at four months corrected age often resolves as preterm tone normalizes here.

B. Refer promptly for developmental assessment and early intervention rather than waiting for a definitive cerebral palsy diagnosis before starting therapy for this case.

C. Order brain MRI first and defer therapy until imaging identifies a structural lesion because early intervention should follow a definitive neurologic diagnosis.

D. Continue routine high-risk follow-up without therapy because standardized intervention is most effective after persistent motor delay is documented at one year.

192. A term infant after a difficult delivery develops firm, erythematous-purple subcutaneous plaques over the shoulders and back but is otherwise well. Which diagnosis is most likely?

A. Subcutaneous fat necrosis of the newborn, which can follow perinatal stress and warrants later monitoring for hypercalcemia here.

B. Erythema toxicum, which produces large indurated plaques in the subcutaneous fat and is strongly linked to neonatal hypercalcemia.

C. Epidermolysis bullosa, which causes deep firm nodules from pressure injury rather than superficial blisters after minimal trauma.

D. Congenital candidiasis, which produces firm sterile nodules without scaling or pustules and is not associated with maternal infection.

193. A phenotypic female newborn has nonpitting edema of the hands and feet, a webbed neck, and coarctation of the aorta. Which chromosomal condition should be suspected?

A. Klinefelter syndrome, which affects phenotypic males and typically does not produce neonatal lymphedema or coarctation.

B. Turner syndrome, usually caused by complete or mosaic monosomy X and associated with neonatal lymphedema and left-sided cardiac lesions.

C. Trisomy 13, which can cause multiple anomalies but is not classically associated with webbed neck and neonatal peripheral lymphedema here.

D. 22q11.2 deletion syndrome, which is associated with conotruncal lesions and hypocalcemia rather than monosomy-X-type lymphedema.

194. A 24-week infant is receiving supplemental oxygen. Why is continuous oxygen-saturation targeting important rather than simply using high inspired oxygen to eliminate desaturation?

A. High inspired oxygen prevents retinopathy by suppressing retinal vaso-obliteration and accelerating normal retinal vascularization.

B. Oxygen saturation has little relationship to tissue oxygen exposure, so fixed 100% oxygen is safer than titrated supplementation in extreme prematurity.

C. Preterm infants are resistant to oxygen toxicity because fetal hemoglobin prevents reactive oxygen species from forming in immature tissues.

D. Excess oxygen exposure can contribute to oxidative injury such as retinopathy and lung damage, while inadequate oxygen also carries important risks.

195. A newborn has micrognathia, glossoptosis, and episodic upper-airway obstruction that worsens when supine. Which initial airway strategy is often helpful in Pierre Robin sequence?

A. Give bronchodilators as first-line therapy because the obstruction is caused by lower-airway smooth-muscle constriction rather than tongue position for the presentation described.

B. Use positioning and individualized airway support, such as prone or side positioning under monitored care, before escalating to nasopharyngeal or surgical options when needed.

C. Perform immediate tracheostomy in most cases because noninvasive positioning or nasopharyngeal support cannot relieve glossoptotic obstruction.

D. Force supine positioning because gravity pulls the tongue forward in micrognathia and reliably opens the airway.

196. A newborn after shoulder dystocia holds one arm adducted and internally rotated with the elbow extended and forearm pronated. Grasp is preserved. Which injury is most likely?

A. Spinal muscular atrophy, which causes symmetric generalized weakness rather than an isolated unilateral birth-related arm posture.

B. Radial nerve palsy, which produces wrist drop but usually preserves shoulder rotation and elbow flexion in the pattern described.

C. A lower brachial plexus injury involving C8-T1 roots, which would more strongly affect hand function while sparing shoulder abduction.

D. An upper brachial plexus injury involving primarily C5-C6 roots, producing the classic Erb pattern with preserved hand grasp here.

197. Parents at 22 to 23 weeks' gestation ask whether active neonatal resuscitation should be attempted if delivery occurs tonight. Which counseling approach is most appropriate?

A. Use shared decision-making that explains prognosis and uncertainty, explores family goals and values, and offers ethically appropriate resuscitative or comfort-focused options within local practice standards.

B. Ask the obstetric team to decide without the parents because neonatal clinicians should not discuss prognosis before the infant is born.

C. Promise a precise individual survival percentage and recommend the same treatment for most families because values should not influence periviable decisions given the gestational and clinical findings described.

D. Avoid discussing comfort care because withholding intensive treatment is ethically impermissible whenever a fetal heartbeat is present at delivery.

198. A preterm infant receiving ibuprofen for a hemodynamically significant PDA develops oliguria and rising creatinine. Which pharmacologic mechanism contributes to this adverse effect?

A. Cyclooxygenase inhibition lowers prostaglandin production, which can reduce renal blood flow in a prostaglandin-dependent neonatal circulation.

B. Ibuprofen blocks antidiuretic hormone receptors, producing free-water diuresis that typically precedes neonatal kidney injury.

C. Ibuprofen activates prostaglandin receptors in the renal artery, causing vasodilation and pathologic glomerular hyperfiltration with oliguria.

D. Ibuprofen is converted to a direct tubular toxin primarily by mature adult liver enzymes, so nephrotoxicity is not related to prostaglandin inhibition.

199. A newborn has recurrent electrographic seizures after an acute hypoxic-ischemic brain injury. Glucose and electrolytes are normal. Which medication is commonly used as first-line antiseizure treatment in this setting?

A. Oral carbamazepine is universally first-line because it controls acute symptomatic neonatal seizures better than intravenous medications.

B. Phenobarbital remains a common first-line treatment for neonatal seizures when no specific channelopathy suggests another agent in this case.

C. No antiseizure medication should be given because neonatal electrographic seizures cannot contribute to brain injury or clinical instability.

D. Acetazolamide is preferred because reducing cerebrospinal fluid production is the principal mechanism for stopping HIE-related seizures.

200. A post-term infant born through thick meconium has respiratory distress, coarse breath sounds, and a chest radiograph with patchy bilateral opacities and hyperinflation. Which diagnosis is most likely?

A. Meconium aspiration syndrome with airway obstruction, chemical pneumonitis, surfactant dysfunction, and risk of pulmonary hypertension here.

B. Transient tachypnea from delayed fetal lung-fluid clearance, which usually causes mild disease with perihilar streaking and fissural fluid.

