

Case Based Pediatrics For Medical Students and Residents

Questions and Answers

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Answer Set Companion

Answers to Questions

Section I. Office Primary Care

Chapter I.1. Pediatric Primary Care

1. False. Proximity to the patient is also an important factor. A general surgeon practicing in a small town might be the best person to handle a suspected case of appendicitis, for example.
2. False. Although some third party payors have standards written into their contracts with physicians, and the American Academy of Pediatrics has created a standard, not all pediatricians adhere to these standards.
3. True. Many factors are involved, including the training of the primary care pediatrician and past experience with similar cases.
- 4.d
- 5.e

Chapter I.2. Growth Monitoring

1. BMI (kg/m^2) = weight in kilograms divided by the square of the height in meters.
2. First 18 months of life.
3. a) If the child's weight is below the 5th percentile, or b) if weight drops more than two major percentile lines.
4. 85th percentile.
5. 30 grams, or 1 oz per day.
6. At 5 years of age. Those who rebound before 5 years have a higher risk of obesity in childhood and adulthood.
7. It does not provide an accurate index of adiposity since it does not differentiate between lean tissue and bone from fat.
8. Congenital pathologic short stature: infant born small and growth gradually tapers off throughout infancy. Constitutional growth delay: weight and height drop in their percentiles near the end of infancy, parallel the norm through middle childhood, and accelerate toward the end of adolescence. Adult size is normal. Familial short stature: Infant and parents are small. Growth runs parallel to and just below the normal curves.
9. Predicted adult height = (mother's height + father's height) divided by 2, and adding 6.5 cm for males, and subtracting 6.5 cm for females, with a range of 2 standard deviations (one standard deviation is about 5 cm).

Chapter I.3. Developmental Screening of Infants, Toddlers and Preschoolers

- 1.c
- 2.a
- 3.c
- 4.e
- 5.d
- 6.b

Chapter I.4. Immunizations

- 1.e
- 2.c
- 3.a. It should be noted that the current parenteral influenza vaccine is not a live attenuated virus. However, a non-parenteral intranasal live attenuated influenza vaccine is available.
- 4.d
- 5.c
- 6a.passive
- 6b.active
- 6c.passive
- 6d.active
- 6e.active
- 6f.passive

Chapter I.5. Hearing Screening

1. True
2. TORCH: toxoplasmosis, rubella, CMV, herpes
3. False
4. Best test for this age group: Behavioral tests that rely on operant conditioning, such as visual reinforcement audiometry (VRA) involves testing one's response to specific tones projected within a soundproof room from different locations.
5. Screening failure is attributable to middle ear disease. Yet, this does not completely rule out a sensorineural defect.
6. True

Chapter I.6. Anticipatory Guidance

- 1.False
- 2.False
- 3.c
- 4.True
- 5.e
- 6.False
- 7.e

Chapter I.7. Common Behavioral Problems in Toddlers and Young Children

- 1.d
- 2.c
- 3.e
- 4.e
5. Reward good behavior and do it quickly and often. Avoid accidentally rewarding bad behavior. Punish some bad behavior by using mild punishment.
6. Time-out can be used initially with one or two targeted behaviors and once the parent and child get used to the technique, it can be expanded to more problem behaviors. Getting started with time-out should occur after caregivers agree on this as a form of mild punishment. It should then be explained to the child before it is initially used so the child can understand what to expect the first time it is used. The child should immediately be placed in a very boring and safe predetermined location using up to ten words in less than ten seconds from the time the target behavior occurred. The child should be placed in time-out for one minute for every year of life (for example a five year old would sit in time out for five minutes) up to a maximum of about 10 minutes. A small portable timer should always be used to remind the child when the time-out is over. Once the timer rings the child will be asked why they went to time-out. Once they produce the answer, the parent drops the issue and goes about their daily activities as usual. Time-out is not designed to make a child feel bad or humiliated.
7. Pediatricians should be available to offer counseling on routine visits with their patients. When the pediatrician observes bad behaviors in the office they should observe how the parent handles them and offer advice in a nonjudgmental way if they note errors. Pediatricians may also provide tips on effective parenting when the child is very young and be particularly sensitive to the needs of first time parents who may not know the correct way to discipline. One good way to find out how a parent is likely to discipline, is to ask them how they were disciplined as a child and the pediatrician can adjust their advice accordingly. It is very important to remain nonjudgmental and calm as you describe these techniques, as you don't want to add additional stress to a parent who is already taking on a very difficult task of raising a child. Be compassionate, listen and gently advise.
8. A pediatrician would likely want to advise a parent to see a specialist like a child psychiatrist or child psychologist if the problem seems to be more than they can handle. Some of these behaviors include extreme aggression and violence or if the child is engaging in dangerous behaviors. If the child is threatening or trying to hurt or kill themselves or others, this needs to be taken very seriously. The pediatrician will need to clinically assess the situation and decide if an emergency room visit is warranted. Threats of self-harm or harm to others should always be considered as a potential emergency.

Chapter I.8. Disabilities and Physician Interactions with Schools

- 1.b
- 2.a
- 3.d
- 4.d
- 5.False

Chapter I.9. Autism and Language Disorders

1. b, c, e
2. a, b, c, e
3. all are correct
4. False, medications are used symptomatically for particular behaviors or related affective disorder.
5. a, b, d, e

Chapter I.10. Attention Deficit/Hyperactivity Disorder

1. False
- 2.a,c,d
- 3.d
- 4.e
- 5.c
- 6.a

Chapter I.11. Medical Insurance Basics

1. False. The service is not covered in this patient's plan even if it is deemed medically necessary.
2. True. The patient must be informed beforehand that the service may not be covered and that he or she will be expected to pay if they wish to have the service done.
3. False. Contracts between third party payers and providers stipulate that balance billing is not allowed when fees exceed maximum allowable charge on a covered service.
4. True. See Hawaii Revised Statutes Chapter 432e.
5. False. An insurer must observe its operating budget, which is dependent on the premiums received. Insurers cannot generate new money; they can only redistribute what they collect after expending reasonable amounts for operations. Reserves are for unforeseen emergencies. Repeated withdrawals from reserves threaten the solvency of the third party payer.

Chapter I.12. Pediatric Dental Basics

1. True
2. Disorders of tooth eruption and positioning (premature, delayed, or failure of eruption, malocclusion or abnormal alignment), abnormalities of tooth number (supernumerary tooth), size and shape (macrodontia, microdontia, or twinning), structure (AI or DI), and color (intrinsic or extrinsic staining).
3. False. Dentinogenesis imperfecta is the condition that may occur with osteogenesis imperfecta.
4. Streptococcus mutans
5. Fluoride supplementation, good oral hygiene that includes brushing and flossing, limiting the amount but more importantly the frequency of intake of sweets (especially the habit of bedtime bottle feeding, eating in between meals and at bedtime), regular dental visits.
6. It is very likely that this history is not correct. These appear to be baby bottle caries, which is the most likely cause. It may be that mother feels guilty that she is not following your advice so she is denying that the child continues to go to bed with a bottle. Another possibility is that she is giving the child juice in a bottle at night and does not consider this to be "bottle feeding". Grandparents living in the same household will often interfere with childhood rearing practices, since they may insist on letting the child have a bottle to prevent the child from crying.
7. The best thing to do with the tooth is to push it back into its original location after a gentle rinse, if the child is cooperative. Otherwise, the tooth can be placed in saline gauze or milk. The tooth should not be scrubbed.

Section II. Nutrition

Chapter II.1. Nutrition Overview

1. False. Formula still lacks the immunological advantages of breast milk.
2. False. Vegetarian diets are NOT recommended for the first two years of life.
3. True.
4. Yes, at 6 months in children in a community with a non-fluorinated water supply.
5. b. 50% of energy from FAT.
6. No, this child will lose weight (failure to thrive). This child is consuming 40 ounces per day which is only 800 calories per day. This child needs 900 calories (100 cal/kg/day) just for maintenance alone. Growth requires a caloric intake in excess of maintenance.
7. Roughly 64 calories. Protein=4 calories/gram, carbohydrate=4 calories/gram, fat=10 calories/gram. 12 calories from protein, 32 calories from carbohydrates, 20 calories from fat, no calories from sodium total calories=64 calories (roughly).
8. This child is receiving 10% (10 gram/100cc) intralipids at 1cc/hr, or 24 cc/day, which is 2.4 grams per day, which is 24 calories from fat per day. He is getting D12.5% (12.5 gm/100c) at 5.5cc/hr, or 132 cc/day, which is 16.5 grams of dextrose per day, which is 66 calories from carbohydrates per day. He is getting 2 grams of amino acids per 100cc, which means that he gets 2.64 grams of amino acids per day, which is 10.5 calories from protein per day. He is getting a total of 100.5 calories per day, which is 118 calories per kg/day. Since his maintenance caloric requirement is 100 calories/kg/day, he is getting more than maintenance which should give him the potential to grow.

Chapter II.2. Breastfeeding

1. Approximately 60% of women breastfeed immediately post-partum, 20% are still breastfeeding at 6 months, and less than 5% are still breastfeeding at 1 year.
2. The Healthy People initiative set a target to increase the proportion of mothers who exclusively breastfeed to 75% at post-partum, 50% at 6 months, and 25% at 1 year.
3. The AAP recommends exclusive breastfeeding for the first 4-6 months of life, with continued breastfeeding to at least 12 months of age, and thereafter for as long as mutually desired.
4. Advantages of breastfeeding include health, nutritional, immunologic, developmental, psychological, social, economic, and environmental benefits. The major disadvantages to breastfeeding include time and energy required of the mother, decreased paternal (father) participation, and lack of universal social acceptance of breastfeeding practices by the public.
5. Anatomic and physiologic changes that occur in the breast include: a) differentiation of epithelial alveolar cells into secretory cells for milk production. b) proliferation of glandular tissue and ductile development by progesterone. c) copious milk production following placental expulsion due to prolactin unopposed by progesterone. d) milk ejection or milk let-down reflex by oxytocin.
6. Carbohydrate, protein, and fat composition differ. Human milk contains lactose as the main carbohydrate source, high whey to casein protein ratio, and variable fat stores which are dependent on maternal diet. Formulas have variable carbohydrate source which include lactose, starch or other complex carbohydrates. Protein sources can also vary by formula type: casein, whey, soy or protein hydrolysate. Fat sources in infant formula can vary as well: triglycerides with long or medium chains, etc. Breastmilk has more absorbable iron, calcium and zinc than formula.
7. Barriers to successful breastfeeding include: physician misinformation and apathy, insufficient prenatal breastfeeding education, inappropriate interruption of breastfeeding, early hospital discharge, and late hospital follow-up care.
8. Indicators for inadequate breastfeeding include: less than 6 urinations per day and 3-4 stools per day by day 5-7 of life, decreased activity level, difficulty arousing, weight loss of greater than 15% of birth weight within the first week of life.
9. Provide good breastfeeding education at the prenatal visit, be well educated on anatomy and physiology of breastfeeding, advocate for breastfeeding policies.

Chapter II.3. Infant Formulas

1. Breastfeeding is regarded first and foremost except when it is not practical, desired or medically contraindicated.
2. From a practical standpoint, whether it is breast milk or infant formula, a healthy term infant is the best regulator of the frequency and quantity of their nutritional intake. However, since we are scientists at heart; during the first 6 months of life approximately 95-115 kcal/kg/day is recommended.
3. In a term infant, iron deficiency is uncommon before 4-6 months of age because of the abundance of iron stores at birth. To compensate for the depletion of iron stores by growth, dietary iron must be provided to exclusively breastfed infants. Iron fortified formulas can prevent iron deficiency in formula fed infants. Guidelines from the Committee on Nutrition of the AAP recommend 2-3 mg/kg/day of elemental iron.
 - 4a. They are about the same. Human milk contains approximately 2/3 kcal/cc (20 kcal/oz). The standard infant formula usually remains close to this range.
 - 4b. Whey:Casein of human milk is 70:30 as compared to a ratio of 18: 82 for cow milk. Please refer to the text to review the clinical significance of this profile difference.
 - 4c. The carbohydrate content is about the same.
 - 4d. Lipids constitute approximately 50% of the calories in human milk (5.7 g/100kcal) and standard infant formula (4.4-6.0 g/100kcal).
5. The clinical significance of the difference in whey:casein ratio between human and bovine milk is illustrated when unmodified casein-predominant cow milk enters the acidic environment of the human stomach and forms a relatively hard curd of casein and minerals. This curd can be difficult for an infant to digest. Thus, the AAP recommends that cow's milk not be used until after the first birthday.
6. Lactose is the main carbohydrate in mammalian milk. The lactose concentration of human milk is 7g/dL, cow milk contains 5 g/dL. Lactose is added to most standard infant formula to achieve the concentration of human milk. Soy formulas do not contain lactose; they contain sucrose, glucose polymers, or a mixture of the two.

