SPECIALTY REVIEW IN NEONATOLOGY
OFFICIAL ANSWER KEY TO THE
PRETEST EXAMINATION

If you disagree with any of the answers, please contact Matthew Saxonhouse at matthew.saxonhouse@atriumhealth.org. As with any examination, there may be an alternative answer that you may feel should be correct. If this is the case, please contact the email address provided and Dr. Saxonhouse would be happy to discuss the question with you. The purpose of this examination is not to see how high of a score you can achieve, but rather to develop an understanding of the material that will be covered during the course and areas of weakness that you have going into the course.

1) B
One of the more common malformations, single umbilical arteries are observed in 0.5 – 1.0% of all infants. They are 3-4 times more common in twins compared with singletons and they may be associated with urogenital tract or cardiac anomalies. The presence of other anomalies on exam does warrant a more detailed evaluation consisting of chromosomal analysis and/or renal US.
Reference: Vogt BA, MacRae Dell K, The kidney and urinary tract. In, Neonatal-Perinatal Medicine. Martin RJ, Fanaroff AA, Walsh MC, eds. Elsevier. 2011. 9th edition. Pgs 1686-1687.

2) D
The woman presented in the vignette has demonstrated multiple diastolic BPs ≥ 90mmHg. The definition of preeclampsia is: DBP ≥ 90 mmHg or SBP ≥ 140 mm Hg documented more than twice; severe if DBP ≥ 110 mmHg or SBP ≥ 160 mm Hg. She does not have any prior medical history thus not supporting a diagnosis of superimposed preeclampsia. She has no other findings consistent with HELLP syndrome
Reference: Hypertensive disorders in pregnancy. In, Williams Obstetrics. Cunningham et al, eds. McGraw-Hill. 2005. 22nd Edition. 761-808.

3) C
The ultrasound (US) findings demonstrate images consistent with a double bubble, or duodenal atresia. Of note, US images displayed on a sheet of paper may not be of the best quality but it is important to be familiar with the appearance of the more common lesions. Being that this question is a 3rd level question, one must also be familiar with the triple screen findings that support Trisomy 21. Of the choices, choice C is correct. If you could not identify the lesion, then just knowing that only Trisomy 21 or 18 are supported by triple screen findings, this could narrow your choices to either C or D. Choice D would support Trisomy 18.
Reference: Prenatal diagnosis and fetal therapy. In, Williams Obstetrics. Cunningham et al, eds. McGraw-Hill. 2005. 22nd Edition. 323.

4) C
The fetal heart rate tracing and mother's contractions represent late decelerations. Late decelerations are usually due to uteroplacental insufficiency and demonstrate fetal hypoxemia. Thus, choice C is the best answer.
Reference: Intrapartum assessment. In, Williams Obstetrics. Cunningham et al, eds. McGraw-Hill. 2005. 22nd Edition. 451-453.

5) D
The demonstration of higher post-ductal saturations supports a diagnosis of Transposition of the great arteries with elevated pulmonary pressures. This finding in combination with the others listed strongly supports fetal exposure to Isotretinoin (Retinoic Acid).
Reference: Jones KL. *Smith's Recognizable Patterns of Human Malformation, 6th edition pgs. 660-661.*

6) B
Cardiac abnormalities (structural or arrhythmias) represent ~ 25% of all cases of non-immune hydrops fetalis. The fetus presented in the vignette may have had an arrhythmia that has recently resolved prior to the ultrasound but resulted in the findings.
Reference: Wolf RB, Moore TR. Amniotic fluid and nonimmune hydrops fetalis. In, Neonatal-Perinatal Medicine. Martin RJ, Fanaroff AA, Walsh MC, eds. Elsevier. 2011. 9th edition. Pgs 390-397.

7) B
When performing resuscitation in any newborn infant, proper guidelines published from the neonatal resuscitation program should be followed. For this particular infant, you do not know what was performed prior to your arrival. Rather, you need to start from the beginning and from the choices listed, choice B is the best option.
Reference: Katwinkel J (ed). Textbook of neonatal resuscitation. 6th edition. AAP and AHA; 2010.

8) C
Intrauterine hypoxia initially results in increased blow flow to the heart, brain, and adrenal glands.
Reference: Katwinkel J (ed). Textbook of neonatal resuscitation. 6th edition. AAP and AHA; 2010.

9) A
Based on the one-minute examination, the infant's APGAR score is 3. See Table below:

Clinical Sign	Score = 0	Score = 1	Score = 2
Respirations	None	Gasping, poor, irregular	Strong cry
Heart Rate	None	< 100 bpm	> 100 bpm
Color	Cyanotic	Acrocyanosis	Pink
Muscle Tone	Floppy	Some flexion of extremities	Active
Reflex Irritability	None	Grimace	Cough, sneeze, or cry

Reference: Reference: Katwinkel J (ed). Textbook of neonatal resuscitation. 6th edition. AAP and AHA; 2010.

10) C
Although following standard NRP guidelines would recommend initiating positive pressure ventilation via bag and mask ventilation, the infant presented in the vignette is unique in that you are suspecting a diagnosis of congenital diaphragmatic hernia. NRP recommendations for these infants are to place an endotracheal tube immediately with appropriate gastric decompression.
Reference: Katwinkel J (ed). Textbook of neonatal resuscitation. 6th edition. AAP and AHA; 2010.

11) B
A. Blood returning from the placenta to the fetus by way of the umbilical vein, normally has a pO2 of **20-30** mmHg.
B. The most oxygenated blood in the fetus travels from the placenta via the umbilical vein into the inferior vena cava and ~ 1/3 of this blood travels through the foramen ovale to the left atrium then to the left ventricle and out the aorta to the major blood vessels of the brain.
C. Blood returning from the brain, via the superior vena cava, is directed primarily from the right atrium to the right ventricle, through the pulmonary artery **and the patent ductus arteriosus to the descending aorta.**
D. Approximately **45%** of the descending aorta blood flow goes to the placenta by way of the umbilical arteries.
E. The pulmonary circulation receives approximately **7%** of the fetal blood volume.
Reference: Fyler DC (ed): Nadas' Pediatric Cardiology. Philadelphia, Hanley & Belfus, 1991, pg. 58.