C. Respiratory distress syndrome from primary surfactant deficiency, which is most typical in extreme prematurity with diffuse low-volume lungs.

D. Pulmonary interstitial emphysema from ventilator injury, which requires prior positive-pressure exposure and appears as interstitial air collections.

Answers

1. C. Extremely preterm infants face interrelated medical and developmental risks. Coordinated follow-up complements primary care by tracking growth, neurodevelopment, sensory function, feeding, respiratory health, and access to early services.

2. C. Infants born to HBsAg-positive mothers require prompt postexposure prophylaxis with both hepatitis B vaccine and HBIG at separate sites within 12 hours of birth. Follow-up includes completion of the vaccine series and later postvaccination serologic testing.

3. D. Specificity is the proportion of people without the disease who test negative. High specificity means relatively few false-positive results, but it does not by itself determine the positive predictive value.

4. B. The 22q11.2 deletion can disrupt pharyngeal pouch development, producing conotruncal heart defects, hypocalcemia from hypoparathyroidism, and thymic hypoplasia with variable T-cell deficiency.

5. D. ROP screening targets infants with immature retinal vascularization, especially those born very preterm or at very low birth weight, with selected larger or more mature infants screened when their clinical course is unusually unstable.

6. D. Erythema toxicum is a common benign neonatal eruption that appears in the first days of life in otherwise well infants. Systemic illness, grouped vesicles, or extensive erosions should prompt consideration of infectious or blistering disorders instead.

7. B. Infants with congenital hypothyroidism can appear normal at birth because of maternal thyroid hormone exposure. Rapid confirmation and early levothyroxine treatment are essential to protect neurodevelopment.

8. A. Modern bilirubin management uses hour-specific thresholds that vary with gestational age and neurotoxicity risk factors such as hemolysis or clinical instability. The same bilirubin concentration can therefore require different action in two infants.

9. C. Number needed to treat is the reciprocal of the absolute risk reduction. A reduction from 30% to 20% is an absolute reduction of 0.10, giving an NNT of 10 over the study's specified time horizon.

10. B. Abrupt deterioration with unilateral absent breath sounds and evidence of pleural air is concerning for tension pneumothorax. In an unstable infant, decompression should not be delayed for radiographic confirmation.

11. A. Human milk alone often does not provide enough protein and minerals for optimal VLBW growth at typical feeding volumes. Fortification is used to increase nutrient density while retaining the benefits of human milk.

12. C. Early hypocalcemia is more common in infants of diabetic mothers and preterm or stressed infants. Symptomatic or marked hypocalcemia requires confirmation, evaluation of magnesium and other contributors, and appropriate calcium replacement.

13. A. Congenital CMV must be demonstrated very early, generally within 21 days, because later viral detection cannot reliably distinguish congenital from postnatal infection. PCR of saliva is commonly used with urine confirmation; antibody testing is not diagnostic for congenital infection.

14. A. Intention-to-treat analysis preserves the original randomized groups and therefore much of randomization's protection against confounding. Per-protocol or as-treated analyses can be informative but are more vulnerable to selection bias.

15. C. Palliative care is an approach to comfort and quality of life rather than a single treatment or location. It can coexist with selected life-prolonging interventions and supports the infant and family through decision-making, symptom management, and bereavement.

16. D. Severe symptomatic neonatal hypoglycemia is an emergency because prolonged neuroglycopenia can injure the brain. IV dextrose is used promptly while recurrent or persistent hypoglycemia is evaluated for causes such as hyperinsulinism, endocrine disease, or metabolic disorders.

17. A. Classic PKU results from deficient phenylalanine hydroxylase activity, leading to high phenylalanine and low downstream tyrosine. Early dietary management prevents severe intellectual disability and other neurologic consequences.

18. D. Postnatal systemic corticosteroids are not universal therapy. In selected infants with prolonged ventilator dependence and high BPD risk, a lower-dose regimen may facilitate extubation, but the decision requires individualized risk-benefit assessment.

19. D. A large left-to-right PDA can increase pulmonary blood flow and left-heart volume loading while reducing diastolic systemic perfusion. Echocardiographic findings such as left atrial enlargement and systemic diastolic flow reversal support hemodynamic significance when interpreted with the clinical picture.

20. C. Preterm birth interrupts third-trimester calcium and phosphorus accretion. Adequate mineral intake, along with vitamin D and appropriate overall nutrition, helps reduce metabolic bone disease of prematurity.

21. B. Failure to pass an orogastric tube with copious secretions and choking strongly suggests esophageal atresia. A distal tracheoesophageal fistula is common and can allow air into the stomach while increasing aspiration risk.

22. D. Pulmonary hemorrhage presents with acute respiratory deterioration and blood from the airway, often with diffuse radiographic opacification. Extremely preterm infants with cardiovascular instability or significant ductal shunting are at increased risk.

23. C. Prolonged parenteral nutrition is associated with cholestatic liver disease, particularly in extremely preterm or septic infants with limited enteral intake. Management includes advancing enteral nutrition when feasible and optimizing PN composition while excluding other causes of cholestasis.

24. B. A modest degree of hypercapnia may be tolerated when pH and clinical status are acceptable, particularly if this helps avoid injurious ventilation. The goal is not uncontrolled hypercapnia but lung-protective gas exchange.

25. A. Carbon dioxide is an important regulator of cerebral vascular tone. Excessive ventilation with severe hypocapnia can reduce cerebral perfusion and has been associated with neurologic injury, so extreme PaCO2 reduction should be avoided.

26. D. Large VSDs may cause few symptoms immediately after birth because pulmonary vascular resistance is initially high. As resistance falls over the first weeks, the left-to-right shunt increases and pulmonary overcirculation can produce heart failure and poor growth.

27. D. Mother's own milk is the preferred enteral nutrition for VLBW infants when available. Because unfortified human milk may not meet all protein and mineral needs of a growing preterm infant, fortification is often added as feeds advance.

28. B. Late vitamin K deficiency bleeding can cause catastrophic intracranial hemorrhage in infants who did not receive prophylaxis, especially with exclusive breast-feeding or unrecognized cholestasis. Coagulation studies show deficiency of vitamin-K-dependent factors while platelets may remain normal.