Chapter II.4. Fluids and Electrolytes

1. b
2. c
3. 4kg: $4 \times 100 = 400$ cc over 24 hours. 3 mEq Na per 100 cc, 2 mEq K per 100 cc. D5-1/4NS + 20 mEq KCl per liter run at 17 cc/hour. 25 kg: $1500 + 5 \times 20 = 1600$ cc over 24 hours. Maintenance electrolytes are the same. D5-1/4NS + 20 mEq KCl per liter run at 67 cc/hour.
4. Since normal osmolarity is about 300, the Na concentration in NS must be about half that (since Na and Cl ions make up the total osmolarity), which is 150 mEq/L. 1/2NS is half that (75 mEq/L), 1/3NS is 50 mEq/L and 1/4NS is 38 mEq/L.
5. An intravascular volume expanding fluid is required to resuscitate severe dehydration and hypovolemic shock. D5-1/4NS is not an intravascular volume expanded (see text). NS and LR are intravascular volume expanders. The resident should not have used the term "isotonic" since what he/she really meant, was to administer an intravascular volume expanding IV solution.
6. The patient has normal kidneys, which will regulate his overall fluid status. Even normal infants drink about 250 cc/kg (about 2.5 times maintenance), which is why they use a lot of diapers. Since formula is only 2/3 of a calorie per cc, he needs more than maintenance to reach maintenance caloric intake. His excess fluid volume will be urinated out. Maintenance fluid volume is the volume which results in minimum work for the kidney. If less than maintenance fluid is taken in, the kidney must work (consume energy) to retain fluid. If more than maintenance fluid is taken in, the kidney must work to excrete excess fluid. Kidney energy consumption (work) is minimized at some point between these two extremes and this is the "maintenance volume". Patients receiving fluid volumes less than or greater than maintenance will not likely develop fluid balance problems as long as their kidneys are functioning normally. However, if they are very ill, it would be best to minimize renal stress by optimizing their fluid balance.
7. Oral rehydration with WHO ORS should be implemented immediately. Pedialyte is for maintenance fluid, is suboptimal for rehydration and is only useful for children with mild dehydration. This child is not ill enough to utilize one of the 5 IV sets available. According to studies, the mortality rate for oral rehydration and IV rehydration are the same for this type of dehydration.

8. 24 hour maintenance volume is 1300 cc. This is split up into three even 8 hour blocks. Maintenance electrolytes are 3 mEq Na and 2 mEq K per 100 cc. Deficit volume is 1120 cc (7% of 16 kg), half of which is given in the first 8 hour block with the other half distributed over the next two 8 hour blocks (1/4 for each 8 hour block). Since dehydration has occurred over a 4 day period, 60% of the deficit comes from the ECF (672 cc) and 40% comes from the ICF (448 cc). Thus, the sodium replacement for ECF fluid is 140 mEq per liter and the potassium replacement for ICF is 140 mEq per liter. The Na deficit is replaced as the deficit fluid is replaced over the next three 8 hour blocks (1/2 + 1/4 + 1/4). Half of the K deficit is replaced distributed evenly over the three 8 hours blocks (1/6 + 1/6 + 1/6). The results of these calculations are shown below:

Weight 16 kg 7% dehydration	24 hours	First 8 hours	Second 8 hours	Third 8 hours
Maintenance volume	1300 cc	433 cc	433 cc	433 cc
Maintenance Na	39 mEq	13 mEq	13 mEq	13 mEq
Maintenance K	26 mEq	9 mEq	9 mEq	9 mEq
Deficit volume	1120 cc	560 cc	280 cc	280 cc
Deficit Na (60%)	94 mEq	47 mEq	24 mEq	24 mEq
Deficit K (40%)	63 mEq	10 mEq	10 mEq	10 mEq
Maintenance+Deficit volume	2420 cc	993 cc	713 cc	713 cc
Maint+Def Na	133 mEq	60 mEq	37 mEq	37 mEq
Maint+Def K	89 mEq	19 mEq	19 mEq	19 mEq
IV rate		124 cc/hr	89 cc/hr	89 cc/hr
Na concentration		60 mEq/L	52 mEq/L	52 mEq/L
K concentration		19 mEq/L	27 mEq/L	27 mEq/L

D5-1/3NS+19 mEq KCl per liter run at 124 cc/hour for 8 hours, then D5-1/3NS+27 mEq KCl per liter run at 89 cc/hour for 16 hours. The KCl should actually be approximated to 20 mEq/L for the first 8 hours, then 25 mEq/L for the next 16 hours. This would make it easier for the nursing staff to carry out the order.

Chapter II.5. Failure to Thrive

- 1.False
- 2.True
- 3.False
- 4.True (can help detect renal disorders)
- 5.False
- 6.True
- 7.True

Chapter II.6. Malnutrition and Vitamin Deficiencies

1. A. kwashiorkor. B. marasmus
2. True
3. True
4. b, c, e
5. True
6. a, b, d
7. b, d
8. True
9. a, b, d, e

Section III. Neonatology

Chapter III.1. Routine Newborn Care

1. Vitamin K prophylaxis, antibiotic eye prophylaxis, bathing, and hepatitis B immunization. Breast feeding should also be considered to be an infection prevention/modifying measure.
2. Newborn blood and metabolic disease screening, hearing screening, physical examination.
3. False
4. True
5. False
6. True

Chapter III.2. Neonatal Hyperbilirubinemia

- 1.e
- 2.True
- 3.b
- 4.c
- 5.True
- 6.True
- 7.False
- 8.False
- 9.False
- 10.d

Chapter III.3. Newborn Resuscitation

1. Antepartum risk factors: None. Intrapartum risk factors: emergency cesarean section, non-reassuring fetal heart tones, use of general anesthesia, narcotics administered to mother within 4 hours of delivery, and abruptio placentae.
2. Fluid in alveoli is absorbed and air fills the air sacs, umbilical cord is clamped disconnecting the infant from the placental circulation and pulmonary vasculature must relax allowing increased pulmonary blood flow and decreased right-to-left shunting.
3. Breathing, heart rate and color.
4. Three or more trained persons would ideally be available for an extensive resuscitation requiring medication administration.
5. Ventilation of the lungs is the most important and most effective step in cardiopulmonary resuscitation of the compromised newborn infant.
6. If the infant continues to be apneic, is gasping, has a heart rate of less than 100 bpm and/or has persistent central cyanosis despite 100% free flow oxygen, then positive pressure ventilation with a bag and mask should be administered. Breaths should be delivered at a rate of 40 to 60 per minute.
7. Noticeable chest wall rise, bilateral breath sounds and improved color and heart rate are indications that ventilation is adequate.
8. If the infant's heart rate remains less than 60 bpm following the initial 30 seconds of positive pressure ventilation, chest compressions must be started and assisted ventilation continued. Three compressions should be administered for every one assisted ventilation so that 90 compressions plus 30 breaths are given each minute.
9. Liver laceration and rib fracture.
10. The recommended dose is 0.1 to 0.3 ml/kg of a 1:10,000 solution (equal to 0.01 to 0.03 mg/kg). It can be administered through an endotracheal tube or through an umbilical vein catheter.

Chapter III.4. High Risk Pregnancy

- 1.false
- 2.d
- 3.b
- 4.true
- 5.b
- 6.false

Chapter III.5. Common Problems of the Premature Infant

- 1.true
- 2.a,c
- 3c
- 4.d
- 5.b
- 6.c
- 7.d
- 8.c
- 9.d
- 10.b
- 11.false

Chapter III.6. Respiratory Distress in the Newborn

1. TTN
2. TTN symptoms occur soon after birth. Later onset of symptoms should suggest other disorders.
3. Meconium, blood, amniotic fluid.
4. Air leaks such as a tension pneumothorax.
5. Surfactant deficiency, which causes some alveoli to collapse next to alveoli which are emphysematous. Some atelectatic alveoli are adjacent to rigid bronchi. These conditions lead to a reticulogranular infiltrate (ground glass) and air bronchogram pattern on the chest radiograph.
6. Group B Streptococcus, gram negative rod organisms (usually E. coli) and Listeria monocytogenes.
7. Cyanotic congenital heart disease.

Chapter III.7. Cyanosis in Newborns

1. Hypoplastic right heart syndrome/Pulmonary atresia (these two are part of a spectrum) and transposition of the great vessels.
2. d. All of the choices are correct.
3. False
4. c
5. b
6. Ventricular septal defect (VSD), overriding aorta, pulmonic stenosis, right ventricular hypertrophy. The severity of the pulmonic stenosis is the most important factor in determining the degree of cyanosis.
7. False.
8. True.

Chapter III.8. Neonatal Hypoglycemia

1. False
2. a
3. c
4. $GIR = (\text{dextrose } \% \times \text{ml/kg/d}) / 144$. Start at 6-8 mg/kg/min and titrate.
5. a, b and c are all correct.

Chapter III.9. Neonatal Seizures

1. false
2. c
3. false, since it is poorly absorbed from the infant GI tract.
4. d
5. true
6. a

Chapter III.10. Neonatal Sepsis

1. Blood and urine cultures, if not already done.
2. Clinical sepsis with poor perfusion and neutropenia; possible septic shock with narrow pulse pressure.
3. a) Repeat CBC to monitor the neutropenia and thrombocytopenia. b) Volume bolus to improve perfusion. c) Follow-up exam of abnormal tone and cry after instituting supportive therapy. d) Start broad spectrum antibiotics parenterally. e) Transfer from the normal nursery to a higher level nursery or intensive care unit for continuous monitoring of vital signs.
4. Seven to ten days empirically, given the clinical presentation and depending on culture results. Serial CRPs may also be used to assist with duration of treatment.
5. Any 2 from the battery reviewed by Sinclair (14) gave 62% for sepsis proved or probable.
6. Again any 2 from the above reference (14) gives 98% negative predictive accuracy for sepsis proved or probable. However, the CBC and differential alone will give you two out of this battery.
7. Yes. At least one ml should be obtained for blood cultures.
8. Yes
9. No. This has implications for the current AAP protocol for monitoring infants whose mothers did not receive prophylaxis.
10. 2.5 per 1000 live births, with mortality rate of 8.7% (18). Clinical sepsis is cited as 3.6 per 1000 live births with mortality of 4.3%. Figures are much higher for VLBW infants.

Chapter III.11. Congenital and Perinatal Infections

1. Small for gestational age, microcephaly, jaundice, pale skin, petechiae, blueberry muffin spots, hepatomegaly, and splenomegaly
2. A congenital infection is an infection seen in the newborn infant that was acquired transplacentally during the first, second, or early third trimester. A perinatal infection is acquired either around the time of delivery or during the 1st week of extrauterine life.
3. Rubella virus, cytomegalovirus (CMV) Toxoplasma gondii, Treponema pallidum, human immunodeficiency virus (HIV), human parvovirus B19 and Epstein-Barr virus (EBV)
4. True
5. Periventricular calcifications are seen in congenital CMV while diffuse calcifications in the brain are seen in congenital toxoplasmosis.
6. False
7. Hepatitis B vaccine and hepatitis B immune globulin.
8. False

Chapter III.12. Necrotizing Enterocolitis

1. False, an estimated 25% show visible bloody stool.
2. c. Dopamine may actually reduce the risk of NEC by increasing mesenteric blood flow.
3. False, the development of resistant organisms presently discourages routine prophylactic antibiotic use.
4. Reduced intestinal motility increases the chances of bacterial overgrowth.
5. Acceptable answers include: 1) oral feeding cessation, 2) nasogastric decompression, 3) intravenous fluid therapy, 4) systemic antibiotics, 5) umbilical catheter removal, 6) acid-base electrolyte balance monitoring, 7) early consultation with a surgeon.

Section IV. Genetics

Chapter IV.1. Prenatal Genetic Screening and Testing

- 1.d
- 2.false
- 3.c
- 4.c
- 5.d
- 6.c
- 7.b
- 8.a
- 9.true
10. b

Chapter IV.2. Congenital Anomalies and Teratogenesis

- 1.d
- 2.c
- 3.b
- 4.a
- 5.d
- 6.d
- 7.c

Chapter IV.3. Common Chromosomal Disorders

1. Trisomy 13, Trisomy 18
2. Trisomy 18. Heart failure and pneumonia
3. Fragile X syndrome; Huntington disease; Friedreich ataxia; and myotonic dystrophy
4. VALIDATE: VSD, Atlanto-occipital instability, Leukemia, Immunodeficiency, Duodenal atresia, Alzheimer's disease, Thyroid dysfunction, Endocardial cushion defect.
5. Ovarian dysgenesis
6. Elevated estradiol to testosterone ratio
7. b,c,d

Chapter IV.4. Inborn Errors of Metabolism

1. False: Many infants with metabolic defects classified as storage disorders (lipid storage disorders) and fatty acid oxidation defects will present at many months of age.
2. e. And, there are many other disorders that can be on the list of possibilities, including child abuse (shaken baby).
3. c, d, f. The other answers are incorrect because: a. Newborn screening is not a diagnostic tool; it merely indicates need for further definitive testing. b. Obviously, physicians do not need more paperwork. e. Ideally, newborn screening could identify all metabolic disease, however, since cost and technology are prohibitive, the current principles are to screen for diseases which have a "significant" prevalence in a population and have some potential for treatment.
4. True: Unfortunately, there are no permanent cures, only lifelong supportive measures to mitigate the effects of the metabolic disease.
5. c

Chapter IV.5. Inherited Connective Tissue Disorders

1. Presence of associated physical findings. Family history. Location of fracture (femur and radius vs tibia and radius), type of fracture (comminuted mid shaft vs epiphyseal and greenstick). Radiographic appearance of the fractures (i.e., presence of osteopenia).
2. Careful fracture history, identifying weak bones, and targeting physical therapy to strengthen those bones.
3. Any of the following: pectus carinatum (or excavatum sufficiently severe to require surgery), reduced upper to lower segment ratio, positive wrist and thumb signs, scoliosis greater than 20 degrees of curvature, reduced extension of elbows, medial displacement of medial malleolus causing pes planus, protrusio acetabuli.
4. Aortic root dilation causing aneurysm and dissection.
5. Any three of the following: hyperextensible doughy skin, atrophic scars, joint hypermobility, connective tissue fragility, and bruising.
6. Marfan syndrome, unlike homocystinuria, is not associated with mental retardation.