12) B
The infant presented in the vignette clearly has findings of cyanotic congenital heart disease. Although all of the choices may present with cyanosis, the single second heart sound and left superior axis on ECG strongly support the diagnosis of tricuspid atresia.
Reference: Zahka KG, Erenberg F. Congenital defects. In, Neonatal-Perinatal Medicine. Martin RJ, Fanaroff AA, Walsh MC, eds. Elsevier. 2011. 9thth edition. Pgs 1245-1266.

13) D
The findings listed support a congenital infection with either Rubella or CMV. Infants with congenital rubella syndrome, if infected < 8-weeks of gestation, have a 50% chance of cardiac disease with patent ductus arteriosus being most common.
Reference: see below

14) C
The clinical findings presented support a diagnosis of Williams Syndrome. The most common cardiac lesion in infant with Williams Syndrome is supravalvular subaortic stenosis.
Reference for 13 and 14: Jones KL. *Smith's Recognizable Patterns of Human Malformation, 6th edition pgs. 120-123.*

15) D
Although the infant presented in the vignette most likely has a ductal dependent congenital heart lesion, such as the hypoplastic left heart syndrome or severe coarctation of the aorta, one should not assume that this is the diagnosis until proper evaluation by a Cardiologist is performed (unless a prenatal diagnosis was obtained). It is important for the outside physician to also entertain the diagnoses of severe neonatal sepsis or an inborn error of metabolism. To hopefully begin stabilization of this critical infant, the following procedures should be performed (if the clinician is able to): stabilization of airway (if PGE is to be started prior to transfer, then intubation is recommended), proper fluid resuscitation, IV infusion of PGE, termination of all protein intake with IV infusion of carbohydrates only, and immediate start of Ampicillin and Gentamicin. By following this algorithm, you have entertained all three diagnoses and initiated proper initial stabilization of this potentially very critical infant.
Reference: Katwinkel J (ed). Textbook of neonatal resuscitation. 6th edition. AAP and AHA; 2010.

16) D
The formula for oxygen consumption is the Fick principle
= CO (dL/min) x (CaO2 – CvO2)
= CO x (1.34 cc/g Hb) (Hb concentration) (arterial sat – venous sat)
CO = cardiac output
CaO2 = oxygen content of arterial blood
CvO2 = oxygen content of mixed venous blood

For this question, all of the answers provided are in dL/minute reminding you to change the units from what you are given. You are provided with the cardiac output in L/min and need to convert this to dL/min before proceeding. In addition, the saturation values provided should be converted to decimal points to answer the question properly. Using the values provided, the question should be answered as:
1.4 L/min = 14 dL/min
14g/dl x (1.34 x 14) (.97-.63) = 14 x 18.8 x .34 = 89.5 dL/min
Reference: Zahka KG. Principles of neonatal cardiovascular hemodynamics. In, Neonatal-Perinatal Medicine. Martin RJ, Fanaroff AA, Walsh MC, eds. Elsevier. 2011. 9th edition. Pgs 1233-1234; Neonatology Review. Brodsky D, Martin C. Hanley & Belfus, INC. 2010. Formulas in Appendix section.

17) B
D-Transposition of the Great Arteries is the most common cyanotic heart defect identified in the first week of life. This fact alone allows you to answer the question correctly. The additional findings of higher saturations in the right foot compared with the right hand also strongly supports this diagnosis. Elevated pulmonary pressures with shunting of oxygenated blood from the pulmonary artery to the descending aorta create these clinical findings.
Reference: Zahka KG, Erenberg F. In, Neonatal-Perinatal Medicine. Martin RJ, Fanaroff AA, Walsh MC, eds. Elsevier. 2011. 9th edition. Pgs 1245-1247.

18) D
Central cyanosis occurs when the concentration of deoxygenated hemoglobin exceeds 3g/dL. Therefore, if the infant in this question has a hemoglobin of 12g/dL, then they would first appear cyanotic if > 3g/dL was deoxygenated. The saturation would be 12 – 3 / 12 = 9/12 = 75%.
Reference: Hansen TN, Weisman LE (eds): Neonatal Respiratory Disease (3rd edition). Pennsylvania, Hanbooks in Health Care Co, 200, pg 57.

19) C
The EKG presented demonstrates a prolonged QT interval. Of the choices provided, hypocalcemia is the best answer, as hypercalcemia may result in a shortened QT interval.
Reference: Sperling, M. Pediatric Endocrinology. 3rd ed. Philadelphia. Saunders; 2008.

20) A

The infant presented in the vignette likely has either esophageal atresia with a distal tracheo-esophageal fistula or isolated esophageal atresia. The esophagus and trachea first appear at the 21st day of gestation as a median ventral diverticulum of the primitive pharynx. By 34-36 days' gestation, lateral ridges divide the diverticulum to form the trachea and esophagus. Tracheoesophageal anomalies occur at this age, during the embryonic stage of lung development.

Reference: Hansen TN, Weisman LE (eds): Neonatal Respiratory Disease (3rd edition). Pennsylvania, Hanbooks in Health Care Co, 2003, pg 222..

21) B

The infant in the vignette is displaying symptoms of respiratory distress that immediately respond to supplemental oxygen. Choices C and D represent fixed shunts that would not respond to oxygen therapy. If the infant was sedated and not breathing, raising the oxygen concentration would not necessarily raise the oxygen saturations. Therefore, choices B and E remain. Respiratory distress syndrome (RDS) is rare in a term infant plus no maneuvers were provided to improve the compliance of the stiff lungs that are caused by RDS. Thus, choice B is the best answer.

Reference: Hansen TN, Weisman LE (eds): Neonatal Respiratory Disease (3rd edition). Pennsylvania, Hanbooks in Health Care Co, 200, pgs 31-35.

22) C

The effect of altitude on the paO_2 (arterial oxygen) can best be solved by the following equation:

$(pB\#1 - pH20) \times FiO_{2\#1} = (pB\#2 - pH20) \times FiO_{2\#2}$

Using the values provided:

(687 – 47) x .55 = (760-47) x (? Value) =

352 = 713 (?)

(?) = 0.494 = 49% oxygen

Reference: Neonatology Review. Brodsky D, Martin C. Hanley & Belfus, INC. 2010. Formulas in Appendix section

23) B

Reference: Hansen TN, Weisman LE (eds): Neonatal Respiratory Disease (3rd edition). Pennsylvania, Hanbooks in Health Care Co, 2003, pgs 288-291.