29. C. Central venous catheters are a major neonatal thrombosis risk. Management is individualized: remove an unnecessary catheter when feasible, consider anticoagulation for clinically important thrombosis, and reserve thrombolysis for selected limb- or organ-threatening situations because bleeding risk is substantial.

30. A. Meconium ileus is a classic neonatal manifestation of cystic fibrosis and should prompt CFTR-directed evaluation even when newborn screening is pending. Complicated cases may include perforation, volvulus, or meconium peritonitis.

31. D. Antenatal magnesium sulfate is used for fetal neuroprotection when early preterm birth is imminent and has been associated with a lower risk of cerebral palsy. It serves a different purpose from antenatal corticosteroids, which mainly accelerate organ maturation.

32. C. Neonatal meningitis can progress rapidly and should be treated empirically after appropriate cultures are obtained, without unnecessary delay. The regimen is chosen for age, likely pathogens, resistance patterns, and local protocols, and HSV coverage is added when clinically indicated.

33. A. In congenital diaphragmatic hernia, bag-mask ventilation can inflate intrathoracic bowel and worsen lung compression. Initial management emphasizes endotracheal ventilation, gastric decompression, gentle lung-protective support, and stabilization before repair.

34. D. Transient abnormal myelopoiesis is a clonal megakaryoblastic proliferation unique to infants with trisomy 21, usually involving GATA1 variants. Many cases regress, but severe organ involvement can be life-threatening and survivors have an increased later risk of myeloid leukemia of Down syndrome.

35. C. A visually significant congenital cataract can deprive the developing visual system of patterned input and lead to amblyopia. Early ophthalmologic management addresses the cataract and then requires optical rehabilitation and amblyopia therapy.

36. C. An omphalocele is a membrane-covered midline defect at the umbilical ring and may contain liver or bowel. It has important associations with aneuploidy, cardiac disease, and syndromic conditions such as Beckwith-Wiedemann syndrome.

37. A. A regular narrow-complex tachycardia near 250 to 300 beats/min in a stable infant is typical of supraventricular tachycardia. After vagal maneuvers, rapid IV adenosine is an appropriate acute therapy; synchronized cardioversion is used for hemodynamic instability.

38. D. Noonan syndrome often includes webbed neck, pectus abnormalities, characteristic facial features, and pulmonary valve stenosis or hypertrophic cardiomyopathy. It can affect any sex and is commonly caused by variants in the RAS/MAPK pathway.

39. A. Severe infection can produce diffuse inflammatory lung injury with secondary surfactant dysfunction, reduced compliance, and hypoxemia. The diagnosis requires exclusion of conditions better explaining the respiratory failure, particularly cardiac causes of edema.

40. D. Neurologic abnormalities during severe unconjugated hyperbilirubinemia are an emergency because they may represent acute bilirubin encephalopathy. Escalation of bilirubin treatment, including exchange transfusion when indicated, is time-sensitive.

41. C. Cystic PVL is a major risk factor for cerebral palsy and other developmental impairment. Damage near descending motor pathways can lead to spasticity, classically affecting the legs more than the arms in spastic diplegia.

42. B. ECMO is a rescue option for selected term or near-term infants with severe, reversible cardiorespiratory failure that persists despite maximal conventional treatment. Candidacy depends on gestation, bleeding risk, anatomy, prognosis, and center-specific criteria.

43. A. Extremely preterm infants are vulnerable to invasive candidiasis, especially with central lines and antibiotic exposure. Management includes systemic antifungal therapy, source control such as catheter removal when possible, repeat cultures, and evaluation for disseminated disease.

44. A. Small or variable residuals are common in preterm infants and are not by themselves diagnostic of NEC or obstruction. Concerning systemic signs, abdominal findings, bloody stool, radiography, and trend over time are more informative.

45. C. Hypernatremia with polyuria and inappropriately dilute urine after central nervous system injury strongly suggests central diabetes insipidus. Management requires careful free-water replacement and treatment with desmopressin when appropriate.

46. C. Bilateral choanal atresia can cause cyclic cyanosis that improves with crying because the newborn can temporarily breathe through the mouth. Establishing an oral airway and securing ventilation when needed precede definitive otolaryngologic repair.

47. B. Severely growth-restricted infants can have low stores of phosphorus and other electrolytes. Rapid anabolic nutrient delivery, especially amino acids and glucose, may drive phosphate and potassium into cells and produce a neonatal refeeding-like syndrome.

48. D. Anemia of prematurity reflects several factors: shorter red-cell survival, rapid somatic growth, phlebotomy, and a blunted erythropoietin response to anemia. The low reticulocyte response helps distinguish it from active hemolysis.

49. B. Maternal intraamniotic infection is a major risk factor for neonatal early-onset bacterial sepsis. Management depends on gestational age, the infant's examination, and the institution's evidence-based early-onset sepsis strategy.

50. B. Ampicillin plus an aminoglycoside is a common empiric regimen for suspected early-onset neonatal sepsis, covering organisms such as GBS and susceptible gram-negative bacilli while cultures are obtained. Local resistance patterns and clinical circumstances can modify therapy.

51. A. Maternal IgG is actively transported across the placenta, with much of the transfer occurring in the third trimester. Extremely preterm infants therefore start life with less passive systemic antibody than term infants.

52. A. Confounding occurs when a third factor is associated with both the exposure and the outcome and distorts their observed relationship. Illness severity is a classic concern when treatment intensity is compared in nonrandomized neonatal data.

53. D. When respiratory status improves after surfactant, minimizing invasive ventilation can reduce ventilator-associated injury. Extubation to CPAP or another appropriate noninvasive mode is considered when spontaneous effort and gas exchange are adequate.

54. B. Osteogenesis imperfecta is a major cause of congenital bone fragility and can present with fractures, osteopenia, deformity, and blue sclerae. Severe neonatal presentations require careful respiratory, orthopedic, and genetic evaluation.

55. C. If the heart rate remains below 60 beats/min despite effective ventilation, coordinated chest compressions are indicated. Neonatal resuscitation generally uses a 3:1 ratio because respiratory failure is the usual mechanism of arrest.

56. B. Randomization is a powerful method for reducing confounding in comparative intervention research because treatment assignment is not determined by patient prognosis or clinician selection. Blinding and allocation concealment further reduce bias when feasible.

57. B. When pulmonary blood flow is ductal dependent, prostaglandin E1 is used to maintain the ductus until catheter-based or surgical therapy is available. Closing the ductus would worsen cyanosis.