Chapter IV.6. Genetic Testing and Gene Therapy

1. False. Newborn screening is not diagnostic. Rather, it is a screen for illness with VERY poor specificity, which, if positive, must be followed with a more specific diagnostic test.
2. Sequence knowledge of the disease locus and mutant alleles and the 1:1 correlation of test to disease allele. For disease conditions with multiple mutant alleles, all possibilities must be specifically tested.
3. The disease does not affect the patient until adulthood when she can make her own decisions. There is no effective prophylactic treatment for a child that will prevent the illness before she reaches adulthood. Testing may be appropriate for a 17 year old who desires pregnancy, has the consent of her parents, and who plans to make the decision to become pregnant based on the information of the test.
4. Bone marrow transplant.
5. Vectors transport engineered nucleic acids (DNA or RNA) into existing human cells.
6. 1) DNA based: Insertion of intact functional gene. Insertion of intact functional promoter or exons to correct production. Insertion of DNA for the purposes of disrupting expression of a gene. Insertion of single stranded DNA for the purposes of binding to mRNA and preventing translation. 2) RNA based: Insertion of RNA to be reverse transcribed and incorporated into DNA. Insertion of RNA to be translated immediately. Insertion of RNA ribozyme to destroy mRNA. Insertion of anti-sense RNA to prevent translation of mRNA.

Chapter IV.7. Basic Genetic Principles

1. b. An autosomal dominant condition which is lethal in infancy is not going to survive in the gene pool. Such conditions must be autosomal recessive to survive in the gene pool. Most autosomal recessive conditions are enzyme deficiencies. An X-linked enzyme deficiency is also a possible answer, but this is less likely and it is not one of the choices given.
2. b,d. Enzyme deficiencies must be homozygous for the condition to manifest, because a 50% reduction of the enzyme level is generally sufficient to carry out the biochemical reaction involved, such that no clinical disease results. The observed inheritance pattern is autosomal recessive. Enzymes on the X-chromosome such as RBC G6PD are not present on the Y-chromosome, so enzymes can also be inherited in an X-linked recessive fashion. An enzyme deficiency is not likely to manifest from a spontaneous new mutation, because it would have to coincidentally occur in both alleles for this to occur.
3. e. Trisomy 21 results from meiotic nondisjunction in about 95% of patients. About 4% have a Robertsonian translocation. A small percentage of patients are mosaic. An even rarer cause of trisomy 21 is the 21q21q translocation, a chromosome comprised of two chromosome 21 long arms. It is thought to originate as an isochromosome.
4. c. There is a far greater probability of males expressing recessive alleles in their phenotypes if they are carried on X chromosomes. For females to have such traits, they would have to inherit the recessive allele for them on both of their X chromosomes.
5. e. An exchange of fragments of chromatids between non-homologous chromosomes during the first meiotic division is termed a translocation.

Section V. Allergy and Immunology

Chapter V.1. Common Allergies and Management

- 1.d
- 2.a
- 3.a
- 4.e
- 5.b
- 6.b
- 7.a
- 8.c
- 9.d
- 10.e

Chapter V.2. Anaphylaxis and Other Acute Allergic Reactions

1. false
2. Epinephrine. Pediatric dosage for epinephrine is 0.01mg/kg up to a max dose of 0.5mg per dose or 0.5ml of 1:1000 SQ/IM Q15minutes for two doses and then Q4 hours as needed. The adult dosage is 0.2-0.5ml of a 1:1000 epinephrine solution.
3. Adjunctive therapies includes antihistamines, bronchodilators, and perhaps glucagon and corticosteroids.
4. e. This is erythema multiforme.
5. b

Chapter V.3. Food Allergies

- 1.b. Tingling in the mouth after eating fruits suggests the possibility of an oral allergy syndrome. Dizziness after eating Chinese food is more likely due to an adverse non-allergic reaction to MSG. Facial redness after drinking a glass a wine may be due to tyramine.
- 2.a
- 3.a
- 4.e
- 5.e
- 6.f. Chinese and southeast Asian foods are frequently cooked with peanut oil. None of the above are safe. Ice cream is potentially contaminated by nuts since nuts are frequently served with ice cream or mixed with ice cream. Dry pet food and chili frequently contain peanuts. Pastry may contain peanuts even if they are called other types of nuts such as almonds.
- 7.a. Lactose is merely a disaccharide. Lactose by itself is not part of milk protein. However, if the source of lactose is a dairy product, then this should be avoided. All of the other products including "non-dairy" creamers and canned tuna may contain milk or milk products.

Chapter V.4. Corticosteroids

- 1.d. Norepinephrine is a hormone of the adrenal medulla, not the adrenal cortex. Corticosteroids are by definition hormones of the adrenal cortex.
- 2.d. Dexamethasone is a high-potency, long-acting glucocorticoid. Prednisone, prednisolone, and triamcinolone are intermediate-potency glucocorticoids.
- 3.a. Eosinophils, lymphocytes, and monocytes are reduced in the peripheral circulation after corticosteroid administration. Although neutrophil numbers are increased, their bactericidal activity is decreased.
- 4.d. Safely tapering corticosteroids in a patient who has taken corticosteroids for more than 10 days, involves reducing the previous week's levels by 25%, and the patient should be monitored clinically for signs of corticosteroid withdrawal (malaise, anorexia, headache, lethargy, nausea, fever, loss of cardiovascular tone, with hypotension, shock, and death) and a worsening of the condition that the corticosteroids were originally given for.

5.a. Th1 cells are stimulated by IL-12 from APC to cause a cellular immune response. Th2 cells, upon stimulation by IL4, cause a humoral response. IL-12 will inhibit IL-4 production as well. Glucocorticoids cause a decrease in IL-12 secretion by APC and IL-12 responsiveness in Th1 cells. This inhibition of IL-12 frees IL-4 to have a more unopposed effect, triggering an enhanced humoral response.

6.c. Glucocorticoids inhibit production of arachidonic acid, prostaglandins, thromboxanes, leukotrienes, and nitric oxide, all of which are involved in the inflammatory response. Neutrophils are increased in the peripheral blood, not decreased.

7. a,b,c are correct. 0.2 mg/kg of dexamethasone would probably be the best answer, although its duration is longer than that of methylprednisolone. This should not be a problem for status asthmaticus. 10 mg/kg of hydrocortisone has equivalent glucocorticoid activity, but it has unnecessary mineralocorticoid activity. 2 mg/kg of prednisone is roughly the same as 2 mg/kg of methylprednisolone, but prednisone would have to be given orally since it cannot be given IV. 20 mg/kg of dexamethasone is clearly an overdose, which results from multiplying by 10 instead of dividing by 10. A good clue would be that dexamethasone comes in 10 mg vials. A 400 mg dose would require 40 vials. This should clearly prompt questioning by pharmacy and nursing staff. Whenever a pediatric dose requires more than one vial, the dose should be questioned.

8. The symptoms of croup and status asthmaticus are largely due to the inflammatory response induced by the viral infection. The virus itself causes less of a problem compared to the body's inflammatory response. Corticosteroids suppress the inflammatory response resulting in less laryngeal and bronchial inflammation. It cannot be assumed that this is true for all viral infections. For example, in viral pharyngitis, the symptoms of a sore throat and nasal congestion may be suppressed with corticosteroids. However, it may cause more harm than good. In the case of croup and status asthmaticus, numerous studies have supported the net benefit of corticosteroids in these two conditions. In bacterial meningitis due to H. flu, a similar benefit has been demonstrated, but for bacterial meningitis due to other organisms and for viral meningitis, the benefit has not been clearly demonstrated.

Chapter V.5. Immune Deficiency

- 1.e
- 2.c
- 3.c
- 4.e
- 5.c
- 6.b
- 7.c
- 8.e
- 9.c
- 10.b

Chapter V.6. Hematopoietic Stem Cell Transplantation and Graft Versus Host Disease

1. e
2. False. HLA matching is the best predictor.
3. d
4. True
5. b

Section VI. Infectious Disease

Chapter VI.1. Virology

1. Poor pappy adds hep to her pox: Parvovirus, papovavirus, adenovirus, hepadnavirus, herpesvirus, poxvirus. The first three are naked, the latter three are enveloped.
2. Naked viruses cause acute infection only. Some enveloped viruses are capable of chronic infection.
3. PEECoRnA: polio, entero, echo, coxsackie, rhino, hepA.
4. VZV and HSV are similar in that they both cause acute vesicular infections with lifelong latency and recurrence. EBV and CMV are similar in that they both cause infectious mononucleosis type syndromes. CMV and HSV both cause congenital viral infection malformation syndromes.
5. Rhinovirus, RSV, parainfluenza virus, coronavirus, adenovirus. Influenza virus may be included also.
6. Pete can float toward the coast backward: picorna, calci, flavi, toga, corona, retro.
7. None. Only enveloped viruses can cause chronic infection.
8. Raspberry filled parfaits are often burned: rhabdo, filo, paramyxo, arena, orthomyxo, bunya.
9. Viruses are too small to be seen on light microscopy. On electron microscopy, nearly all naked viruses have an icosahedral shape.
10. Herpesvirus (HSV, VZV, CMV), picornavirus (poliovirus, enteroviruses), flavivirus (encephalitis), togavirus (encephalitis), rhabdovirus (rabies), bunyavirus (encephalitis).

Chapter VI.2. Basic Bacteriology

1. This is Staph epi which is almost always resistant to methicillin and cephalosporins.
2. The peritoneal fluid is likely to grow multiple stool organisms. E. coli will predominate. A polymicrobial anaerobic infection is also likely. To properly culture anaerobes, an anaerobic culture swab sent in special anaerobic media (e.g., thioglycolate) must be sent.
3. They are usually polymicrobial and they have a foul odor.
4. Tetanus, botulism, diphtheria, toxic shock, staphylococcal scalded skin syndrome, scarlet fever, etc.
5. Early antibiotic treatment results in a slightly shorter course of symptoms, but the main reason to treat is to prevent suppurative complications and rheumatic fever.
6. Lancefield classification to determine if this organism is group A, group B, etc.
7. Pneumococcus or Staph epi (contaminant).
8. Most likely gram positive cocci is pneumococcal meningitis. Most likely gram negative cocci is meningococcal meningitis.
9. This is an inappropriate order. The stool will be full of enterobacteriaceae, anaerobes, and enterococcus. The gram stain will show mostly gram negative rods and perhaps a few gram positive cocci.
10. It is not possible to determine this with certainty in most instances. However, healthy patients who are no longer ill by the time the culture comes back are unlikely to have had Staph epi bacteremia. Thus, in these patients, the Staph epi is most likely a contaminant. In patients with indwelling plastic (central catheters, ventriculoperitoneal shunts), it should be assumed that the Staph epi is a clinically important infection, probably colonizing the plastic tubing.

Chapter VI.3. Fever

1. True
2. c
3. False
4. False
5. 6 months for boys, 24 months for girls.
6. False. At the most, teething might causes a very slight temperature elevation.
7. False. Otitis media is not considered to be a reliable source of causing a high fever. Other conditions, such as UTI, need to be considered.
8. False.

Chapter VI.4. Inhibitory and Bactericidal Principles (MIC & MBC)

1. When the level of the antibiotic is so high that all organisms are killed.
2. When the level of the antibiotic is so low that organism growth is inhibited, but they are not killed.
3. No. MIC/MBC or Schlichter tests are only useful when a very long course of antibiotics are anticipated and the patient must be changed to oral antibiotics to complete the antibiotic course as an outpatient. These tests are necessary to determine if it is possible to attain sufficient blood levels with the oral antibiotics to predict therapeutic success. The most common clinical scenarios would be for osteomyelitis, septic arthritis and bacterial endocarditis.
4. A Schlichter test should be performed when the lab is unable to measure levels of the antibiotic that is to be used.
5. When we don't have an organism (cultures are negative).
6. We are never totally sure. We do know that compared to blood levels, most antibiotics have lower levels in bone and in joint fluid, but higher levels in urine.

Chapter VI.5. Antibiotics

1 and 2. There are at least four, and probably five, and possibly six. No doubt in the future, there will be more. How do these cephalosporins differ from each other and what characteristic places them in a given generation? The answer to this question is not an easy one. If you enter "fourth generation cephalosporin" into Medline's search engine, you will find some articles on fourth generation cephalosporins. Similarly, searches for fifth and sixth generation cephalosporin yields some articles. If I was a slick marketer of drugs, I would simply call my new cephalosporin "Tenth Generation" and almost everyone would buy it. However, what specific characteristic of the cephalosporin makes it clinically useful over other cephalosporins? If the drug was a tenth generation cephalosporin, but it had no clinical advantage over an existing third generation cephalosporin, then there is no need for a such a tenth generation cephalosporin. The generation is not nearly as important as the specific property of the cephalosporin which makes it clinically useful over another cephalosporin.

3. Osteomyelitis: Most likely Staph aureus. Bacterial meningitis: Pneumococcus, meningococcus, Haemophilus influenzae type B (HiB).