24) C
To properly answer this question, the following formulas should be used:
Oxygen delivery to alveoli (cc/kg/minute) = alveolar minute ventilation x FiO2
alveolar minute ventilation = tidal volume x respiratory rate

Using the values provided from the question:
Alveolar minute ventilation = 9 x 45 = 405
Oxygen delivery to alveoli = 405 x 0.25 = 101.25 = 101 ml oxygen per minute
Reference: Neonatology Review. Brodsky D, Martin C. Hanley & Belfus, INC. 2010. Formulas in Appendix section

25) E
Reference: Hansen TN, Weisman LE (eds): Neonatal Respiratory Disease (3rd edition). Pennsylvania, Hanbooks in Health Care Co, 2003, pg 45.

26) A
SP-A is the most abundant of the surfactant proteins constituting nearly 5% by weight of the surfactant.
Reference: Hansen TN, Weisman LE (eds): Neonatal Respiratory Disease (3rd edition). Pennsylvania, Hanbooks in Health Care Co, 2003, pgs 45-47.

27) A
Oxygen carrying capacity is equal to the oxygen content of blood. The formula for oxygen content is equal to oxygen bound to hemoglobin (Hgb) plus dissolved oxygen or O_2 bound to Hgb plus dissolved O_2

O_2 bound to Hgb = [(1.34 cc O_2 /g Hgb) x Hgb (mg/dL) x oxygen saturation
Dissolved O_2 = [0.003 cc O_2/ dL torr) x pa O_2 (torr)]

Increasing cardiac output will not change the oxygen content, only oxygen delivery. This eliminates choices C and D. Using the values provided in the question, the oxygen content is:
1.34 x 10 x 0.92 + .003 x 60 = 12.3 + 0.18 = 12.5.

If we change the Hgb to 15 mg/dL, the content increases to:
1.34 x 15 x 0.92 + .003 x 60 = 18.5 + 0.18 = 18.7 (choice A).

If we increase to PaO_2 to 400, the content increases to:
1.34 x 10 x 1 *(assume saturation 100% if PaO2 400)* + 0.003 x 400 = 13.4 + 1.2 = 14.6 (choice B)

Therefore, choice A is the best answer.
Reference: Hansen TN, Weisman LE (eds): Neonatal Respiratory Disease (3rd edition). Pennsylvania, Hanbooks in Health Care Co, 2003, pg 25; Neonatology Review. Brodsky D, Martin C. Hanley & Belfus, INC. 2010. Formulas in Appendix section

28) A
Factors affecting Oxyhemoglobin Dissociation Curve

Shift Curve to Left *Impair oxygen delivery/increase oxygen affinity*	**Shift Curve to Right** *Improve oxygen delivery/decrease oxygen affinity*
Alkalosis	Acidosis
Decreased temperature	Increased temperature
Decreased 2,3-DPG concentrations	Increased 2,3-DPG concentrations
Increased fetal Hemoglobin	Decreased fetal Hemoglobin

Reference: Hansen TN, Weisman LE (eds): Neonatal Respiratory Disease (3rd edition). Pennsylvania, Hanbooks in Health Care Co, 2003, pg 25.

29) E
Oxygenation Index =
{Mean airway pressure x inspired oxygen concentration / PaO2} x 100

{20 x 1 / 39} x 100 = 51.3 = 51.
Reference: Carlo W, Di Fiore JM. Assessment of pulmonary function. In, Neonatal-Perinatal Medicine. Martin RJ, Fanaroff AA, Walsh MC, eds. Elsevier. 2011. 9th edition. Pg 1095.

30) D
The prenatal risk factors and emergent delivery plus the infant's clinical symptoms support a diagnosis of respiratory distress syndrome. In addition, the chest x-ray demonstrates ground glass appearance, air bronchograms, and decreased lung volumes. However, it is important to pay close attention to all of the details of the chest x-ray as the ETT is also in too far and probably contributing to the infant's distress.
Reference: Hamvas A. Pathophysiology and management of respiratory distress syndrome. In, Neonatal-Perinatal Medicine. Martin RJ, Fanaroff AA, Walsh MC, eds. Elsevier. 2011. 9th edition. Pgs 1106 – 1116.

31) C
Close attention to the clinical history and CXR findings demonstrate a pleural effusion on the patient's right side as the cardiac silhouette is displaced towards the left. The PICC line on the right is not in good position thus supporting the diagnosis of a pleural effusion from TPN given the infant's NPO status.
Reference: Abu-Shaweesh JM. Respiratory disorders in preterm and term infants. In, Neonatal-Perinatal Medicine. Martin RJ, Fanaroff AA, Walsh MC, eds. Elsevier. 2011. 9th edition. Pgs 1153 – 1154.

32) C

The CXR demonstrates cystic changes on the infant's left side. Although this can be mistaken for intestines thus supporting a diagnosis of a left sided congenital diaphragmatic hernia, review of the CXR demonstrates a normally located stomach bubble with NG tube in place. In addition, a normal bowel gas pattern is demonstrated. These findings support a diagnosis of a CPAM.

Reference: Reference: Abu-Shaweesh JM. Respiratory disorders in preterm and term infants. In, Neonatal-Perinatal Medicine. Martin RJ, Fanaroff AA, Walsh MC, eds. Elsevier. 2011. 9th edition. Pgs 1154 – 1155.

33) D

Please see the table below describing the laboratory values for the different urea cycle defects:

Enzyme Deficiency	Orotic Acid	Glutamine	Alanine	Citrulline	Arginine
N-acetylglutamate Deficiency	Normal or low				
Carbamyl phosphate synthetase	Normal or low	Increased	Increased	Decreased	Decreased
Ornithine carbamyl transferase	Increased	Increased	Increased	Decreased	Decreased
Arginosuccinic acid synthetase (citrullinemia)	Increased			Increased	Decreased
Arginase	Increased				
Arginosuccinic lyase	Increased			Normal	Decreased

Reference: Zinn AB. Inborn errors of metabolism. In, Neonatal-Perinatal Medicine. Martin RJ, Fanaroff AA, Walsh MC, eds. Elsevier. 2011. 9th edition. Pgs 1673-1677.