58. A. Open disclosure respects families and supports patient safety. The clinical team should address immediate harm, communicate what is known and uncertain, document accurately, and use safety systems to analyze and reduce recurrence.

59. A. Deferred cord clamping for at least roughly 60 seconds is recommended for many preterm infants who do not require immediate resuscitation. Intact-cord milking is not a substitute in extremely preterm infants because safety concerns include severe intraventricular hemorrhage.

60. D. Spontaneous intestinal perforation is distinct from classic NEC and often appears in extremely preterm infants as a focal intestinal perforation with relatively limited surrounding inflammatory bowel changes. Surgical drainage or laparotomy may be required depending on the infant's condition.

61. A. Large IVH can impair CSF absorption or flow and lead to posthemorrhagic ventricular dilatation. Serial head measurements and cranial imaging are used because intervention may be needed before signs of markedly raised intracranial pressure appear.

62. A. Trisomy 13 often produces severe midline developmental abnormalities including holoprosencephaly, cleft lip or palate, microphthalmia, and polydactyly. The pattern distinguishes it from other common aneuploidies.

63. C. Gastroschisis leaves bowel exposed without a protective sac, causing major evaporative heat and fluid loss and risk of injury. Initial care protects the bowel, supports temperature and perfusion, decompresses the stomach, and prepares for staged or primary closure.

64. B. Congenital rubella can produce the classic combination of cataracts, congenital heart disease, and hearing impairment. Prevention depends on maternal immunity and vaccination before pregnancy because live rubella vaccine is not given during pregnancy.

65. C. Pneumatosis with systemic and gastrointestinal signs is strongly concerning for NEC. Enteral feeding is stopped, the stomach is decompressed, antibiotics and supportive care are given, and surgery is involved when perforation or progressive disease is suspected.

66. C. The classic neonatal renal vein thrombosis pattern includes hematuria, thrombocytopenia, and renal enlargement. Doppler imaging supports the diagnosis, and management depends on extension, renal function, and thrombosis risk.

67. D. Red-cell alloimmunization can cause progressive fetal hemolytic anemia. Severe anemia increases cardiac output and cerebral blood-flow velocity and can culminate in high-output failure and hydrops; fetal transfusion is used in appropriately selected cases.

68. A. The combination of extreme prematurity, early respiratory distress, low lung volumes, and diffuse granular opacities is classic for surfactant-deficient respiratory distress syndrome. Surfactant lowers alveolar surface tension and improves compliance.

69. C. For appropriately selected term or near-term infants with moderate to severe hypoxic-ischemic encephalopathy, therapeutic hypothermia started within the first 6 hours improves neurodevelopmental outcomes. Treatment requires protocolized monitoring and controlled rewarming.

70. B. Neonatal seizures are frequently electrographic-only or have subtle clinical signs. Continuous EEG or appropriately interpreted amplitude-integrated EEG with confirmatory EEG improves seizure detection and helps avoid treating nonepileptic movements.

71. D. Systemic blood flow in hypoplastic left heart syndrome is ductal dependent. Prostaglandin E1 maintains or restores ductal patency while ventilation, perfusion, acid-base status, and transfer for definitive cardiac care are managed.

72. C. In d-transposition, systemic and pulmonary circulations run largely in parallel, so survival depends on mixing. When an atrial communication is restrictive and hypoxemia is severe, balloon atrial septostomy can provide rapid improvement in mixing while definitive repair is planned.

73. D. Inappropriate insulin action during hypoglycemia suppresses glycogenolysis, gluconeogenesis, lipolysis, and ketogenesis. The need for high glucose infusion plus low ketones and free fatty acids is a classic biochemical pattern of hyperinsulinemic hypoglycemia.

74. D. Congenital CMV can cause hearing loss that is present at birth, delayed, fluctuating, or progressive. Passing the initial screen therefore does not eliminate the need for ongoing audiologic follow-up in an affected infant.

75. A. Early bacterial pneumonia can mimic or coexist with RDS, especially in preterm infants. Maternal infection history, systemic instability, cultures, and the overall clinical course help distinguish infection from isolated surfactant deficiency.

76. B. For term and late-preterm infants requiring respiratory support, resuscitation can generally begin with room air. Oxygen is then titrated using preductal pulse oximetry and the infant's clinical response rather than routinely starting at 100%.

77. B. Congenital myotonic dystrophy can be severe in newborns, with hypotonia, respiratory failure, feeding weakness, and contractures. Maternal myotonia may be subtle and is an important diagnostic clue to the underlying trinucleotide-repeat disorder.

78. C. Neonatal HSV can progress rapidly and may involve skin, eyes, mouth, CNS, or multiple organs. IV acyclovir should be started promptly when disease is suspected, with treatment duration guided by disease classification and virologic response.

79. C. Early CPAP can stabilize functional residual capacity and reduce exposure to invasive ventilation in spontaneously breathing preterm infants. Surfactant is then given selectively when disease severity warrants it.

80. A. Aminoglycosides are cleared primarily by the kidneys and can contribute to nephrotoxicity. Rising creatinine or oliguria should prompt reassessment of dosing, levels when indicated, other nephrotoxins, and the ongoing need for therapy.

81. D. Fetal hyperinsulinemia in maternal diabetes can stimulate myocardial growth, especially septal hypertrophy. The cardiomyopathy is often transient as the postnatal hyperinsulinemic state resolves, though severe outflow obstruction can require careful management.

82. B. The phenotype and complete atrioventricular septal defect are classic for trisomy 21. Newborns with Down syndrome also warrant assessment for feeding problems, hematologic abnormalities, thyroid disease, hearing issues, and other associated conditions.

83. B. Obstructed total anomalous pulmonary venous return is a surgical emergency. Pulmonary venous obstruction produces severe pulmonary edema and hypoxemia, and medical stabilization is only a bridge to definitive repair.

84. C. Aeration of the lungs and increasing oxygen tension markedly reduce pulmonary vascular resistance. At the same time, removal of the placenta raises systemic vascular resistance, helping reverse fetal shunt patterns and establish serial circulation.

85. D. Pulmonary interstitial emphysema is an air-leak syndrome associated with fragile preterm lungs and positive-pressure ventilation. Radiographs show linear or cystic interstitial air collections that can impair ventilation and venous return.