4. For osteomyelitis, we could cover the Staph aureus with an anti-Staph aureus penicillin such as oxacillin, nafcillin or methicillin or a first generation cephalosporin such as cefazolin. However, resistance to these drugs is currently about 25% to 30%. Although there is a good chance the patient will respond, in 25% to 30% of cases, this treatment will fail and the patient will suffer the consequences of inadequate treatment which would include: death from sepsis, Staph pneumonia, spread of the osteomyelitis, chronic osteomyelitis requiring an amputation, etc. None of these complications are minor, therefore, 75% coverage is inadequate. We need 100% coverage empirically since osteomyelitis is a serious infection. Thus, IV vancomycin is the treatment of choice here. For the bacterial meningitis case, we need an antibiotic to effectively cover these organisms and additionally, we need an antibiotic that will penetrate the blood brain barrier into the CSF. Chloramphenicol would be satisfactory here, but we don't use this because of its side effects. IV ceftriaxone or cefotaxime would penetrate the CSF well and cover meningococcus and HiB, and most pneumococcus, but pneumococcus has a small frequency of high level resistance to cephalosporins, so vancomycin must be added.

5. What organism is most likely? Mycoplasma or viral. Pneumococcus is unlikely since she is afebrile. The best antibiotic choice would be an erythromycin.

6. Although trimethoprim/sulfamethoxazole (Bactrim or Septra) is commonly recommended because of its broad coverage for this indication, this drug causes Stevens-Johnson syndrome more commonly than others. If the parents accept this increased risk, then this should be documented on the chart. Most parents are not willing to accept this increased risk since other antibiotics are available. Amoxicillin will probably work, but there is a high frequency of resistance which is generally not a problem for simple cystitis, but in a febrile 18 month old, there may be some degree of pyelonephritis as well. Resistance to cephalosporins is infrequent. Thus, an acceptable answer here would also be a first generation cephalosporin such as cephalexin. IM ceftriaxone can also be given at the initial patient encounter to ensure high initial antibiotic levels and initial compliance.

7. Cost, compliance, convenience, efficacy, etc. While EES is \$10 and azithromycin is \$70, some patients may choose to pay more if the more expensive drug has significant advantages. Additionally, since most patients have drug plans, the difference may be negligible (e.g., \$5 vs. \$10). Compliance is essential for the drug to be effective. EES must be taken four times a day for 10 days while azithromycin is once a day for five days. Additionally, EES may have more GI side effects. Clearly a once a day medication is more convenient than a q.i.d. medication. If both medications are efficacious, perhaps it is best to discuss these differences with the patient and give them some input in the decision.

Chapter VI.6. Otitis Media and Otitis Externa

1. 6 to 18 months of age.
2. Attendance in day-care, second-hand cigarette smoke exposure, craniofacial abnormalities, bottle-feeding in the horizontal position.
3. Pneumatic otoscopy (myringotomy/tympanocentesis is the gold standard, but not the best diagnostic tool because of its invasiveness).
4. AOM: otalgia, fever, hearing loss, associated with upper respiratory tract infection; TM that is opaque or erythematous and bulging with poor mobility, perforation. OME: commonly asymptomatic but may have hearing loss; retracted TM.
5. Streptococcus pneumoniae, non-typable Haemophilus influenzae, Moraxella catarrhalis.
6. Amoxicillin
7. Amoxicillin-clavulanic acid, cefuroxime axetil, intramuscular ceftriaxone
8. Significant conductive hearing loss; young infant since they cannot communicate their symptoms; associated suppurative upper respiratory tract infection; concurrent permanent conductive and sensorineural hearing loss; speech-language delay because of effusion and hearing loss; alterations in the tympanic membrane such as a retraction pocket; middle ear changes such as adhesive otitis media or involvement with the ossicles; previous surgery for otitis media; frequent recurrent episodes; and persistence of the effusion for 3 months or longer in both ears or 6 months or longer in one ear.
9. Conductive and sensorineural hearing loss, mastoiditis, cholesteatoma, labyrinthitis, facial paralysis, meningitis, brain abscess, and lateral sinus thrombosis.
10. Pseudomonas aeruginosa.
11. Excessive wetness, lack of cerumen, preexisting skin problems, and trauma.
12. 2% acetic acid or dilute alcohol.

Chapter VI.7. Sinusitis

1. Amoxicillin 45-50 mg/kg/day. A higher dose should be prescribed if pneumococcal resistance is likely.
2. Up to 10% will progress.
3. Allergic rhinitis, viral infections, cystic fibrosis, foreign body.
4. Mucosal thickening of at least 4mm, air fluid levels, opacification.
5. Periorbital cellulitis.

Chapter VI.8. Mastoiditis

1. S. pneumoniae, H. influenzae (non-typable), and M. catarrhalis.
2. S. pneumoniae, S. pyogenes, and S. aureus.
3. Meningitis, epidural empyema, subdural empyema, venous sinus thrombosis.
4. Facial nerve paralysis, deafness, labyrinthitis, petrositis, Bezold abscess.
5. In the older child the ear is up and out and in the infant it is down and out.
6. False
7. True

Chapter VI.9. Oral and Upper Respiratory Infections

1. any of the answers may be correct depending on your practice setting.
 - a. for difficult to reach families or someone you don't trust to follow up.
 - b. is probably what you would do for most families you felt comfortable with follow up (i.e., you could reach them on the phone if you needed to).
 - c. is what you might do if you are playing the odds; it's probably viral.
 - d. is what you might do during an epidemic.
2. a, c and d (b - HIV antibody test - is usually negative during this period and PCR for p24 antigen, RNA or reverse transcriptase is required).
3. d
4. b (antitoxin must be given with antibiotics)
5. a, b and d (no one is sure what causes PFAPA)

Chapter VI.10. Pertussis

- 1.a. A false negative can occur in those who have received amoxicillin.
2. None of the choices are correct. Choices a and e are the closest to being correct, but technically, these answers are incorrect. Mycoplasma pneumonia might show up as a pneumonia on a CXR, but this would be non-specific for mycoplasma. Additionally, some mycoplasma infections may not cause a pneumonia. Foreign body aspiration might show up on a CXR, but these often require special views such as an expiratory view or a lateral decubitus view. Foreign body aspiration is frequently occult.
3. a-2, b-1, c-3, d-4
- 4.d. Suctioning of nose, oropharynx, or trachea always precipitates coughing, occasionally causes bronchospasm or apnea, and should be done prn only.
5. Increased intrathoracic and intra-abdominal pressure during coughing can result in conjunctival hemorrhages, petechiae on the upper body, epistaxis, hemorrhage in the central nervous system and retina, pneumothorax and subcutaneous emphysema, and umbilical and inguinal hernias. A child protective services report is not necessarily indicated since pertussis could cause this. Other clinical or psychosocial findings inconsistent with pertussis may lead one to report this to child protective services.

Chapter VI.11. Pulmonary Infections

- 1.c. Overall, viruses cause the majority of pneumonias in children; however, the incidence of viral pneumonia decreases with age, becoming less common in older children and adolescents.
- 2.b
- 3.False
- 4.False. Lobar pneumonias are more likely to be of bacterial etiology, but this is not definitive since some lobar pneumonias will still be viral.
- 5.d
- 6.a
- 7.True
- 8.d
- 9.False

Chapter VI.12. Croup and Epiglottitis

1. d.
2. False. Routine airway visualization is stressful and may precipitate respiratory arrest. If epiglottitis is unlikely, then airway visualization appears to be safe. In the event of respiratory arrest, laryngoscopy will be necessary for tracheal intubation.
3. d is the best answer. c is also correct in that nebulized albuterol does have some efficacy in croup, but nebulized epinephrine is better.
4. d.
5. Most textbooks would suggest that this is false in that a longer observation period is generally recommended. However, most patients are low risk and can be discharged soon after dexamethasone and epinephrine are administered. Severe patients or those who do not respond as well should be observed for longer periods of time.

Chapter VI.13. Cellulitis

- 1.b
- 2.a
- 3.a
- 4.d
- 5.a

Chapter VI.14. Meningitis

1. This is most likely a viral meningitis. He is older, so his risk of bacterial meningitis is lower. He has been fully immunized, which presumably means that he has had H. influenzae, type B vaccine. He has probably had pneumococcal vaccine, but this can't be automatically assumed. He is alert, ambulatory, and not toxic in appearance, which all suggest that he does not have an overwhelming infection such as bacterial meningitis.

2. This is most consistent with viral meningitis. Although he has a high percentage of segs, this is still consistent with early viral meningitis. Cases of bacterial meningitis which have not been pre-treated with antibiotics almost always have more than 90% segs. The gram stain does not show any organisms which makes bacterial meningitis less likely. This laboratory analysis of his CSF suggesting viral meningitis, is consistent with his clinical appearance which also suggests viral meningitis (see the answer to #1 above).

3. Pneumococcus, meningococcus and Haemophilus influenzae type B. Pneumococcus is usually sensitive to penicillins and cephalosporins, but some resistance has emerged so vancomycin should be given in addition to cefotaxime or ceftriaxone. Meningococcus is sensitive to penicillin so cefotaxime or ceftriaxone provides sufficient coverage. H. influenzae type B is sensitive to cefotaxime and ceftriaxone, but this organism is not a common cause of bacterial meningitis due to widespread immunization against this organism.

4. CSF 1 shows bacterial meningitis. The increased number of cells in the CSF with a predominant number of neutrophils makes this a strong likelihood possibility. In addition, he also has a very low glucose CSF level (CSF, blood glucose ratio of 25%) and an increased protein value sometimes. Cases of early viral meningitis can present with an increased number of cells and neutrophils but usually the CSF glucose is normal or not lower than 40% of the blood CSF value.

CSF 2 is normal. The normal number of WBCs in the CSF depends upon the age of the patient. The younger and more immature the infant is, the higher the value is. CSF glucose value depends upon the value of glucose in the blood and upon the integrity of the blood brain barrier. In patients with normal meninges the CSF value is usually about 75% of the blood level. When the meninges become inflamed, the active transport of glucose across the blood brain barrier becomes altered and the ratio drops proportionately to the degree of inflammation. Most viral meningitis produce less changes than bacterial meningitis accordingly CSF glucose values are lower in bacterial meningitis.

CSF 3 shows viral meningitis. Most cases of viral meningitis will present with a moderate increase in the number of white cells and a percentage of neutrophils not higher than 60-70%.

CSF 4 is inconclusive. The high percentage of neutrophils indicates that bacterial meningitis is possible. It would be wise to administer antibiotics until more information can be obtained. The gram stain result will be helpful. If it is positive for organisms, then this indicates bacterial meningitis. If the gram stain is negative, bacterial meningitis still cannot be totally ruled out. The child's clinical condition is not part of this table, but in reality, a child who is alert, active and playful is more likely to have viral meningitis, as opposed to a lethargic, toxic child who is more likely to have bacterial meningitis. This will probably turn out to be a case of viral meningitis despite the high percentage of neutrophils, since an early viral meningitis will often have high neutrophil percentages. A repeat LP 12 to 24 hours from the first LP will be helpful. A repeat LP which demonstrates a clear shift toward mononuclear cells, is consistent with viral meningitis, while no shift, or only a slight shift would suggest bacterial meningitis. Culture of the CSF will be most definitive if it is positive, but this result will not be available for at least 24 hours.

Chapter VI.15. Encephalitis

1. a. viral
2. HSV, St. Louis encephalitis, and rabies virus.
3. a. HSV
4. Japanese encephalitis-decorticate or decerebrate posturing, Eastern equine encephalitis-highest mortality, Post-infectious encephalitis-involvement of multiple CNS levels, St. Louis encephalitis-SIADH, La Cross encephalitis-Aedes triseriatus.
5. e. Rabies virus
6. False. Antiviral therapy has only decreased mortality, NOT morbidity.

Chapter VI.16. Sepsis

- 1.a
- 2.b
- 3.b
- 4.d
- 5.a

Chapter VI.17. Kawasaki Disease

1. Presence of fever ranging between 38 and 41 degrees C, and four out of five principal diagnostic criteria which include: discrete conjunctival injection without exudates, changes in the mouth, polymorphous erythematous rash, changes in the hands and feet, and unilateral cervical lymphadenopathy.
2. Intravenous gamma globulin treatment.
3. Children <1 year of age and those untreated with IVIG.
4. Stevens-Johnson syndrome, erythema multiforme, serum sickness.
5. Measles, adenovirus, toxic shock syndrome, scarlet fever, staphylococcal scalded skin syndrome.

Chapter VI.18. Staphylococcal and Streptococcal Toxic Shock Syndromes

1. True
2. True
3. True
4. True
5. False. The mortality rate for Strep TSS is 30-70%. The mortality rate for Staph TSS is much lower.
6. True
7. True
8. True
9. True
10. True. Examples include impetigo and paronychia.