34) B

The infant described in the vignette has features consistent with one of the Mucopolysaccharidoses. The majority of these are inherited in an autosomal recessive pattern. However, the family history provided supports an inheritance consistent with X-linked inheritance. Hunter syndrome is the only Mucopolysaccharidosis inherited in a X-linked pattern thus supporting choice B.

Reference: Jones KL. Recognizable patterns of human malformation. 1997. Sixth edition. Elsevier Saunders. Pages 518-534.

35) B
See below

36) D
Reference for 35 and 36: Jones KL. Recognizable patterns of human malformation. 1997. Sixth edition. Elsevier Saunders. Pages 478-479; 88-89.

37) B
The mother of the infant has a sister with Carpenter syndrome. We can assume that she does not have the syndrome; therefore, we need to determine her risk of being a carrier for the syndrome. Because it is an autosomal recessive disorder, her chance of being a carrier is 2/3 or 0.67.

The chance that the father of the baby is a carrier (we can also assume that he does not have the syndrome as that information was not provided to us) can be calculated by using the Hardy-Weinberg equation. The disease incidence of an autosomal recessive disorder is q^2. Therefore:
$q^2 = 1/9000$, so q equals 1/94.9 and p equals 93.9/94.9, as p + q = 1. The carrier frequency is 2pq = 2 (93.9/94.9) (1/94.9) = 2 (0.99)(0.01) = 0.0198 = 0.02 is the probability he is a carrier.
Applying the fact that the mother's chance of being a carrier is 0.67 and the father's chance is 0.02, they then have a 25% chance of having a neonate with Carpenter syndrome.
So: 0.67 x 0.02 x 0.25 = 0.003 = 0.3%.
References: 1. Genetics in Medicine Thompson and Thompson, Chapter 9
2. Clinical Genetics by Andrew Read and Dian Donnai Chapter 10

38) C
See below
39) A
Reference for 38 and 39: Jones KL. Recognizable patterns of human malformation. 1997. Sixth edition. Elsevier Saunders. Pages 198-199; 64.

40) C
Homocysteine is not considered an essential amino acid.
Reference: Poindexter B, Denne S. Nutrition and metabolism in the high-risk neonate. Neonatal-Perinatal Medicine. Martin RJ, Fanaroff AA, Walsh MC, eds. Elsevier. 2011. 9th edition. Pgs 651-668.

41) B
Reference: Textbook of Human Lactation, Hale & Hartmann: Chapter 4.

42) B
A 10-day old, 34-week infant with symptoms of respiratory distress should be receiving anywhere from 100-140 ml/kg/day. Based on this, choices D and E can be eliminated. Choice A is on the restricted side but not totally incorrect. If an infant is receiving only TPN, then his/her caloric intake should be about 70-100 kcal/kg/day and should follow the rule that about 50% of the total calories should be from carbohydrates, 40% should be from fat, and ~ 10% from protein. Applying these principles, choice B is the best answer. Choice C is close but provides 150 ml/kg/day and 90 kcal/kg/day with 62% of total calories from carbohydrates.
Reference: AAP Pediatric Nutrition Handbook , 2009

43) B
Reference: Textbook of Human Lactation, Hale & Hartmann: Chapter 4.

44) E
Reference: McRae Dell K. Fluids, electrolytes, and acid-base homeostasis. In, Neonatal-Perinatal Medicine. Martin RJ, Fanaroff AA, Walsh MC, eds. Elsevier. 2011. 9th edition. Pgs. 669-684.

45) C
Reference: Poindexter B, Denne S. Nutrition and metabolism in the high-risk neonate. Neonatal-Perinatal Medicine. Martin RJ, Fanaroff AA, Walsh MC, eds. Elsevier. 2011. 9th edition. Pgs 651-668.

46) D
Reference: Backman DA, Fawcett LB, Brent RL. The effects of maternal drugs on the developing fetus. In, Avery's Neonatology-Pathophysiology & management of the newborn. MacDonald MG, Mullett MD, Seshia M, eds. Lippincott Williams & Williams. 2005. 6th edition. Pg 251.

47) C
Reference: Behman RE, Kliegman RM, Arvin AM (eds): Nelson Textbook of Pediatrics (15th edition). Philadelphia, WB Saunders Co, 1996, p. 146-147.

48) B
Na+ deficit (mEq) = [Na+ desired (mEq/L) – Na+ current (mEq/L)] x 0.6 x weight (kg) = [130 – 114] x 0.6 x 0.85kg = 16 x 0.6 x 0.85 = 8.2 mEq.
Reference: Andreoli SP. Renal failure in the newborn infant. In, Workbook in Practical Neonatology. Polin RA, Yoder MC, Burg FD, eds. 3rd Edition. 2001. Saunders. Pgs 322-338.

49) D
Point a = respiratory alkalosis
Point b = respiratory acidosis with metabolic compensation
Point c = normal
Point d = metabolic acidosis with respiratory compensation
Point e = metabolic acidosis
Point f = respiratory acidosis
Point g = metabolic alkalosis
Reference: Hansen TN, Weisman LE (eds): Neonatal Respiratory Disease (3rd edition). Pennsylvania, Hanbooks in Health Care Co, 2003.

50) B
The infant described in the vignette has a normal anion gap acidosis.
$Na + K – Cl + HCO_3^- = 125 + 5 – 110 + 15 = 5.$
The infant's urine pH is 4.5 supporting a diagnosis of type II or proximal renal tubular acidosis. The pathology is due to decreased or absent proximal tubular reabsorption of HCO_3^- with normal distal acidification. If the urine pH was > 6.2, then this would support a diagnosis of type I or distal renal tubular acidosis in which H+ cannot be secreted in the distal tubule.
Reference: McRae Dell K. Fluids, electrolytes, and acid-base homeostasis. In, Neonatal-Perinatal Medicine. Martin RJ, Fanaroff AA, Walsh MC, eds. Elsevier. 2011. 9th edition. Pgs. 681-683.