86. B. Sensitivity is the proportion of people with the disease who test positive. A highly sensitive test has relatively few false-negative results, although predictive values still depend on disease prevalence.

87. B. Antenatal urinary tract dilation requires structured postnatal reassessment because severity and associated features determine the need for repeat imaging, voiding cystourethrography, functional studies, prophylaxis, or urology follow-up.

88. A. Neonatal lupus results from transplacental maternal anti-Ro/SSA or related antibodies. Skin and hematologic manifestations usually resolve as maternal antibodies disappear, whereas congenital heart block can be permanent.

89. A. Routine suctioning, including routine tracheal suctioning, is not recommended solely because meconium is present. Ventilation should not be delayed; suction is reserved for suspected airway obstruction that interferes with effective ventilation.

90. B. Feeding-related coughing and desaturation raise concern for impaired suck-swallow-breathe coordination and aspiration. A multidisciplinary feeding assessment and, when indicated, instrumental evaluation can guide nipple flow, pacing, positioning, or temporary alternate feeding routes.

91. C. Newborns rely heavily on brown-fat-mediated nonshivering thermogenesis, but extremely preterm infants have limited brown fat and immature skin barriers. They therefore require active environmental measures to reduce evaporative, conductive, convective, and radiant heat loss.

92. A. Bilious emesis in a newborn is a surgical warning sign. Malrotation with volvulus can compromise the superior mesenteric blood supply and requires urgent imaging and surgical evaluation rather than routine observation.

93. D. Maternal hyperglycemia stimulates fetal beta-cell hyperplasia and hyperinsulinemia. After cord clamping, the transplacental glucose supply stops while insulin remains high, producing early neonatal hypoglycemia.

94. A. Classic 21-hydroxylase deficiency impairs cortisol and often aldosterone synthesis while increasing androgen production. Salt-wasting infants can present in the first weeks with dehydration, hyponatremia, hyperkalemia, shock, and genital virilization in affected 46,XX infants.

95. C. Relative risk compares event probabilities between groups. Dividing 0.12 by 0.06 gives 2.0, meaning NEC occurred twice as often in the formula-fed group in this cohort; this does not by itself prove causation.

96. D. Hemolytic disease from red-cell alloimmunization is mediated by maternal IgG antibodies against fetal erythrocyte antigens. Severe disease can cause fetal anemia, hydrops, neonatal hyperbilirubinemia, and the need for transfusion or exchange therapy.

97. B. Neonatal growth is multidimensional. Serial weight, length, and head circumference trends help distinguish proportional growth from excess fluid or isolated weight gain and guide nutritional adjustments.

98. A. Critical coarctation can be ductal dependent for systemic blood flow and may present with shock as the ductus constricts. Prostaglandin E1 can reopen or maintain ductal patency and is started while cardiology and surgical management are arranged.

99. B. Pasteurized donor human milk is recommended when mother's own milk is unavailable for many VLBW infants, especially to reduce NEC risk compared with formula. It still requires appropriate handling and usually fortification to meet preterm nutritional needs.

100. C. Newborns normally lose extracellular water after birth, and preterm infants can have substantial early weight loss during diuresis. Sodium trend, urine output, perfusion, intake, and clinical status help distinguish physiologic loss from dehydration.

101. B. The listed abnormalities span multiple components of the VACTERL association. Because VACTERL is an association rather than a single molecular diagnosis, evaluation should also consider syndromes and exposures that can mimic the pattern.

102. B. G6PD deficiency can cause neonatal jaundice through increased bilirubin production and oxidative hemolysis, and severe cases may progress rapidly. Testing should be considered when the clinical pattern or ancestry raises suspicion, recognizing that some assays can be misleading during acute hemolysis.

103. D. Caffeine clearance is slow in premature infants, producing a long half-life that supports once-daily maintenance in typical regimens. Routine serum-level monitoring is not required in every infant but can be useful when response, adherence, dosing, or toxicity is uncertain.

104. B. Severe thrombocytopenia in an otherwise well newborn with a normal maternal platelet count is a classic presentation of fetal and neonatal alloimmune thrombocytopenia. Intracranial hemorrhage can occur antenatally, so prompt platelet-directed management is crucial.

105. A. SIADH causes impaired free-water excretion, leading to hypotonic hyponatremia with urine that remains inappropriately concentrated. Central nervous system and pulmonary disease are important neonatal triggers.

106. D. Placental insufficiency can produce chronic fetal hypoxemia, which increases erythropoietin and erythropoiesis. Growth-restricted infants therefore have an increased risk of polycythemia, although dehydration can further raise a measured hematocrit after birth.

107. C. Small enteral volumes can provide trophic stimulation to the immature intestine while parenteral nutrition supplies most calories and protein. Feeding advancement still depends on the infant's stability and tolerance.

108. D. At birth, neonatal serum creatinine reflects maternal concentration to an important degree. Subsequent trajectory, urine output, gestational age, and other findings are needed to assess neonatal kidney function accurately.

109. D. Prader-Willi syndrome commonly presents in the newborn period with hypotonia, poor feeding, and reduced spontaneous movement. The later phenotype includes excessive appetite, obesity risk, developmental issues, and endocrine abnormalities.

110. A. Neonatal arterial ischemic stroke often presents with focal seizures, while weakness may be subtle initially. MRI with diffusion imaging defines the infarct, and evaluation considers placental, cardiac, hematologic, and perinatal risk factors.

111. C. Visible jaundice during the first 24 hours is abnormal and can reflect hemolysis, infection, or other pathology. A bilirubin level should be obtained promptly and interpreted in relation to postnatal age, gestational age, and risk factors.

112. D. Renal tubular bicarbonate loss can cause hyperchloremic, normal-anion-gap metabolic acidosis. Prematurity itself may contribute to limited tubular acid-base handling, but persistent or severe abnormalities require evaluation for renal tubular disease and other causes.

113. B. Neonatal shock is a clinical syndrome of inadequate tissue perfusion, and hypotension may be a late or inconsistent finding. Capillary refill, temperature gradient, urine output, mental status, lactate trend, and hemodynamics must be integrated.

114. D. Benign neonatal sleep myoclonus is distinguished by occurrence during sleep, cessation with arousal, and absence of an ictal EEG correlate. Recognizing it can prevent unnecessary antiseizure treatment while ensuring concerning alternative causes have been excluded.