Chapter VI.19. Tuberculosis

- 1.False
- 2.True
- 3.False
- 4.False
- 5.True
- 6.True
- 7.True
- 8.True
- 9.False
- 10.False

Chapter VI.20. Human Immunodeficiency Virus (HIV) Infections

- 1.e
- 2.a
- 3.e
- 4.b
- 5.c
- 6.e

Chapter VI.21. Sexually Transmitted Infections

1. Abdominal pain, adnexal tenderness on bimanual exam, and cervical motion tenderness.
2. Human papillomavirus (HPV) is estimated to be the most common STI among young, sexually active people in the United States, though many HPV infections are asymptomatic. According to the CDC, an estimated 5.5 million people of all ages contract HPV each year in the United States. On the other hand, chlamydia is the most commonly reported infectious disease in the United States.
3. Adolescents adopt high risk behaviors including early onset of sexual activity, multiple sexual partners, or drug/alcohol use which may impair judgment. Adolescents generally are less able to access health care due to embarrassment about their condition, financial constraints, or transportation barriers.
4. d. Quinolones are no longer recommended for the treatment of gonorrhea in Hawaii or infections acquired in Asia. In 2000, the CDC collected 5,461 isolates for its Gonococcal Isolate Surveillance Project (GISP). 14.3% of the GISP isolates in Hawaii were found to be quinolone-resistant *N. Gonorrhoeae* (QRNG), compared to 0.2% of samples collected within the continental United States and Alaska. Furthermore, since QRNG is becoming more common in West Coast areas, the use of fluoroquinolones in California is probably inadvisable.
5. False. Acyclovir and other antivirals only reduce viral shedding, but they do not eliminate the risk of transmission. Suppressive therapy reduces the frequency of symptomatic genital herpes recurrences by 70% to 80% for patients with 6 or more recurrences a year.
6. FTA-Abs is more specific, but it is still not totally diagnostic of syphilis since patients with yaws will still have a positive FTA-Abs. Other false positive results of FTA-Abs may occur with patients with various medical problems. The differential diagnosis of a positive treponemal antibody test includes other treponemal diseases such as pinta, yaws, and endemic syphilis.
7. Criteria for hospitalization include: surgical emergencies (e.g., appendicitis) that cannot be excluded, pregnancy, failure to respond to outpatient treatment, suspected noncompliance or intolerance to outpatient treatment, nulligravid status, severe illness (including nausea, vomiting, or high fever), suspected tubo-ovarian or other pelvic abscess.

Chapter VI.22. Common Viral Exanthems

- 1.a
- 2.e
- 3.b
- 4.c
- 5.b

Chapter VI.23. Epstein-Barr Virus Infections

1. The answer is b. In this case, the group A streptococcus probably represents colonization rather than the etiology of the patient's symptoms. Infectious mononucleosis may have a similar presentation to streptococcal pharyngitis, and must be considered if a patient is not responding clinically to treatment with antibiotics. Diagnosis may be made with a Monospot test as well as the presence of atypical lymphocytes on CBC. EBV titers are not usually needed in diagnosis, but may be considered if the Monospot is negative and EBV infection is to be ruled out. Treatment with acyclovir or corticosteroids has not been proven to be of clinical benefit in uncomplicated cases of infectious mononucleosis.
2. The answer is b. Primary EBV infection occurs more commonly in childhood and is often asymptomatic. In children who do develop symptomatic EBV infection, heterophil antibodies are more often negative. Lymphocytic interstitial pneumonitis may occur in children with HIV. Complications occur less commonly in children than in adults.
3. The answer is d. The first three have all been found to be associated with EBV infection. Kaposi's sarcoma is associated with a different human herpes virus, referred to as human herpes virus-8 or HHV-8.
4. The answer is b. The Monospot test is a highly sensitive test, although ten percent of EBV-associated infectious mononucleosis may be negative. There are also a number of organisms that may cause an infectious mononucleosis-like syndrome but are not associated with formation of heterophil antibodies. The most common cause of a heterophil-negative infectious mononucleosis-like syndrome is CMV, which this patient likely has. Obtaining antibody titers specific against EBV and CMV may clarify the diagnosis. The atypical lymphocytes that may be seen with either EBV or CMV infection represent activated T lymphocytes, which proliferate in response to infected B lymphocytes.

5. The answer is a. The syndrome of infectious mononucleosis results from primary infection with EBV, particularly when it is delayed until adolescence or young adulthood. It is usually transmitted through close contact with oral secretions of an infected individual. The virus is ubiquitous, and almost all adults over age 40 show serologic evidence of prior infection. Splenic rupture is a rare complication of EBV-associated infectious mononucleosis.

Chapter VI.24. Polio

1. Picornaviridae family (Pico=small, RNAviridae=RNA virus).
2. Asymptomatic presentation is up to 95% of the cases.
3. The correct answer is b, exclusive IPV immunization.
- 4a. OPV (for endemic countries).
- 4b. IPV (Household contact, especially since Grandpa might be changing the diapers).
- 4c. OPV (May receive 3rd and/or 4th oral doses).
- 4d. IPV (Immunization through all IPV schedule).
- 4e. OPV (Mass vaccination campaign to control outbreaks).
5. The proposed mechanism includes the dropout of neurons that were reinnervated after the initial paralytic poliomyelitis infection due to increased metabolic stresses.
6. True. The March of Dimes was originally named the National Foundation for Infantile Paralysis.

Chapter VI.25. Rabies

- 1.b,e
- 2.e
- 3.False
- 4.b,d
- 5.a

Chapter VI.26. Rocky Mountain Spotted Fever

1. False. Primarily south-Atlantic and south-central states.
2. False. A history of tick bite or exposure is obtained in only 60% of cases.
3. True. Therapy should never be withheld until a definitive diagnosis is made. Poorer outcome with increased mortality is associated with delay in initiating treatment.
4. False. Rash typically starts on the hands/wrists and feet/ankles. Involvement of the palms and soles is classic.
- 5.c
- 6.b

Chapter VI.27. Lyme Disease

1. True.
2. False. Bell's palsy due to Lyme disease should NOT be treated with corticosteroids.
3. False.
4. True.
5. True.
6. True.
7. False.
8. False.
9. True.
10. False. Positive Lyme serology in low risk cases are usually false positives.
11. True

Chapter VI.28. Leptospirosis

- 1.b, 2.d, 3.c
- 4.e. None of the above. Leptospira are difficult to culture. Culture requires special laboratory techniques not available at most clinical labs. Thus, the diagnosis is usually confirmed by serology.
- 5.a
- 6.e. Jaundice indicates icteric leptospirosis, which is a more serious condition which has a higher mortality rate. Azotemia is an additional marker of severity.

Chapter VI.29. Cat Scratch Disease

1. True.
2. False. Cat scratch disease is more common in humid climates because humidity is necessary for the existence of cat fleas.
3. False. Cat scratch disease adenopathy develops slowly, usually over 10-14 days.
4. False. With hepatosplenic CSD, LFTs are usually normal, and only 50% of patients have concomitant lymphadenopathy.
5. True.
6. True.

Chapter VI.30. Malaria

1. a. *P. falciparum* is unique among malarial species in that it has mechanisms to adhere to vascular endothelial walls. This produces a microvascular disease, leading to poor perfusion and metabolic acidosis. This hypoperfusion can affect almost any organ in the body, but this is of greatest significance in that it can cause cerebral malaria, which can cause a change in consciousness and seizures. Long-term effects due to cerebral malaria can also be seen. *P. vivax* is the most common form of malaria, but produces a more milder form of the disease.

2. d. The fever of malaria can produce any pattern of fever, with *P. falciparum* most known for its lack of recognizable fever patterns. Classically, the release of merozoites from red blood cells all in one group at similar times causes an inflammatory response, the production of TNF-alpha, and a characteristic pattern of fever depending on the particular species. The fever occurs approximately every 48 hours (called tertian malaria) for *P. falciparum*, *P. vivax*, and *P. ovale*, and 72 hours (called quartan malaria) for *P. malariae*.

3. c. The life cycle of malaria is very complex. It starts with malarial sporozoites being released from the anopheles mosquito. In the pre-erythrocytic stage sporozoites travel to the liver, with the patient being asymptomatic during this time. Sporozoites form schizonts, which eventually produce thousands of merozoites. These merozoites are released from hepatocytes, and infect red blood cells, giving rise to the erythrocytic stage of the life cycle. The erythrocytes burst after infection, releasing merozoites, which is the major cause of the cyclical fever. These merozoites can infect new blood cells, or form gametocytes. Male and female gametocytes are taken up by the mosquito, where they reproduce and form new sporozoites, completing the life-cycle during the mosquito's next blood meal.

4. c. Sporozoites infecting the liver can form into schizonts and can also form hypnozoites. These can remain dormant in the liver, causing an infection months later. The dormant liver-stage of the malarial life cycle, seen in *P. vivax* and *P. ovale*, is effectively treated with primaquine.

5. g. The microvascular disease of *P. falciparum* can affect almost any tissue of the body, giving rise to the many clinical features of malaria.

6. e. Prophylaxis for malaria includes using permethrin impregnated mosquito nets, avoiding mosquito bites using 35% DEET, and chemoprophylaxis most commonly with chloroquine or mefloquine. The anopheles mosquito usually bites from dusk to dawn, not during the day, and it is during these times that travelers should be particularly careful.

Chapter VI.31. Protozoans and Parasites

1. *Diphyllobothrium latum* (fish tapeworm), *Clonorchis sinensis* (Asian liver fluke),
2. Pinworms (*Enterobius vermicularis*).
3. *Trichinella spiralis*, *Taenia solium*.
4. *Trichomonas vaginalis*, *Giardia lamblia*.
5. *Ancylostoma duodenale*, *Necator americanus*
6. Malaria (*Plasmodium vivax*, *falciparum*, *haematobium*, *malariae*), filariasis (*Wuchereria bancrofti*, *Brugia malayi*).
7. *Tryp cruzi* [reduviid bug vector, which is not really a fly, but it is a biting bug], *T. rhodesiense* and *T. gambiense* [tsetse fly vector], leishmania [sandfly vector], *Onchocerca* [*Simulium* blackfly vector], loa loa eye worm [*Chrysops* fly].
8. *Taenia solium* (neurocysticercosis), *Naegleria fowleri*, Toxoplasmosis, *Loa loa* (eye).

Chapter VI.32. Candida and Fungal Infections

1. b. *T. tonsurans* is the most common cause of tinea capitis in the United States.
2. False. Tinea capitis, "black dot" pattern, is caused by *T. tonsurans*. This is an endothrix infection, thus would not be visible by Wood's lamp. Diagnosis is best done with KOH prep or culture.
3. False. Tinea pedis is most common in preadolescent and adolescent males.
4. True. *C. albicans* often colonizes the gastrointestinal tract. In 57% of patients with oropharyngeal candidiasis, candidal diaper dermatitis is also seen (6).
5. d. All of the above. Tinea versicolor lesions present differently depending on the individual's natural skin color. In light skinned individuals they often appear as reddish brown scaly lesions. In darker skinned individuals they can appear as either hyperpigmented or hypopigmented macules.
 - 6a. tinea only.
 - 6b. candida only.
 - 6c. both.
 - 6d. both.
 - 6e. both.
 - 6f. both.

Chapter VI.33. Necrotizing Fasciitis

- 1.c. *Bacteroides* is the most common bacteria isolated in polymicrobial NF. *Staphylococcus*, *streptococcus*, and *clostridium* are also commonly found.
- 2.a. Plain films are routinely used to differentiate cellulitis and NF. MRI and CT are currently under investigation for utility, however, they are costly and time consuming. Answers b and c could be correct, but ultrasound (answer d) is not useful. If NF is suspected, surgical exploration is necessary and will yield the same information.
- 3.b. The M protein inhibits the activation of complement and prevents phagocytosis. The other virulence factors listed belong to the streptococcal species, but have different roles in causing infection.
- 4.a. *Clostridium* causes gas gangrene and crepitus, which characterizes Type III NF. The other bacteria listed are causes of Type I or Type II NF.
- 5.d. First line therapy for streptococcal NF is penicillin according to current guidelines. Unfortunately, one does not initially know that the NF is due to GABHS. Most anaerobes are penicillin sensitive. Adding clindamycin may be useful even if the organism is penicillin sensitive since it may inhibit protein synthesis (toxin production) in non-replicating organisms. For other organisms, anti-microbial therapy should be based on culture and sensitivity results when they are obtained.

Chapter VI.34. Lymphadenitis and Lymphangitis

1. Persistent enlargement despite empiric therapy, persistent enlargement or no improvement with negative laboratory work up, solid fixed mass, mass located in the supraclavicular area, accompanying constitutional signs of persistent fever or weight loss.
2. Self limited, systemic viral infections such as adenovirus, influenza, and RSV are most common. EBV and CMV also can present as acute bilateral cervical lymphadenitis.
3. *Staph aureus* and *Strep pyogenes* (group A strep). Suppuration is more likely to be present with *Staph aureus*.
4. Complete surgical excision of the node is required to avoid development of a draining fistula.
5. Nontuberculous mycobacteria and cat scratch disease are common. EBV, CMV, toxoplasmosis, histoplasmosis, HIV are other infectious etiologies. Malignant diseases such as leukemia, lymphoma and solid tumors such as neuroblastoma, rhabdomyosarcoma and nasopharyngeal carcinoma also need to be considered.