51) B
The calculation for the fractional excretion of Na or FE_{Na} =
(Urine Na x Plasma Cr) / (Urine Cr x Plasma Na) x 100
Using the values provided = (2 x 1) / (147 x 1) = 0.014 x 100 = 1.4%

Interpretation for neonates: < 1% is normal; 1-2.5% is pre-renal; > 3% is intrinsic renal failure. Thus, choice B.
Reference: Andreoli SP. Renal failure in the newborn infant. In, Workbook in Practical Neonatology. Polin RA, Yoder MC, Burg FD, eds. 3rd Edition. 2001. Saunders. Pgs 322-338.

52) C
At first, it appears that the infant has congenital adrenal hyperplasia based on the low Na and elevated K. However, the infant's aldosterone level is elevated supporting a diagnosis of pseudohypoaldosteronism. In CAH, aldosterone levels are low. K+ is also usually elevated to a greater extent than in this example.
Reference: McRae Dell K. Fluids, electrolytes, and acid-base homeostasis. In, Neonatal-Perinatal Medicine. Martin RJ, Fanaroff AA, Walsh MC, eds. Elsevier. 2011. 9th edition. Pg. 675.

53) C

The infant presented in the vignette has signs and symptoms consistent with Galactosemia. Galactosemia is a deficiency in the enzyme galactose-1-phosphate-uridyltransferase. Galactokinase deficiency may present with cataracts, but elevated blood glucose values exist.

Reference: Zinn AB. Inborn errors of metabolism. Neonatal-Perinatal Medicine. Martin RJ, Fanaroff AA, Walsh MC, eds. Elsevier. 2011. 9th edition. Pgs 1621-1679.

54) D

The profound metabolic acidosis and slightly elevated ammonia level are most consistent with an organic acidemia. A urea cycle defect, if caught early, would present with a respiratory alkalosis and severely elevated ammonia level. The lack of renal failure and elevated ammonia levels rules against a ductal dependent congenital heart lesion.

Reference: Zinn AB. Inborn errors of metabolism. Neonatal-Perinatal Medicine. Martin RJ, Fanaroff AA, Walsh MC, eds. Elsevier. 2011. 9th edition. Pgs 1621-1679.

55) C

Placing an infant in a double-walled incubator, compared with a single-walled incubator reduces the amount of radiant heat loss.

Reference: Sedin G. The thermal environment. In, Neonatal-Perinatal Medicine. Martin RJ, Fanaroff AA, Walsh MC, eds. Elsevier. 2011. 9th edition. Pgs 555-569.

56) B

The infant has a relatively normal serum calcium value but low serum phosphorous value. In addition, the alkaline phosphatase level is significantly elevated. The infant is also only on fortified breast milk feedings and not a multivitamin. These findings are most consistent with vitamin D deficiency. The infant is likely receiving a normal amount of phosphorous but likely using it for bone growth reflecting the low serum value.

Reference: Sperling, M. Pediatric Endocrinology. 3rd ed. Philadelphia. Saunders; 20

57) D

Nephrogenic DI	Neurogenic DI
Increased UOP due to failure of kidneys to respond to ADH	Increased UOP due to inadequate production of ADH
Increased plasma osmolality	Increased plasma osmolality
Decreased urine osmolality	Decreased urine osmolality
Normal to high ADH levels	Low ADH levels
No change in urine osmolality after fluid restriction	No change in urine osmolality after fluid restriction
Increase in ADH levels after fluid restriction	No change in ADH levels after fluid restriction
No change in urine osmolality after ADH administration	Increase in urine osmolality after ADH administration

Reference: Sperling, M. Pediatric Endocrinology. 3rd ed. Philadelphia. Saunders; 2008.

58) D
The findings of hypocalcemia and hyperphosphatemia suggest primary hypoparathyroidism. However, the infant has an elevated PTH level supporting pseudohypoparathyroidism.
Reference: Sperling, M. Pediatric Endocrinology. 3rd ed. Philadelphia. Saunders; 2008.

59) C
The clinical findings support a diagnosis of Zellweger Syndrome or cerebro-hepato-renal syndrome. This is a rare autosomal recessive syndrome marked by the absence of hepatic and renal peroxisomes. Increased levels of very long chain fatty acids confirm the diagnosis in suspected infants.
Reference: Jones KL. Recognizable patterns of human malformation. 1997. Sixth edition. Elsevier Saunders. Pages 238-239.

60) A
LAD-1 deficiency may present in the neonatal period. A history of recurrent infections, delayed separation of the umbilical cord, leukocytosis, and absence of pus supports this diagnosis. Confusion may be made with Chronic Granulomatous Deficiency as this may also present in the neonatal period. However, recurrent infections with catalase positive bacteria or fungi strongly supports this diagnosis.
Reference: Saxonhouse MA, Sleasman JW: Immunodeficiency Diseases of the Neonate, In Neonatal Hematology, DeAlarcon PA and Werner EJ, editors. Cambridge University 2005. Pgs 280-309.

61) C
Although all of the immunoglobulin levels evaluated are low, they fall within the expected ranges for neonates of this post-conceptional age. Whether or not they predispose the infant to infection remains to be determined. Thus, these values represent the expected nadir of immunoglobulin levels for neonates of this post-conceptional age.
Reference: Saxonhouse MA, Sleasman JW: Immunodeficiency Diseases of the Neonate, In Neonatal Hematology, DeAlarcon PA and Werner EJ, editors. Cambridge University 2005. Pgs 280-309.

62) D
The persistent symptoms of diarrhea, failure to thrive, and scaly eruption combined with the eosinophilia strongly support a diagnosis of severe combined immunodeficiency disease (SCID).
Reference: Saxonhouse MA, Sleasman JW: Immunodeficiency Diseases of the Neonate, In Neonatal Hematology, DeAlarcon PA and Werner EJ, editors. Cambridge University 2005. Pgs 280-309.

63) D
Due to the fact that gonococcal ophthalmia or disseminated infection occasionally can occur in infants born to mothers with gonococcal infections, infants born to mothers known to have gonorrhea should receive a single dose of IV Ceftriaxone. Cefotaxime may also be used if there are concerns with hyperbilirubinemia.
Reference. Pickering LK (ed). Red Book: 2009 Report of the Committee on Infectious Diseases. 28th Edition, 2009.

64) D
Due to difficulties in eradication of Candidal sepsis when a central line is in place, it is recommended that all central lines be removed if an infant has candidal sepsis. Another central catheter may be placed after 3-negative blood cultures.
Reference. Pickering LK (ed). Red Book: 2009 Report of the Committee on Infectious Diseases. 28th Edition, 2009.