115. C. Beckwith-Wiedemann syndrome is characterized by fetal or neonatal overgrowth, macroglossia, abdominal wall defects, and frequent neonatal hypoglycemia from hyperinsulinism. Affected children also need syndrome-specific tumor surveillance.

116. D. Preterm infants have reduced fetal iron accretion, frequent phlebotomy losses, and rapid growth. Enteral iron supplementation is therefore commonly used once feeding is established, with dosing individualized to gestation and transfusion history.

117. C. Placental insufficiency associated with preeclampsia can reduce fetal platelet production, causing early thrombocytopenia, especially in growth-restricted infants. Severe or persistent thrombocytopenia still requires evaluation for other causes.

118. B. Corrected age accounts for the weeks an infant was born before term and is commonly used when judging early growth and developmental milestones, often through about two years. Persistent concerns still require evaluation rather than being attributed automatically to prematurity.

119. D. CHARGE is a clinical pattern strongly associated with pathogenic CHD7 variants. Airway, feeding, hearing, cardiac, and endocrine problems can make the neonatal course complex.

120. D. Aminoglycosides are eliminated mainly by glomerular filtration, which is immature and variable in preterm infants. Dosing therefore considers gestational and postnatal age, renal function, and drug levels when indicated.

121. A. Predictive values depend on pretest probability or prevalence. When disease is more common, a positive result is more likely to represent a true positive, whereas negative predictive value generally decreases.

122. C. Coarctation classically impairs lower-body perfusion, producing diminished femoral pulses and an upper-to-lower extremity pressure gradient. The findings can be subtle while the ductus remains open and become dramatic as ductal flow decreases.

123. B. Chromosomal microarray is commonly used as an initial genomic test in infants with multiple unexplained congenital anomalies because it detects submicroscopic copy-number changes. More targeted testing or broader sequencing may follow depending on the phenotype.

124. C. Hypertrophic pyloric stenosis classically presents at several weeks of age with progressive nonbilious projectile vomiting and persistent hunger. Prolonged vomiting can cause hypochloremic, hypokalemic metabolic alkalosis; fluid and electrolyte correction precedes pyloromyotomy.

125. A. MCAD deficiency impairs use of medium-chain fatty acids during fasting, limiting both energy generation and ketone production. Illness or prolonged fasting can therefore cause sudden hypoketotic hypoglycemia, hepatic dysfunction, encephalopathy, or death.

126. C. Subcutaneous fat necrosis can be followed by clinically important hypercalcemia, sometimes after the skin findings are improving. Calcium should be monitored because symptoms can include irritability, vomiting, poor feeding, dehydration, and nephrocalcinosis.

127. A. Caffeine is the standard pharmacologic therapy for apnea of prematurity after secondary causes of apnea are considered. It increases respiratory drive and has a long half-life that permits convenient dosing.

128. A. The germinal matrix has fragile capillaries and is prominent in very preterm infants. Hemorrhage often occurs in the first days after birth and can extend into the ventricles or brain parenchyma.

129. C. Evaluation of an exposed neonate includes a quantitative nontreponemal serum test compared with the maternal titer and assessment of maternal treatment and infant findings. Cord blood is not preferred because of contamination and testing concerns.

130. C. DIC is a consumptive coagulopathy triggered by severe illness such as sepsis, asphyxia, or shock. The combination of thrombocytopenia, prolonged clotting times, low fibrinogen, and increased fibrin breakdown supports the diagnosis.

131. D. Fetomaternal hemorrhage can cause severe fetal or neonatal anemia, shock, hydrops, or stillbirth depending on the volume and timing. A negative antiglobulin test and evidence of fetal cells in the maternal circulation support blood loss rather than immune hemolysis.

132. D. Twin-twin transfusion syndrome occurs in monochorionic gestations when placental vascular anastomoses create net blood flow from donor to recipient. The donor commonly develops oligohydramnios and growth restriction, while the recipient develops polyhydramnios and cardiovascular volume overload.

133. B. Pale stools, dark urine, and elevated direct bilirubin are red flags for neonatal cholestasis. Conditions such as biliary atresia are time-sensitive, so evaluation should not be delayed by assuming prolonged physiologic or breast-milk jaundice.

134. C. Adequate amniotic fluid and fetal lung distention are important for lung development. Early, prolonged oligohydramnios can cause pulmonary hypoplasia, which may produce severe respiratory failure after birth.

135. B. Preterm infants may have slower and more variable morphine clearance, and active metabolites can accumulate when renal function is impaired. Respiratory status, sedation, blood pressure, and cumulative exposure should be monitored carefully.

136. A. Marked neonatal polycythemia can increase blood viscosity and contribute to neurologic, metabolic, and perfusion abnormalities. In a symptomatic infant with a very high venous hematocrit, partial exchange may be considered after confirming the value and assessing other causes of symptoms.

137. B. Coagulase-negative staphylococci are common skin contaminants but are also important causes of catheter-associated late-onset sepsis in very preterm infants. Repeated positive cultures, compatible symptoms, and device history help distinguish true infection from contamination.

138. C. Classic galactosemia can present after milk exposure with liver dysfunction, jaundice, vomiting, cataracts, and a notable association with E. coli sepsis. Lactose and galactose must be removed promptly while confirmatory testing and supportive care proceed.

139. C. Stable preterm infants generally benefit from enteral feeding rather than prolonged fasting, with advancement individualized to gestation, illness severity, and tolerance. Human milk is preferred when available.

140. B. Delayed meconium passage, distention, and explosive stool after rectal examination are classic clues to Hirschsprung disease. Diagnosis is established by demonstrating absence of ganglion cells on rectal biopsy.

141. C. Prematurity affects several layers of host defense at once. Immature barriers, limited neutrophil reserve and function, lower complement activity, reduced transplacental IgG, invasive devices, and prolonged hospitalization all contribute to infection risk.

142. D. Transient tachypnea is associated with delayed fetal lung-fluid absorption and is more common after cesarean delivery without labor. Hyperinflation, perihilar interstitial markings, and fissural fluid support the diagnosis.

143. C. The classic teaching triad for congenital toxoplasmosis is chorioretinitis, hydrocephalus, and intracranial calcifications, though presentations vary. Confirmatory testing and prolonged antiparasitic therapy are coordinated with infectious disease specialists.