Section VII. Cardiology

Chapter VII.1. Congestive Heart Failure

- 1.c
- 2.False
- 3.c
- 4.False
- 5.b
- 6.False
- 7.a

Chapter VII.2. Acyanotic Congenital Heart Disease

1. False. The physiologic pulmonary hypertension present in a newborn can prevent blood flow across a septal defect or PDA. These can be detected several hours after birth or several days after birth. Other congenital heart disease lesions may remain occult for longer period of time.
2. False. An aberrant right subclavian artery originating below a coarctation will produce equal pressures in the right arm and leg.
3. VSD, ASD, PDA. Of these, VSD is the most common.
4. False. Development of collateral vessels to the lower body can produce palpable femoral pulses.
5. True.
6. Congestive heart failure and pulmonary edema may cause hypoxia. If the hypoxia is severe enough, visible cyanosis will result, although this can be overcome with oxygen and other treatments for pulmonary edema and congestive heart failure. Long standing excessive pulmonary blood flow leads to pulmonary hypertension and Eisenmenger's complex, right to left shunting and cyanosis.
7. False. They cannot hear the murmur of a VSD on day 1 because on day 1, pulmonary vascular resistance is still high, which restricts left to right flow through the VSD. On day 2, pulmonary vascular resistance is lower, so left to right shunting through the VSD increases making the murmur louder.

Chapter VII.3. Cyanotic Congenital Heart Disease

- 1.d
- 2.d
- 3.c
- 4.b
- 5.d
- 6.b
- 7.c

Chapter VII.4. Rheumatic Fever

- 1.d
- 2.d
- 3.a
- 4.b
- 5.c
- 6.e

Chapter VII.5. Carditis

1. b.
2. b. Choice a is too short of a course and choice c is the preferred treatment for methicillin sensitive *S. aureus* infective endocarditis.
3. d.
4. a. The patient had positive blood cultures (1 major), and (3 minors) fever greater than 38 degrees C, a predisposing structural cardiovascular lesion (VSD), and evidence of an immunologic phenomenon (microscopic hematuria).
5. e. No antibiotics are needed, because this particular patient has no risk factors for infective endocarditis.
6. e.
7. b. Although c may be associated with viral myocarditis, viral pericarditis is most likely self-limiting.
8. c. Answers b and d may not show any abnormal findings.
9. e.

Chapter VII.6. Arrhythmias

1. Atrioventricular reentrant tachycardia (AVRT) and AV nodal reentrant tachycardia (AVNRT).
2. Ebstein anomaly and L-transposition of the great vessels.
3. With the presence of a bundle branch block or with antidromic conduction.
4. Vagal maneuvers and intravenous adenosine.
5. False.

Chapter VII.7. Vascular Rings and Slings

1. b
2. c
3. c
4. b
5. b
6. right aortic arch, left aortic arch, connecting to the descending aorta.
7. A vascular ring involves the aorta and its branches. A vascular sling involves the pulmonary artery. In the vascular sling, the left pulmonary artery arises from the right pulmonary artery and compresses the trachea posteriorly.

Section VIII. Pulmonology

Chapter VIII.1. Interpretation of Blood Gases and Pulse Oximetry

1. This cannot be determined without knowing the hemoglobin or hematocrit of each patient. Patient A could paradoxically have a lower oxygen content if he has a substantially lower hemoglobin (severely anemic) than patient B.
2. This is a respiratory acidosis with metabolic compensation.
3. This patient has bronchopulmonary dysplasia (chronic lung disease) with chronic CO₂ retention and metabolic compensation. An alternative answer would be an adult with chronic emphysema. An incorrect answer is acute respiratory failure, because if the respiratory failure were acute, the patient would not have enough time for metabolic compensation and his bicarb would be 24 or lower.
4. Visible cyanosis requires a certain amount of deoxygenated hemoglobin which is why the answer to this question depends on the hemoglobin or hematocrit. Patients with low hematocrits require a lower pO₂ for visible cyanosis compared to patients with higher hematocrits. So there is no single answer to this question. For example, a patient with cyanotic congenital heart disease may have a high hemoglobin to compensate. If his chronic oxygen saturation is 80%, he can compensate by having a higher hemoglobin such as a hemoglobin of 16. He will be visibly cyanotic because 20% (100% minus 80% oxygen saturation) of his 16 hemoglobin is desaturated (i.e., 3.2 Hgb is desaturated). For a normal child with a hemoglobin of 13 to have 3.2 desaturated Hgb, this child would have to have 25% (3.2 divided by 13) desaturation (i.e., an oxygen saturation of 75%). Thus, one patient may look bluer at 80% saturation, while another would be less blue at 80% because of different hemoglobins.

5. The pH should be low. The bicarb should be low (metabolic acidosis). There should be some respiratory compensation so the pCO₂ should be low (hyperventilation or Kussmaul respirations). The pO₂ should be fairly normal. So an example might be pH 7.14, pCO₂ 30, pO₂ 100, bicarb 10, BE -17.

6. This is a cardiac arrest so the patient is probably intubated. The high pCO₂ indicates that the patient is being hypoventilated or the endotracheal tube is not in the trachea. Proper placement of the endotracheal tube should be confirmed. The tidal volume and respiratory rate need to be increased to increase the minute ventilation to decrease the pCO₂. Better chest compressions to improve pulmonary blood flow will also facilitate the removal of CO₂. The bicarb is low causing a metabolic acidosis. Sodium bicarbonate can be given intravenously to reverse the metabolic acidosis.

7. Their color is pale. Thus pallor can suggest anemia, poor skin perfusion or hypoxia.

8. CO poisoning.

Chapter VIII.2. Asthma

1. Asthma is best thought of as a chronic inflammatory condition consisting of obstruction of the airways of the lung caused by spasms of the smooth muscle surrounding the airways which, in some cases, can be easily reversed by beta adrenergic bronchodilators. In other cases, corticosteroids may be necessary to reverse the airway obstruction by reducing the inflammatory changes responsible for the airway narrowing. These changes may be caused by a variety of different stimuli.

2. Medications are divided into groups directed towards relaxing bronchial smooth muscles (relievers) and reversing the inflammation (controllers).

3. This answer can be divided into two parts. The first is used to describe the degree of severity of the acute asthmatic episode. These would include rate and effort of respirations, ability to move air through a peak flow meter or spirometer, and oxygen and carbon dioxide concentration in the arterial blood. The second parameter involves the sensitivity of the airways (i.e., the chronic severity classification described in the chapter). Day symptoms, night coughing episodes, peak flow, coughing with exercise, prolonged coughing after upper respiratory infections, and coughing with drinking ice-cold beverages help to categorize the severity of asthma.

4. Wheezing may be heard but if the attack is very severe there may be no wheezing at all (due to poor air exchange). Aeration is a good indicator of acute severity. Evidence of respiratory distress (retractions, tachypnea) indicates increasing severity until respiratory failure occurs (at which point, the patient may tire and exhibit seemingly less respiratory distress). Hypoxemia is also indicative of severity. Peak flow is typically low for acute exacerbations. For mild cases, cough may be present at any phase of an asthmatic episode and may be the only sign that bronchospasm is occurring. A peak flow meter reading before and after a challenge of inhaled bronchodilator may reveal an increase in the airflow indicating the presence of bronchospasm.

5. Always consider the triggering event in formulating the treatment plan. Avoidance of the trigger can be very cost effective. Preventive use of medications can be very useful such as preemptive use of medication with first sign of a cold. Analysis of the symptom's response to initial treatment can guide you in up regulating or down regulating medications. Use of the peak flow meter can serve as an objective means of adjusting medications. If cough and wheezing occur often and there are signs/symptoms of chronic asthma, a maintenance plan of daily medication should be initiated. Efforts should be made to approximate the degree of inflammation in the airways. This estimation can serve to guide you in the type and dosage of anti-inflammatory medications to use. A contingency plan of what medications to use during an acute episode can be helpful and may help to avoid an unnecessary emergency visit to the hospital.

6. The asthma maintenance plans are dependent on the patient's severity class (step 1, 2, 3, or 4). For all "persistent" levels, a daily plan will usually involve a long-acting bronchodilator and corticosteroid, LTRA, cromolyn and/or theophylline two to three times a day. Regular monitoring with peak flow meter readings can help to determine if the treatment is helping to return the lungs to normal function. A "rescue" plan using short acting bronchodilators with optional systemic corticosteroids may be needed for breakthrough wheezing.

7. Allergen exposure is mediated through IgE with resultant immediate and late phase reactions. A variety of mediators are released and cause a cascade of immunologic events culminating in tissue edema, increased mucous production, and sloughing of the epithelial layer of the inner lining of the airways. This affects the free and easy movement of air to the alveoli, which affects air exchange and causes atelectasis as the smaller air ways are completely plugged by the thickened mucous.

8. Triggering mast cells cause release of mediators, which can cause immediate effects on the lung tissue and smooth muscles. Other mediators are formed and released later and serve primarily to attract inflammatory cells. Some of these late mediators help to capture the incoming cells. Other mediators recruit epithelial cells and transform them into participants of the reaction causing them to release more mediators (biologic amplification).

9. The critical issue of steroids in children is that of linear growth. It is now well established that the use of inhaled steroids has significantly less effect on growth than systemic corticosteroids. The length of steroid use (inhaled or systemic), may have some effect on growth but its effect is temporary and in many studies final growth of asthmatics is generally no different than in non asthmatics (i.e., catch up growth occurs if the corticosteroids can be stopped for a period of time long enough for this to occur). Chronic inflammatory suppression (long term use of inhaled corticosteroids) improves the long term outcome of asthma (i.e., less severity in the future).

10. This is where your ability to practice medicine is tested. You need to educate and persuade the parents that your recommendations are in the best interest of their child and that it is based on considering the risks against the benefits. This is ideally done without making the parents feel guilty or intimidated by the potential for fatal outcomes. While our goal may be to maintain the patient's lifestyle and lung function, patients may see their goal as getting off medications as soon as possible. For persistent asthmatics, they should be convinced that this is a chronic disease and long term medications will be required. Long term use of medications is generally very safe and not addictive.

Chapter VIII.3. Cystic Fibrosis

- 1.d
- 2.c
- 3.b
- 4.b
- 5.b
- 6.e
- 7.e
- 8.b
- 9.e
- 10.b

Chapter VIII.4. Chronic Lung Disease of Infancy (Bronchopulmonary Dysplasia)

1. False
2. b
3. e
4. e
5. d
6. False

Chapter VIII.5. Bronchiectasis in Children

1. true
2. false
3. false
4. true
5. false
6. false

Chapter VIII.6. Foreign Body Aspiration

1. False.
2. d.
3. First phase: Acute symptomatic period that immediately follows the incident. May see choking, gagging, coughing, and/or cyanosis. High risk of death. Second phase: Quiescent asymptomatic period. May last minutes to months depending on location, type, and ease of movement of the foreign body. Third phase: Renewed symptomatic period. May see wheezing, chronic cough, fever, hemoptysis. High risk of complication.

4. Organic material is worse to aspirate because it will cause a more intense inflammatory response, thereby increasing the risk for complications. Additionally, most organic material is non-radiopaque making it more difficult to visualize.
5. False. Right and left foreign bodies occur at roughly the same frequency.
6. A blind finger sweep may reposition the foreign body causing a complete airway obstruction.
7. d
8. d
9. True. Whenever a choking episode occurs while a young child is eating nuts, the risk of foreign body aspiration is high. Bronchoscopy should be highly considered here (9).

Chapter VIII.7. Pulmonary Hemosiderosis

1. c. Hypercarbia is not usually seen because compensatory mechanisms usually overcome the problems of reduced gas exchange by increasing minute ventilation (either by increasing rate or depth of ventilation).
2. It is one scheme to help identify the etiology for a condition with numerous causes. Treatment is more likely to be successful after identifying and treating the primary cause.
3. d. Bronchospasm, edema, and mucus can narrow the airway causing obstructive disease similar to asthma. Chronic inflammation can increase interstitial fibrin and collagen deposits which then reduce compliance resulting in giving restrictive disease. Any combination of the two is possible.
4. a. The classic triad is iron deficiency anemia, pulmonary infiltrates and hemoptysis, although hemoptysis is seen less commonly in children. "Pulmonary hemorrhage" does result in hemosiderosis, but it is not part of the classic triad.
5. False. There is controversy over whether a lung biopsy should be undertaken for all patients with significant PH. It could be argued that all patients who have PH without a known etiology (suspected IPH) should have a lung biopsy. But since IPH is a diagnosis of exclusion, a lung biopsy doesn't preclude the other parts of the evaluation, including history, exam, radiology and laboratory studies. Pathology from lung biopsy is seldom diagnostic alone and can only be interpreted in light of the other information.

Chapter VIII.8. Pulmonary Vascular Anomalies

1. >2:1 pulmonary flow to systemic flow.
2. To prevent future complications such as: pneumonia, arrhythmia, and irreversible pulmonary hypertension (13).
3. Recurrent pulmonary infections, bronchiectasis and hemorrhage.
4. Typically, it is left-to-right venous drainage: pulmonary venous/systemic artery to the systemic venous system. Intrapulmonary sequestrations typically shunt systemic blood to the pulmonary vein (systemic artery to the pulmonary vein, which is left to left).
5. Extrapulmonary sequestration (30% are associated with diaphragmatic hernias).
6. 1) Sequestration contains bronchi that do not communicate with the trachea. 2) Two types of sequestration (intrapulmonary and extrapulmonary). 3) Dextrocardia and ASD usually accompany Scimitar syndrome. 4) Intrapulmonary sequestration venous drainage enters the left heart, while the venous drainage of Scimitar and extrapulmonary sequestration enters the right heart circulation.