65) D
Newborn infants are at minimal risk for syphilis if they are born to mothers who completed appropriate penicillin treatment for syphilis more than 4-weeks before delivery, the mother had an appropriate response to treatment (documented fourfold or greater decrease in VDRL), and mother has no evidence of reinfection or relapse. The mother presented in the vignette received adequate treatment with an appropriate VDRL response.
Reference. Pickering LK (ed). Red Book: 2009 Report of the Committee on Infectious Diseases. 28th Edition, 2009.

66) C
The infant presented in the vignette is very concerning for neonatal sepsis. Based on the infant's age, Ampicillin and Gentamicin are recommended. However, the mother's history of fever and chills is concerning for a viral infection. In addition, the infant is lethargic, febrile, and jaundiced raising suspicions for HSV infection. Therefore, adequate fluid resuscitation, antibiotics, and antiviral therapy is strongly recommended.
Reference. Pickering LK (ed). Red Book: 2009 Report of the Committee on Infectious Diseases. 28th Edition, 2009.

67) D
Infants with congenital toxoplasmosis typically present with hydrocephalus and cortical calcifications. Infants with congenial CMV demonstrate periventricular calcifications. Both may demonstrate mild hepatitis and thrombocytopenia.
Reference: Fanaroff AA, Martin RJ (eds): Neonatal-perinatal medicine (6th edition). St Louis, Mosby-Year Book Inc, 1997, p 768.

68) E

The first four choices, although theoretical risks for breast feeding are not contraindications. Choice E is an absolute contraindication for breast feeding. Mother's with active TB and an abnormal CXR should be isolated from their infants until the mother has been appropriately evaluated and both the mother and infant are receiving appropriate antituberculosis therapy, the mother wears a mask, and the mother understands and is willing to adhere to infection control measures.

Reference. Pickering LK (ed). Red Book: 2009 Report of the Committee on Infectious Diseases. 28th Edition, 2009.

69) B

Most abdominal masses presenting in the neonatal period are of renal origin. They include hydronephrosis, renal dysplasia, and polycystic kidney disease.

Reference: Neonatology Review. Brodsky D, Martin C. Hanley & Belfus, INC. 2003. Pg 232.

70) C

The abdominal wall defect described is a gastroschisis. Unlike neonates with omphalocele, infants with gastroschisis usually do not have other nongastrointestinal abnormalities. However, infants with gastroschisis have about a 16% chance of having other gastrointestinal abnormalities such as intestinal atresia, midgut volvulus, and intestinal stenosis.

Reference: Barksdale EM, Chwals WJ, Magnuson DK, and Parry RL. Selected gastrointestinal anomalies. In, Neonatal-Perinatal Medicine. Martin RJ, Fanaroff AA, Walsh MC, eds. Elsevier. 2011. 9th edition. Pgs 1400-1431.

71) C

Lactase levels reach adult levels by 36-weeks gestation.

Reference: Neonatology Review. Brodsky D, Martin C. Hanley & Belfus, INC. 2010. page 274.

72) D

The infant presented in the vignette is a late preterm infant with significant hyperbilirubinemia due to hemolytic anemia. In addition, the infant demonstrates findings consistent with the intermediate phase of acute bilirubin encephalopathy. Infants who demonstrate physical findings at this point should receive a double volume exchange transfusion to prevent the irreversible, advance phase of acute bilirubin encephalopathy and kernicterus.

Reference: AAP *Subcommittee on Hyperbilirubinemia. Clinical Practice Guideline. Management of Hyperbilirubinemia in the Newborn Infant 35 or More Weeks of Gestation. PEDIATRICS Vol. 114 No. 1 July 2004, pp. 297-316.*

73) D

The optimal volume for an exchange transfusion is twice the infant's blood volume. For a term infant, blood volume is about 80-85 ml/kg. For a preterm infant, it is 100 ml/kg. The infant in the vignette is a full-term infant. Using 80 ml/kg, the appropriate volume for an exchange transfusion would be 640 ml. An exchange transfusion results in removal of about 85% of the neonate's red blood cells.

Reference: Luchtman-Jones L, Wilson DB. Hematologic problems in the fetus and neonate. In, Neonatal-Perinatal Medicine. Martin RJ, Fanaroff AA, Walsh MC, eds. Elsevier. 2011. 5th edition. Pgs 1303-1373.

74) D

Line A is alpha globin chain; Line B is the gamma globin chain, Line C is the beta globin chain, and line D is the delta globin chain.

Reference: de Alarcon PA, Johnson MC, Werner EJ. Eryhropoiesis, red cells, and the approach to anemia. In, Neonatal Hematology. De Alarcon PA and Werner E, eds. Cambridge University Press. 2005. Pages 40-57.

75) D

Complete absence of Protein S or C activity may result in purpura fulminans in the neonate. In addition, factor V Leiden mutation may also present in this fashion.

Reference: Albisetti M, Andrew M, Monagle P. Hemostatic abnormalities. In, Neonatal Hematology. De Alarcon PA and Werner E, eds. Cambridge University Press. 2005. Pages 310-348.

76) C

The infant presented in the vignette most likely has late hemorrhagic disease of the newborn. The combination of the mother being a vegetarian, solely breast feeding, and not allowing the infant to receive vitamin K after birth is the likely reason for this illness. The lack of retinal hemorrhages also goes against shaken baby syndrome. In addition, late hemorrhagic disease of the newborn mostly presents with intracranial bleeding. Hemorrhagic disease of the newborn is supported by an isolated prolongation of the Prothrombin time, with normal partial thromboplastin times and normal platelet counts. Thus, choice C. Choice A represents hemophilia A or B; Choice B represents DIC and/or liver disease; Choice D represents isolated thrombocytopenia; and choice E demonstrates possible liver disease with thrombocytopenia or sepsis but is not specific for one illness.

Reference: Albisetti M, Andrew M, Monagle P. Hemostatic abnormalities. In, Neonatal Hematology. De Alarcon PA and Werner E, eds. Cambridge University Press. 2005. Pages 310-348.

77) C

Although there really is no standard of care for management of neonates with thromboses, the infant presented in the vignette has a definite organ/life threatening thrombosis. Current recommendations are to initiate low dose systemic rTPA and low dose heparin for life/organ/limb threatening thromboses.