144. B. A PDA is common in extremely preterm infants, but routine prophylactic or early closure has not shown overall benefit compared with expectant management. Treatment is individualized when the shunt appears clinically and hemodynamically important.

145. D. Supportive, family-centered nonpharmacologic care is first-line for many infants with neonatal opioid withdrawal. Medication is added when clinically important dysfunction persists despite optimized supportive measures; naloxone can precipitate acute withdrawal and is not routine therapy.

146. A. Prolonged early oligohydramnios can impair fetal lung expansion and produce pulmonary hypoplasia. The resulting reduction in lung parenchyma and pulmonary vascular bed can cause severe respiratory failure and pulmonary hypertension.

147. C. NEC is strongly suggested by systemic instability, abdominal findings, bloody stool, and pneumatosis. Management includes bowel rest, decompression, antibiotics, cardiorespiratory support, and surgical consultation when perforation or progressive necrosis is suspected.

148. C. Posterior urethral valves are an important cause of lower urinary tract obstruction in male newborns. Bladder decompression, evaluation of renal function and electrolytes, and definitive urologic management are urgent priorities.

149. A. Meaningful informed consent requires understandable communication. Qualified interpreters reduce errors and protect confidentiality; family members, especially minors, should not be relied on for complex consent when professional language services are available.

150. C. Early developmental tests describe current function and can identify important delays, but their ability to predict complex later outcomes is limited. Former extremely preterm children benefit from ongoing surveillance beyond a single reassuring score.

151. A. Intrapartum prophylaxis is highly effective for prevention of early-onset GBS disease but does not prevent late-onset disease. Late-onset GBS can present with bacteremia, meningitis, or focal infection weeks after birth.

152. C. Antenatal corticosteroids accelerate fetal pulmonary maturation and reduce important complications of preterm birth when delivery is likely soon. They are not replaced by diuresis, cesarean delivery, or tocolysis alone.

153. A. Needle decompression is an emergency maneuver for tension physiology, but persistent or recurrent pleural air generally requires definitive drainage. Continued ventilator support should be adjusted to minimize further air leak.

154. D. A preductal saturation higher than the postductal saturation can occur when high pulmonary vascular resistance drives right-to-left ductal shunting. In this context, the finding supports persistent pulmonary hypertension of the newborn.

155. C. Achondroplasia is most often caused by an activating FGFR3 variant that suppresses chondrocyte proliferation at the growth plate. Foramen magnum stenosis is an important early complication requiring careful neurologic and respiratory surveillance.

156. A. Transient neonatal myasthenia can result when maternal pathogenic IgG crosses the placenta and impairs neuromuscular transmission. Affected infants may have weak cry, poor feeding, hypotonia, facial weakness, or respiratory compromise; symptoms usually improve as maternal antibody is cleared, while supportive respiratory and feeding care may be required.

157. B. Inhaled nitric oxide is a selective pulmonary vasodilator used in appropriate term and near-term infants with hypoxic respiratory failure associated with pulmonary hypertension after lung recruitment and other causes of hypoxemia are addressed. Routine rescue use in preterm RDS without a pulmonary hypertension indication is not equivalent.

158. B. Extremely preterm infants have limited nutrient stores and high growth needs, so parenteral nutrition often supplies protein, energy, and micronutrients while enteral feeds are established. The goal is to minimize cumulative nutrient deficits without causing metabolic complications.

159. A. Secretory IgA is an important component of human milk and contributes to mucosal protection by binding pathogens and antigens at epithelial surfaces. Human milk also contains many other antimicrobial and immunomodulatory factors.

160. D. Preterm growth depends on adequate protein as well as energy. If calories are sufficient but linear and lean-mass growth are poor, the protein-to-energy ratio and actual nutrient delivery should be reassessed.

161. A. T-cell receptor excision circle screening identifies markedly reduced thymic T-cell production and can detect SCID before symptoms. An abnormal result requires prompt confirmatory immunologic testing and infection precautions while the diagnosis is clarified.

162. D. Intravascular epinephrine is preferred when the heart rate remains below 60 despite adequate ventilation and compressions. The recommended IV dose is 0.01 to 0.03 mg/kg, with repeat dosing every 3 to 5 minutes when needed.

163. D. Otoacoustic emissions assess cochlear outer-hair-cell function, while auditory brainstem response evaluates neural transmission through the auditory pathway. High-risk NICU infants can have auditory neuropathy, making neural screening particularly relevant.

164. A. Pulmonary sequestration consists of dysplastic lung tissue without normal tracheobronchial communication and with systemic arterial supply, often from the thoracic or abdominal aorta. Imaging of the feeding vessel is diagnostically useful.

165. C. PDSA cycles are a core quality-improvement method: plan a change, do it on a manageable scale, study what happened, and act on the findings. Repeated cycles help refine implementation before broader spread.

166. B. For a relative measure such as a risk ratio, the null value is 1.0. An interval from 0.68 to 0.99 excludes 1.0, but statistical significance does not establish clinical importance, freedom from bias, or generalizability.

167. C. Major placental abruption can compromise uteroplacental oxygen exchange and may be accompanied by fetal hemorrhage. The combination of anemia, shock, and severe metabolic acidosis is therefore compatible with acute blood loss plus hypoxic-ischemic stress.

168. D. Cyclooxygenase inhibition with ibuprofen is an accepted medical option for a symptomatic hemodynamically significant PDA; acetaminophen is another option in selected settings. Prostaglandin E1 has the opposite purpose and maintains ductal patency.

169. A. Excessive tidal volume is an important contributor to ventilator-induced lung injury. Volume-targeted approaches can reduce fluctuations in delivered tidal volume as compliance changes and are often used as part of lung-protective ventilation.

170. B. The constellation of prenatal growth restriction, prominent occiput, micrognathia, overlapping fingers, and rocker-bottom feet strongly suggests trisomy 18. Cardiac and renal anomalies are also common.

171. D. Listeria can cross the placenta and cause severe early-onset sepsis or pneumonia, sometimes with characteristic disseminated granulomatous lesions. Ampicillin is central to treatment, often combined initially with an aminoglycoside depending on severity and local practice.

172. C. Effective ventilation is the central intervention in neonatal resuscitation. An apneic or gasping newborn should receive positive-pressure ventilation even if the initial heart rate is above 100 beats/min.