Chapter VIII.9. Bronchogenic Cysts and Congenital Cystic Adenomatoid Malformations

- 1.b
- 2.a
- 3.b
- 4.d
- 5.c
- 6.b
- 7.b

Chapter VIII.10. Congenital Airway Problems

1. Laryngomalacia
2. a. inspiratory
3. d. b and c
4. small or malformed cricoid cartilage
5. 20%
6. congenital subglottic stenosis
7. c. 18 to 24 months old
8. central, peripheral

Chapter VIII.11. Sleep Disorders

- 1.a,
- 2.b
- 3.a
4. Sleep attack, cataplexy, sleep paralysis, hypnagogic hallucinations
5. Obstructive: Tonsillar enlargement, Prader-Willi syndrome, Pickwickian. Non-obstructive: Prematurity, Ondine's Curse.
6. Adolescence

Chapter VIII.12. Sudden Infant Death Syndrome (SIDS)

- 1.False
- 2.b
- 3.False
- 4.c
- 5.True
- 6.b
- 7.True

Section IX. Gastroenterology

Chapter IX.1. Infant Colic

- 1.d
- 2.d
- 3.c
- 4.abcd
- 5.true
- 6.d

Chapter IX.2. Abdominal Pain

1. True
2. True
3. Crampy (hollow viscus) versus steady (solid viscus and peritoneal).
4. Mid-abdomen
5. Flank, groin and ipsilateral scrotum or labium

Chapter IX.3. Gastroenteritis and Dehydration

1. Shigella.
2. Rotavirus. It causes fever, vomiting, and watery diarrhea.
3. The diagnosis can be made by antigen detection, identifying cysts in the stool, endoscopy or examination of jejunal contents. It is treated with metronidazole or furazolidone.
4. Sunken fontanelle, absence of tears, sunken eyes, sticky/tacky oral mucosa, delayed capillary refill, reduced skin turgor, inactivity/lethargy, tachycardia, hypotension.
5. With oral rehydration, small frequent volumes 5-20cc every 5-10 minutes, advanced slowly.
6. With IV fluid infusion of normal saline or lactated Ringer's at 20cc/kg. Oral rehydration with ORS is commonly employed in other countries.

Chapter IX.4. Biliary Atresia

1. False. Persistent jaundice needs to be worked up before permanent damage is done by any number of pathological conditions, such as BA. Since there is little risk involved, the threshold to obtain a serum fractionated bilirubin should be low. If there is an elevation of conjugated bilirubin at 14 days of age or earlier, it is by definition neonatal cholestasis.
2. No. With poor uptake into the liver you can only state that there is cholestasis. This may be transient cholestasis due to hepatitis, or it may be due to severe damage to the hepatocytes by several possible causes including biliary atresia. To make a diagnosis, there needs to be normal uptake in the liver with no movement into the bowel, even after 24 hours. Pretreatment with phenobarbital improves the yield on the DISIDA scan.
3. Yes, this histopathology is consistent with the histopathology seen with biliary atresia. However, it is also consistent with idiopathic neonatal hepatitis and therefore a definitive diagnosis can not be made on this biopsy result alone.
4. The presence of clay colored or acholic stools are indicative of cholestasis. The lack of bile flow into the bowel prevents the characteristic stool coloring. The superficial light coloring is due to the sloughing of pigmented cells during the transit in the bowel and does not affect the core of the stool.
5. While there is little chance of long-term survival with the patient's native liver in someone who undergoes the Kasai procedure after 3 months of age, there is some benefit. The Kasai procedure can lead to extended survival time with the native liver, allowing the patient to stabilize baseline health. There is also a benefit in that there will be longer period of time to find a donor and prepare the patient for transplantation. However, each patient is different and some may be better served by primary liver transplantation.

Chapter IX.5. Hepatitis

1. These enzymes are found within the hepatocyte, and therefore are indicative of hepatocellular damage, and not actual function of the liver. The most useful test for liver function is prothrombin time.
2. False. They are usually anicteric.
3. The 15 month old should receive immunoglobulin (too young to receive Hep A vaccine). The 5 year old can receive the Hep A vaccine since she is over 2 years of age. The vaccine is given as two doses 6 months apart.
4. The mother is actually immune to hepatitis B, perhaps from receiving hepatitis B vaccinations in the past or from a previous exposure to hepatitis B. She does not have infection, she is not contagious and in fact, she is immune. This infant does not need HBIG prophylaxis, but should be vaccinated against hepatitis B in the usual fashion.
5. The mother has a positive HBsAg, which means that she is contagious. Therefore, this infant needs HBIG and hepatitis B vaccine prior to 12 hours of age. Because this premie is less than 2 kg, a 3-dose vaccine schedule should be instituted after this infant is over 2 kg, and not counting the initial dose because he was less than 2 kg. After completion of the 3-dose schedule, he should be tested serologically for anti-HBs and HBsAg 1-3 months after completion of the series. If his anti-HBs (<10 mIU/ml) is low and HBsAg is negative, then he should receive 3 additional doses of vaccine at a 0, 1, and 6-month schedule, with anti-HBs testing done 1 month later to determine immunity. The mother's status could be consistent with acute hepatitis B, chronic hepatitis B, or a hepatitis B carrier state. The most important serologic test out of the three listed is the HBsAg, since this test tells us whether the mother is contagious and the newborn requires HBIG prophylaxis.
6. Brain (or nervous system), liver, and eye. Manifestations are neuropsychiatric symptoms, hepatitis, and Kayser-Fleischer rings.
7. Copper.
8. Lung and liver. The pulmonary manifestation is emphysema and hepatic manifestations include prolonged jaundice in infants, neonatal hepatitis syndrome, mild elevations of aminotransferases in toddlers, portal hypertension and severe liver dysfunction in older children, and chronic hepatitis, cryptogenic cirrhosis, and hepatocellular carcinoma in adults.

Chapter IX.6. Gastroesophageal Reflux

1. False. Though most episodes are asymptomatic, reflux is a routine physiologic phenomenon in everyone, at every age. It is gastroesophageal reflux DISEASE that is uncommon in most of childhood.
2. d. Remember regurgitation is effortless, vomiting is forceful and is atypical for uncomplicated GE reflux. It can indicate obstruction or metabolic derangement, and represents a problem that requires an answer in as short a period of time as possible (even if the answer is a diagnosis of routine gastroenteritis).
3. Consider pyloric stenosis, even if only a few of the classic symptoms and signs are present. Waiting for the diagnosis to become more obvious further delays surgical intervention and increases the risk of complications such as hypochloremic alkalosis and dehydration. See differential diagnosis above.
4. This one is arguable, but my personal preference is to start treatment with antacids since it offers a means of immediate relief of any truly peptic pain episode, and younger children are better reinforced by immediacy of the response. Of course a good history and physical should come first to verify the pain does fit a "peptic" pattern, as constipation is more likely at this age.
5. False. The vast majority of uncomplicated pain seems to respond to mechanical measures, avoidance of caffeine, nicotine, and the like, and intermittent antacid use. It is only when the pain episodes remain disruptive more than once weekly that it is generally warranted to proceed to chronic medical therapy, and then only at the minimal doses necessary unless other complications (e.g., Barrett's esophagus) occur.

Chapter IX.7. Gastrointestinal Foreign Bodies

1. The level of the cricopharyngeus muscle in the proximal esophagus, the aortic arch crossover in the midesophagus, and the lower esophageal sphincter.
2. The esophagus because of its orientation.
3. False. A sharp object in the esophagus should be endoscopically removed immediately to prevent perforation.
4. False. The mercuric oxide in disk batteries is not readily absorbed by the GI tract.
5. Accumulations of plant and vegetable matter.
6. A penny cannot fit in an infant's trachea.
7. More gadgets which use disc batteries increases the likelihood that these batteries will be left around the house for young children to put into their mouths.

Chapter IX.8. Constipation

1. Answer d is correct, and the radiologist will appreciate the warning as to why the exam is being requested without prior bowel cleanout (which may otherwise be performed as part of the radiology routine, rendering the same end result as answer c). Answer a will not only miss the diagnosis but may also render diagnosis more difficult later if the pattern is set for stimulation for defecation. Answer b may give the diagnosis if a microcolon can be identified on exam, but can make interpretation of a barium enema difficult. Answer c is wrong for the same reasons as a and b. Answer e is doing too much too soon.

2. Correct answers are both b and c. Anal winks can be expected at any age unless the anus has indeed been badly traumatized. Its absence usually indicates a neurogenic component, and the examiner is prompted to carefully assess the tone of the sphincter and retrospectively look for other signs of aberrant function of the longer neuron sensory and motor tracts or signs of sacral anomalies. If the issue is still in doubt, it can be deferred by one visit. The process can still be addressed by full fecal softening and re-establishment of regular bowel habits since the therapies diverge at a later stage where a timing suppository needs to be added to maintain regular defecation as the weaning progresses and the stool becomes firmer. Full fecal softening is needed initially for both causes to address the flaccidity of the rectum.

3. No, the absence of impaction is worrisome, and the behavioral and social history are likely incomplete. The above pattern suggests voluntary soiling, in which a socially uncomfortable behavior is expressed to avoid an even more uncomfortable behavior, such as sexual abuse.

4. NO! The enemas may have dilated the rectum beyond the reach of the examining digit, and it is common for patients with short segment Hirschsprung's disease to pass the softer stools of breast feeding but have trouble with formula and pureed food. Expert radiographic evaluation is necessary, and the assistance of a pediatric surgeon or gastroenterologist may be helpful.

5. This is the typical appearance of the delayed view in a patient with Hirschsprung's disease. The obstruction is of high enough a grade that the portion of the colon with normal ganglion innervation has set up a "to and fro" pattern of peristalsis, evenly mixing the remaining barium with the increased fluids present in the lumen, rather than transporting the barium to the rectum where the excess fluid is removed (which is the appearance of the normal colon).

Chapter IX.9. Hirschsprung's Disease

1. false
2. true
3. No meconium for the first day of life.
4. false
5. neural crest cells

Chapter IX.10. Gastrointestinal Bleeding and Peptic Ulcer Disease

1. A modified Apt test can be done. Take the loose clots and suspend them in a minimal amount of tap water (you need a visibly pink supernatant composed of free hemoglobin, hence the tap water to lyse the cells). Centrifuge the cells and to 5 cc of pink supernatant add 1 cc of 1% sodium hydroxide. Read in two minutes: adult hemoglobin turns yellow or brown, fetal hemoglobin remains pink. If the supernatant turns yellow, the blood is mother's, and every one can relax.

2. This infant has no sign that the bleeding originates with him, as bleeding sufficient to produce melena should leave him quite shocky. The history gives every sign that he has induced a mastitis (and nipple bleeding) in his mother, and she is able to compensate for the several ounces of blood loss that produced the melanotic stool. You counsel her on proper feeding and handling techniques to keep the infant satisfied without having to overfeed, and have his mother avoid feeding on the affected side until the inflammation subsides. At followup in a week, all are smiling.

3. ABC's first. He shows no sign of acute intravascular volume depletion, but looks a little pale and turns out to be mildly anemic, indicating a longer standing problem. Next, place an NG tube to look for upper GI bleeding but you find no evidence of this. Now what? There is evidence of bleeding in an area bathed in acid, but it is not the stomach (or the duodenum). If he is hemodynamically stable, you have time to pretreat with a histamine-2 receptor blocker to improve the yield of a Meckel's scan looking for ectopic gastric mucosa. This finds a hot spot in the lower mid-abdomen which the technician assures you is not tracer in the bladder. You contact your pediatric surgeon for minimally invasive removal of a presumptive Meckel's diverticulum with acid-secreting ectopic gastric mucosa.

4. The black color is due to blood exposure to acid. Acid fermentation can take place in the cecum. If this occurs and the transit time is relatively slow, bleeding in this area can present as melena. Bleeding from a Meckel's can also result in acid exposure in the lower GI tract.

5. The acid level in the stomach is low (possibly due to antacids and H₂ blockers) and/or the bowel transit time is very rapid. Also, the bleeding may originate from the duodenum which does not expose the blood to acid if the pylorus is tight or the level of stomach acid is low.

6. The history has all the hallmarks of inflammatory bowel disease, but still the common things are more common. The physical examination shows no weight loss (but little net gain over the year), and she has a mild temperature elevation (100.5 degrees) and tachycardia (105) but no specific findings in the abdomen other than a mild increase in the amount of fluid and gas palpable in the small bowel and colon. Along with the CBC and ESR, you obtain a rectal swab for stool culture. There is no anemia, but the WBC count is slightly elevated and the ESR is 6. You are puzzled until the stool culture results return 2 days later, positive for *Campylobacter*. You call to discuss the results and find her new puppy had been ill the week before (dogs can both harbor and become ill from this organism), and the poor race performance actually arose because she was getting fed up with her coach (her father) and had been wanting to quit. Since she is still out of school with the cramping and diarrhea, you start her on erythromycin, offer to act as a go-between on the issue of changing sports, and annotate her chart to remind yourself to monitor for other signs of depression in the future.