Reference: Monagle et al. Antithrombotic therapy in neonates and children. CHEST. 2012. e737s – e801s.

78) B

The infant presented in the vignette likely has methemoglobinemia due to cytochrome-b5 reductase deficiency. If the level is > 40% (as in this infant), treatment should be with methylene blue at a dose of 1-2 mg/kg. Although methylene blue should not be given to infants with G6PD, the infant is stressed but demonstrates no evidence of hemolysis with a normal hemoglobin value.

Reference: Glader B, Allen G. Neonatal hemolysis. In, Neonatal Hematology. De Alarcon PA and Werner E, eds. Cambridge University Press. 2005. pgs. 132-162.

79) B

The infant presented in the vignette likely has *Staphylococcal* scalded skin syndrome. The majority of cases are caused by *Staphylococcal aureus* exotoxin. The history of facial erythema and a positive Nikolsky sign with a sterile culture from the fluid from the bullae strongly support the diagnosis.

Reference: Eichenfeld LF, Frieden IJ, Esterly NB (eds). Neonatal Dermatology. 2nd Edition. Saunders Elsevier. 2008.

80) D

The infant in the vignette has a complete gastric outlet obstruction as there is no air below the stomach bubble. A possible cause of this finding is pyloric atresia. The multiple blisters on the arms and legs are concerning for infection but could also be associated with a form of epidermolysis bullosa. In fact, a genetic cause has been identified for some cases of pyloric atresia that occur in association with epidermolysis bullosa lethalis (Herlitz and Carmi syndromes).

Reference: Safder S, Arora S, and Chelimsky G. Disorders of digestion. In, Neonatal-Perinatal Medicine. Martin RJ, Fanaroff AA, Walsh MC, eds. Elsevier. 2011. 5th edition. Pgs 1381-1400.

81) A

Premature closure of the sagittal suture results in Scaphocephaly or dolichocephaly. This is the most common type of craniosynostosis. Surgery is for cosmetic improvement and if no other lesions are present, usually not associated with increased intracranial pressure or other neurologic complications.

Reference: Neonatology Review. Brodsky D, Martin C. Hanley & Belfus, INC. 2010. page 130.

82) D
If the birth injury resulted in involvement of C4/5, then the resulting phrenic nerve paralysis will result in respiratory distress and decreased diaphragm movement with elevation of the hemidiaphragm on CXR.
Reference: Volpe J. Neurology of the newborn. Saunders Elsevier. 5th Edition. 2008. Pgs 972-976.

83) C
Reference: 1) Cloherty JP, Stark AR (ed): Manual of Neonatal Care (4th edition). Philadelphia, Lippincott-Raven, 1998. p. 537. 2) Volpe J. Neurology of the newborn. Saunders Elsevier. 5th Edition. 2008. Pgs 9-19.

84) B
A = Caput succedaneum; B = Cephalohematoma; C = Subgaleal hemorrhage; D = Extradural hematoma
Reference: Fletcher MA. Physical diagnosis in neonatology. Philadelphia, Lippincott-Raven, 1998, pg. 184.

85) D
Selective neuronal necrosis is the most common pattern of cerebral injury after hypoxic-ischemic encephalopathy. The clinical outcome is mental deficiency, seizures, ataxia, feeding difficulties, and pyramidal cerebral palsy.
Reference: Volpe J. Neurology of the newborn. Saunders Elsevier. 5th Edition. 2008. Pgs 348-356.

86) D
Extrapyramidal or athetoid cerebral palsy is classified as having mixed tone in the same muscle with gross and fine motor skills affected. Hearing deficits and speech abnormalities also exist.
Reference: Volpe J. Neurology of the newborn. Saunders Elsevier. 5th Edition. 2008. Pgs 430-438.

87) B
McCune-Albright Syndrome is characterized by multiple areas of fibrous dysplasia of the long bones but also may include the ribs and spine. Irregular brown pigmentation usually involves the sacrum, buttocks, and upper spine. In addition, infants suffer from hyperthyroidism, hyperparathyroidism, and pituitary adenomas.
Reference: Jones KL. *Smith's Recognizable Patterns of Human Malformation, 6th edition pgs. 594-595.*

88) C
The infant described in the vignette has findings consistent with spastic diplegia.
Reference: Volpe J. Neurology of the newborn. Saunders Elsevier. 5th Edition. 2008. Pgs 430-438.

89) C
Biphasic stridor is usually the result of laryngeal obstruction. It tends to worsen with agitation and it is the most common type of stridor in the neonatal period. Of the various causes of laryngeal obstruction, laryngomalacia is the most common laryngeal anomaly.
Reference: Sprecher RC, Arnold JE. Upper airway lesions. In, Neonatal-Perinatal Medicine. Martin RJ, Fanaroff AA, Walsh MC, eds. Elsevier. 2011. 9th edition. Pgs 1170-1179.

90) B
A decrease in cerebral blood flow may be caused by decreased paCO2, increased paO2, increased hemoglobin concentrations, and decreased fetal hemoglobin concentrations.
Reference: Volpe J. Neurology of the newborn. Saunders Elsevier. 5th Edition. 2008. Pgs 178-181.

91) B
First-order kinetics is characterized by the excretion of a certain percentage of drug per unit time and the rate of drug elimination is directly proportional to the serum drug concentration. Zero-order kinetics is characterized by the excretion of a constant amount of drug regardless of the serum drug concentration.
Reference: Boroujerdi M. Pharmokinetics: Principles and applications. New York: McGRaw-Hill; 2001.

92) B
Ranitidine inhibits the cytochrome P_{450} system. Theophylline is metabolized by the cytochrome P_{450} system. Due to the inhibition of the system by starting ranitidine, the infant's theophylline level would likely increase.
Reference: Boroujerdi M. Pharmokinetics: Principles and applications. New York: McGRaw-Hill; 2001.

93) D
As in most ethics' questions, there may be several answers that seem very reasonable. Providing comfort care and the administration of narcotics is done in many units and hospice centers. However, these procedures are usually carried out after some effort has been made between families, nursing staff, and ethics committees. Therefore, choice D is the most logical and safest answer and best addresses the interests of the family, infant, and staff.
Reference: Beauchamp TL, Childress JF. Principles of biomedical ethics. 5th edition. New York: Oxford University Press, Inc;2001.