173. B. A hypercyanotic spell reflects increased right-to-left shunting across the VSD, often from dynamic right ventricular outflow obstruction. Increasing systemic vascular resistance with knee-chest positioning can reduce the shunt; oxygen, calming, volume, and selected medications may also be used.

174. A. Bronchopulmonary dysplasia is the chronic lung disease of prematurity, with contemporary disease reflecting interrupted alveolar and vascular development plus inflammatory and ventilator or oxygen injury. Severity definitions incorporate respiratory support around 36 weeks postmenstrual age.

175. D. Aplasia cutis congenita is a congenital absence of skin, most commonly on the scalp. Small superficial lesions may heal with local care, while larger or atypically located lesions may warrant imaging for associated skull or vascular defects.

176. D. ABO hemolytic disease most often occurs when a group O mother has an A or B infant because some maternal anti-A or anti-B antibodies are IgG and cross the placenta. Disease is often milder than severe Rh alloimmunization but can cause early significant hyperbilirubinemia.

177. A. Blistering after minimal trauma, especially with a family history, raises concern for epidermolysis bullosa. Immediate care emphasizes gentle handling, wound care, infection prevention, pain control, and subtype-directed genetic and dermatologic evaluation.

178. B. Cerebral sinovenous thrombosis can present with seizures, encephalopathy, or venous infarction in neonates. Risk factors include infection, dehydration, and other prothrombotic states, and treatment is individualized with neurologic and hematologic input.

179. B. Congenital pulmonary airway malformation is a developmental lesion characterized by cystic or solid abnormal lung tissue supplied by the pulmonary circulation. Large lesions can cause fetal mediastinal shift or neonatal respiratory compromise.

180. A. Normothermia is an important resuscitation goal. Preterm infants lose heat rapidly, so environmental warming, plastic wrapping or other measures as appropriate, and close temperature monitoring are used to keep temperature in the normal range without overheating.

181. B. ROP reflects abnormal vascular development in an incompletely vascularized retina. Oxygen exposure is one important modifiable factor, although gestational age, growth, illness severity, and other factors also contribute.

182. B. An open myelomeningocele requires careful protection of exposed neural tissue and prevention of contamination or trauma. Associated Chiari II malformation, hydrocephalus, bladder dysfunction, orthopedic abnormalities, and latex sensitivity also require coordinated evaluation.

183. A. Reflux is common in infants, particularly preterm infants, and temporal association with apnea is inconsistent. Acid-suppressive medications have potential harms and should not be used simply for uncomplicated regurgitation without a clear indication.

184. C. Congenital complete heart block can be tolerated when ventricular rates and cardiac output are adequate, but marked bradycardia with symptoms or ventricular dysfunction is a pacing indication. Immune-mediated conduction tissue injury is not reversed with adenosine.

185. D. Thymic hypoplasia in 22q11.2 deletion ranges from mild to profound T-cell deficiency. Severe cellular immunodeficiency changes vaccine and transfusion practices, so immune phenotype should be established before routine live-virus exposure.

186. B. Very preterm kidneys have limited tubular sodium conservation and relatively high obligatory sodium losses. Sodium intake therefore must be individualized using growth, serum sodium, urine findings, and overall fluid balance.

187. D. RDS is driven by surfactant deficiency and structural immaturity. When a preterm infant on noninvasive support meets clinical criteria for rescue therapy, exogenous surfactant treats the fundamental problem while ventilation remains lung protective.

188. D. Subgaleal hemorrhage occurs in the potential space beneath the scalp aponeurosis and can spread widely across suture lines. Because the space can hold a large fraction of neonatal blood volume, serial examination, hematocrit, perfusion, and urgent resuscitation are essential.

189. C. SMA produces a lower motor neuron pattern with hypotonia, symmetric weakness, areflexia, and sometimes tongue fasciculations. Newborn screening allows presymptomatic diagnosis in many regions, which is important because disease-modifying therapy is most effective when started early.

190. B. Cystic periventricular leukomalacia reflects significant injury to immature cerebral white matter and is strongly associated with later cerebral palsy, especially spastic diplegia. Less obvious noncystic white-matter injury can also affect neurodevelopment.

191. B. High-risk infants benefit from early recognition of abnormal motor patterns and timely referral to developmental and therapy services. Intervention can address function and family goals even before a final diagnostic label is established.

192. A. Subcutaneous fat necrosis produces firm inflammatory plaques or nodules in term or post-term infants after perinatal stress or therapeutic cooling. The lesions often resolve, but delayed hypercalcemia can be clinically important.

193. B. Turner syndrome should be considered in a female newborn with hand and foot lymphedema, webbed neck, or left-sided obstructive cardiac disease. Karyotype or chromosomal testing confirms the diagnosis.

194. D. Both hypoxemia and hyperoxia can harm extremely preterm infants. Oxygen is therefore titrated to unit targets with continuous monitoring rather than maximized indiscriminately.

195. B. Pierre Robin sequence causes upper-airway obstruction from micrognathia and posterior tongue displacement. Severity varies, so management ranges from monitored positioning and feeding support to nasopharyngeal airway, positive pressure, mandibular distraction, or tracheostomy.

196. D. The adducted, internally rotated shoulder with extended elbow and preserved grasp is typical of an upper brachial plexus palsy. Recovery depends on injury severity, and serial motor examination plus early therapy and specialty follow-up are important.

197. A. Periviable counseling should be individualized, transparent about uncertainty, and sensitive to outcomes that matter to the family. Shared decision-making is especially important when more than one ethically acceptable course exists.

198. A. NSAIDs close the ductus by inhibiting prostaglandin synthesis, but prostaglandins also support renal and other organ blood flow in neonates. Reduced renal perfusion and oliguria are therefore recognized adverse effects requiring monitoring.

199. B. Phenobarbital remains widely recommended as first-line therapy for many acute symptomatic neonatal seizures. Etiology matters, because sodium-channel blockers may be favored for certain genetic channelopathies, and ongoing EEG is important to judge response.

200. A. Meconium aspiration can obstruct airways, trigger inflammation, inactivate surfactant, and contribute to air leak and persistent pulmonary hypertension. The post-term setting, meconium exposure, coarse infiltrates, and hyperinflation are characteristic.