94) C
Reference: Neonatology Review. Brodsky D, Martin C. Hanley & Belfus, INC. 2003. Pg. 398.

95) A
A type I error is when the null hypothesis is rejected when the null hypothesis is really true. This usually occurs when the sample size is very small.
Reference: Neonatology Review. Brodsky D, Martin C. Hanley & Belfus, INC. 2003.Pg 397.

96) C
Sensitivity is the probability of a test being positive when true disease is present.

Test	**Disease**	
	+	-
+	**9**	**21**
-	**5**	**9965**

9 / 9 + 5 = 64%
Reference: Neonatology Review. Brodsky D, Martin C. Hanley & Belfus, INC. 2003. Pg. 394.

97) D
The earliest finding of congenital glaucoma is an enlarged cornea.
Reference: Kaufman LM, Miller MT, Gupta BK. Examination and common problems. In, Neonatal-Perinatal Medicine. Martin RJ, Fanaroff AA, Walsh MC, eds. Elsevier. 2011. 9th edition. Pg 1748.

98) B
Odds ratio can be calculated by the following formula:
True positives x true negatives / false positives x false negatives
8 x 150 / 4 x 65 = 1200 / 260 = 4.6.
Reference: Neonatology Review. Brodsky D, Martin C. Hanley & Belfus, INC. 2003. Pg 393.

99) D
Cumulative incidence is the number of new cases in a given time period divided by the total population at risk. For this question, there were 19 cases in 22,000 infants born. 19/22000 = 0.0009
Reference: Neonatology Review. Brodsky D, Martin C. Hanley & Belfus, INC. 2003. Pg. 391.

100) E
Reference: Norman GR, Streiner DL. Biostatiscis: The Bare Essentials. 3rd ed. St. Louis:Mosby; 2008.

101) C
Reference: Norman GR, Streiner DL. Biostatiscis: The Bare Essentials. 3rd ed. St. Louis:Mosby; 2008.

102) D
Reference: Norman GR, Streiner DL. Biostatiscis: The Bare Essentials. 3rd ed. St. Louis:Mosby; 2008.

103) B
Reference: Norman GR, Streiner DL. Biostatiscis: The Bare Essentials. 3rd ed. St. Louis:Mosby; 2008.

104) B
Type 1 or α error. Rejects null hypothesis when the null hypothesis is true. Demonstrates a difference in outcome between two treatments when there is truly no difference = false positive

Type 2 or β error. Do not reject the null hypothesis when the null hypothesis is false. Study demonstrates no difference in outcome between two treatments when there really is a difference = false-negtative.
Reference: Norman GR, Streiner DL. Biostatiscis: The Bare Essentials. 3rd ed. St. Louis:Mosby; 2008.

105) C
The ultrasound suggests bilateral lower urinary tract obstruction. The most common cause of lower urinary tract obstruction is posterior urethral valves. Immediate goals for treatment are to relieve the obstruction through the placement of a drainage catheter.
Reference: Vogt, MacRae Dell and Davis. The Kidney and Urinary Tract. In, Neonatal-Perinatal Medicine: Disease of the fetus and infant. Volume 2. 8th edition. Elsevier. 2008. Page 1678.

106) C
Remember it is the hCG from the placenta that initiates genital differentiation. Around 12 weeks the fetus begins to produce GnRH and the LH and FSH stimulates the fetal gonads to continue with genital development. The hypothalamus needs to provide GnRH so that the pituitary can secrete LH and FSH.
Reference: Anhalt H, Neely K, Hintz R. Ambiguous Genitalia. Pediatr Rev 1996; 17; 213-220.

107) E

An MRI of the brain and cervical spine may show a defect intracranial or at the craniocervical junction causing the tongue fasciculation and hypotonia. Congenital myotonic dystrophy can cause severe neonatal hypotonia however the tongue fasciculations would not be expected. A high arched palate, "myopathic" facies with ptosis and facial weakness would be expected. Grip or percussion myotonia are often absent at this age but will appear later in childhood. EMG/NCS can help determine if it is a lower motor neuron problem at the anterior horn level, axon, myelin, neuromuscular junction, or muscle level. Spinal muscular atrophy is a common cause of severe hypotonia and tongue fasciculations especially in light of a normal mental status. Although Prader Willi syndrome can cause profound hypotonia, tongue fasciculations would not be expected.

Reference: Volpe J. Neurology of the newborn. Saunders Elsevier. 5th Edition.

108) B

The patient in this case has a classic history and symptomatology of a Vein of Galen malformation. Other causes of high output congestive heart failure need to be excluded. Carnitine deficiency may present with a cardiomyopathy but is typically associated with other findings such as muscle weakness and GI issues. Auscultation of the head and neck may detect a loud bruit.

Reference: Cohen AR. Disorders in head shape and size. In, Neonatal-Perinatal Medicine. Martin RJ, Fanaroff AA, Walsh MC, eds. Elsevier. 2011. 9th edition. Ps 1024-1025.

109) D

The crossed adductor sign is present until 7-8 months. The Moro, palmar, and Galant responses should disappear by 6 months of age. The pupillary light response is variably present by 30 weeks gestation and definitely present by 34 weeks gestation.

Reference: Volpe J. Neurology of the newborn. Saunders Elsevier. 5th Edition.

110) D

While autosomal dominant polycystic kidney disease has a higher overall incidence than multicystic dysplastic kidneys (MCDK), it is uncommon in neonatal period. MCDK is the most common cystic renal disease in the newborn period.

Reference: Vogt, MacRae Dell and Davis. The Kidney and Urinary Tract. In, Neonatal-Perinatal Medicine: Disease of the fetus and infant. Volume 2. 8th edition. Elsevier. 2008. Pages 1676-1680.

We thank you for completing the Specialty Review in Neonatology Course Pretest for 2020.

If you have questions about the course, suggestions or comments, please feel free to email Dr. David L. Weisoly, Course Director, at **dweisoly@yahoo.com** or **David_Weisoly@pediatrix.com**.

For questions or input specifically regarding this Pre-test, please contact Dr. Matthew Saxonhouse, Associate Course Director, at **matthew.saxonhouse@atriumhealth.org**