7. As the negative gastric aspirates over the last 2 days indicate no UGI source, you prep him for colonoscopy to look for a lower GI bleeding site. GoLYTELY is used in hopes of diluting the bleeding as blood rapidly absorbs all light even in a thin film, and you anticipate much suctioning and lavage which will markedly extend the time for the procedure. As he will be under anesthesia anyway, you also obtain consent for EGD for completeness' sake. At endoscopy, the EGD study finds the pylorus is tightly shut as there is a large duodenal ulcer (not a simple erosion) with a visible vessel (an indicator of high risk of recurrent bleeding). With this you joyfully cancel the colonoscopy as being unnecessary, and chalk up the experience as a reminder that rapid transit times and the low acid production of early childhood can sometimes prevent the blood from encountering enough acid to turn to acid hematin or melena. Indeed, the higher the volume lost, the more acid is needed and the less likely the reaction. Unfortunately, as the finding was a therapeutic surprise, you are unprepared to address the ulcer in any invasive manner (sclerotherapy, heater probe, etc.) and have to return the patient to intensive care on an IV histamine receptor blocker and carafate and sufficient antacid to keep the pH of the gastric contents, measured every hour, above 6.5 (and well above the 4.5 activation level of pepsin). Preparations are made to return with the proper equipment the next day if he continues bleeding, only to find the bleeding stops with the procedure (and the drop in splanchnic pressures encountered under anesthesia), the current measures are more than sufficient, and no further transfusions are required. The patient makes a rapid and full recovery, with no recurrence in over 5 years (based on actual personal experience).

Chapter IX.11. Inflammatory Bowel Disease

1. CD affects the gut anywhere between the mouth to the anus, while UC affects the colon.
2. UC has a greater risk for cancer. CD only slightly increases the risk for cancer.
3. CD: Transmural inflammation, skip areas, aphthoid lesions, fissuring ulceration, granuloma, fibrosis.

UC: Mucosal inflammation, diffuse involvement, crypt abscesses, crypt distortion.

4. Joints: arthralgias are more common than arthritis. Integument: erythema nodosum and pyoderma gangrenosum. Eyes: episcleritis, uveitis, and rarely, orbital myositis. Hepatobiliary system: sclerosing cholangitis, chronic active hepatitis. Pancreas: pancreatitis. Renal system: nephrolithiasis, hydronephrosis. Coagulation system: hypercoagulability. Bone: decreased bone density.

5. Crohn's disease may be subdivided into 3 categories: 1) The fistulizing type, 2) Patients with fibrostenosing disease, and 3) The inflammatory category. Ulcerative colitis is divided into three categories: mild, moderate, severe.

Chapter IX.12. Malabsorption Conditions

1. Luminal phase, mucosal phase, and transport (removal) phase.
2. Luminal phase.
3. Mucosal phase.
4. False. Younger patients often display a more acute and wider-ranging symptomatology than older children.
5. True.
6. True.

Chapter IX.13. Meckel's Diverticulum

1. Meckel's diverticulum is the most common congenital anomaly of the gastrointestinal tract, affecting about 2% of the population.
2. The embryologic yolk-stalk or omphalomesenteric or vitelline duct
3. Meckel's diverticula appear in males and females at equal frequencies, however, males are 3 times more likely to develop symptomatic or complicated Meckel's diverticula.
4. Gastric mucosa is present in 80% of all heterotopic cases.
5. Most cases of Meckel's diverticula are asymptomatic, are detected at autopsy, or incidentally during unrelated abdominal surgery.
6. The infant or young child who has a massive, painless bout of dark red rectal bleeding most likely has Meckel's diverticulum.
7. The principal complications of Meckel's diverticulum include ulceration, hemorrhage, small bowel obstruction (may be due to volvulus or intussusception), diverticulitis, and perforation.
8. A Meckel's scan (technetium-99m pertechnetate scintigraphy).
9. False negative scans are seen in Meckel's diverticulum that do not contain ectopic gastric mucosa and in Meckel's diverticulum with rapid bleeding that prevents the accumulation of tracer in the diverticulum.
10. Meckel's rule of four 2's: a) Occurs in 2% of the population, b) Only 2% of those with a Meckel's manifest clinical problems, c) Usually located 2 feet proximal to the ileocecal valve and the diverticulum is approximately 2 inches long, d) Symptoms commonly manifest at age 2 years.

Section X. Surgery

Chapter X.1. Wound Management

1. Since epinephrine is a vasoconstrictor, it slows the rate of local anesthetic release into the general circulation permitting a higher total dose of local anesthetic that can be given (useful if the wound is large), it extends the duration of action, and decreases bleeding.

2. Lower tensile strength compared to sutures and thus it can't be used in areas of high tension such as wounds over joints. If it gets wet, the adhesive may fall off prematurely.

3. The research done on the comparisons between sutures and tissue adhesives have shown that they have comparable cosmetic results.

4. Approximately 7 to 10 days

5. Cocaine component: arrhythmia, urticaria, drowsiness, excitation, seizure, vomiting, flushing, and death. TAC should be avoided near mucous membranes. TAC is no longer available in most centers.

6. Significantly contaminated wounds, are at greater risk of infection if closed by primary intention.

7. True. Heavily contaminated wounds will develop infection despite antibiotic treatment.

Chapter X.2. Inguinal Hernias and Hydroceles

1. True

2. a

3. Intermittent inguinal, scrotal or labial swelling that spontaneously resolves.

4. True

5. c

6. True

7. b

8. True

Chapter X.3. Appendicitis

1. Movement alleviates colicky pain but exacerbates peritoneal pain.

2. 4 to 5 cm (1.5 to 2 inches) cephalad on a line drawn between the anterior-superior iliac spine and the umbilicus.

3. Persistent and constant in nature.

4. Genitourinary.

5. Literally "middle pain" caused by a ruptured ovarian follicle which occurs approximately in mid-menstrual cycle.

Chapter X.4. Intussusception

1.c

2.a

3.e

4.c

5.b,d,e

6.e

7.b

8.false

9.a

Chapter X.5. Malrotation and Volvulus

1. a) Ladd's bands compressing and obstructing the proximal small bowel. b) Midgut volvulus.
2. The term "malformation" originates from the embryological formation of the malrotation which is of little or no value for clinicians.
3. Upper GI series. Barium enema and ultrasound are less reliable.
4. Midgut volvulus and sigmoid volvulus. Midgut volvulus is a true surgical emergency involving nearly the entire small bowel which will infarct unless the volvulus is relieved surgically. Sigmoid volvulus, which occurs in the elderly, involves the sigmoid colon and can usually be relieved without surgical means.
5. It is unlikely, but it can happen. About half the patients with a malrotation will present in the neonatal period, with the other half presenting at any other age.

Chapter X.6. Gastroschisis and Omphalocele

1. c
2. c
3. d
4. c
5. c

Chapter X.7. Diaphragmatic Hernia

1. c
2. d
3. d
4. c
5. c

Chapter X.8. Pyloric Stenosis

1. A 3 to 4 week old male infant who presents with progressively severe, non-bilious vomiting, which may be projectile. The vomiting occurs immediately after feeding, after which the infant is still hungry and wants to feed again. On physical exam, the infant may display signs of dehydration. Visible waves of peristalsis may be seen and an "olive" may be palpable.
2. A palpable "olive" is pathognomonic but is very difficult to determine with certainty. If the pylorus cannot be palpated, ultrasound is diagnostic with 90% sensitivity.
3. The "classic" laboratory finding is a hypochloremic, hypokalemic metabolic alkalosis. However, due to more expedient diagnosis, this metabolic abnormality is seen in less than 10% of patients.
4. The initial step in management involves fluid resuscitation and correction of any metabolic abnormalities. HPS is not a surgical emergency, and any fluid deficits or alkalosis should be corrected prior to surgery to decrease surgical/anesthetic risks.
5. Electrolyte patterns are not pathognomonic for pyloric stenosis. The correct answers are a and c. Pattern "a" is a classic early vomiting picture, often seen with HPS. Pattern "c" is a picture of vomiting resulting in dehydration and lactic acidosis. This can also be seen later in the clinical course of HPS as dehydration worsens. Pattern "b" is typical of adrenal crisis (low Na, high K). Pattern "d" is typical of hypernatremic dehydration.

Chapter X.9. Intestinal Atresias, Duplications and Microcolon

1. Duodenal atresia.
2. Esophageal atresia with tracheoesophageal fistula results in a gas within the bowel, esophageal atresia without tracheoesophageal fistula does not.
3. VACTER association; includes vertebral defects, anal atresia, congenital cardiac anomalies, tracheoesophageal fistula with esophageal atresia, radial upper limb hypoplasia and renal defects.
4. Esophageal or duodenal atresias result from failure of the lumen to recanalize. A jejunal or ileal atresia results from an intrauterine ischemic event.
5. Undiagnosed intestinal duplications may cause a bowel obstruction or may undergo malignant transformations in adults.

Chapter X.10. Craniofacial Malformations

1a. The clefting is caused by improper migration of the lateral lip segments in utero. This is a complex process, and sometimes it malfunctions.

1b. Probably not, and reassurance is the best treatment as the parents will inevitably feel some guilt. For future pregnancies, good nutrition (especially folic acid) and avoidance of toxins (alcohol, cigarettes, drugs, medications, environmental) are helpful. For further discussion, see <http://www.cleft.net/reduce>.

1c. The parents will probably need help in learning how to feed their baby, since the baby has less ability to create suction. Making a larger opening in the nipple, and using a broad nipple can help - the baby can get milk by compressing the nipple with the tongue rather than sucking. Breast feeding is possible, but more difficult. For a nice discussion of this, see <http://www.samizdat.com/pp2.html>

1d. The surgeries involve repair of the lip in the first year, repair of the cleft palate at about age 1, repair of the alveolar cleft at 6-10 years, and repair of the cleft nasal deformity as a teenager, after growth is complete. Each of these may involve one major operation, and perhaps one or more refinement operations if desired.

1e. The incidence of cleft lip in the general population is about 1:750. This approximately doubles for each affected family member, so the next baby would have about a 1:375 chance of having a cleft. For more precise evaluation, consultation with a genetic counselor is recommended.

2. In cleft palate, the muscles of the soft palate (levator palatini) are incorrectly aligned: they cannot cross the midline as they normally do. Thus, contraction of these muscles does not pull on the Eustachian tube to open it up, and the ears remained "plugged", causing serous otitis which then can get infected and cause otitis media.

3. Because of the clefting of the palate, the children cannot build up air pressure in the mouth (the air escapes into the nose). Thus, they cannot properly form the sounds which require increased air pressure (b, p, t, k, g, v, and s). As they try to learn to speak, they substitute other sounds for the ones that they cannot make ("compensatory articulations"). As they get older, it becomes increasingly difficult for them to unlearn these habits, so repair of the cleft palate should be done prior to speech development if possible. In addition, hearing is often slightly impaired, as noted above.

Chapter X.11. Abscesses

1. True

2. Staph aureus

3. False, lung abscesses are not.

4. Abscesses are often mixed infections, therefore antibiotic treatment needs to provide adequate coverage of the common bacteria associated with that type of abscess. Some antibiotics (notably clindamycin) may provide synergistic efficacy as well.

5. No

6. Bacteremia, rupture into neighboring tissue, bleeding by erosion into nearby vessels, impaired function of the affected organ or systemic effects such as cachexia and anorexia.

Chapter X.12. Lymphangiomas

1.e. A subdural hygroma is liquefaction of a subdural hematoma.

2.a & b. These are indicative of venous malformations.

3.true

4.d. Radiation is reserved as a last resort.

5.f

Section XI. Hematology

Chapter XI.1. Anemia

1. Classification by red blood cell size (microcytic, normocytic, and macrocytic anemias) and classification by mechanism (decreased production, increased destruction, and blood loss).
2. Low reticulocyte count.
3. History: dark urine. Physical exam: jaundice, scleral icterus, splenomegaly. Lab: elevated LDH, AST, indirect bilirubin; decreased serum haptoglobin; positive direct antibody test (DAT, also known as Coombs test), high reticulocyte count.
4. Bone marrow stain for iron has the highest positive predictive value and specificity, but it is too invasive in most instances. Low serum ferritin is diagnostic of iron deficiency, but its wide range of normal values and its fluctuation with acute inflammation may make interpretation difficult. Serum iron coupled with TIBC and % iron saturation are satisfactory, but this test is subject to some laboratory fluctuation as well. Response to a therapeutic trial of iron is also acceptable as proof of iron deficiency. No actual correct answer to this question.
5. True
6. True
7. False
8. False. Cow's milk contains a modest amount of iron, but little of it is bioavailable.
9. c

Chapter XI.2. Thalassemia

1. Answer is d. Since the child had Hemoglobin Barts on the newborn screen, a form of alpha thalassemia is present. The hemoglobin of 9.1g/dl implies that it is likely Hemoglobin H thalassemia. There is no need to do a hemoglobin electrophoresis, since the type of thalassemia (alpha) is already known. Additionally, Hemoglobin H is so fast moving that it is typically missed on routine hemoglobin electrophoresis, thereby giving "normal" results. In general, therefore, hemoglobin electrophoresis is typically useless in evaluating for alpha thalassemia. This patient and her family should be provided with genetic counseling and education. She should be counseled to avoid supplemental iron, as a true iron deficiency is extremely rare in Hemoglobin H thalassemia. If iron deficiency is ever suspected, iron studies should be done to clearly document a true deficiency before iron supplementation is started.
2. Answer is b. The two most likely etiologies of the anemia in this young lady are iron deficiency or a form of thalassemia. She could most effectively be managed with a trial of iron (for one month). If a repeat CBC shows no change, then either alpha or beta thalassemia should be considered. A hemoglobin electrophoresis would be the next step if the iron trial fails. An increase in Hemoglobin A2 is very suggestive of beta thalassemia. In this case, the mild anemia would indicate a heterozygous beta thalassemia (beta thalassemia minor). Workup may stop there with proper genetic counseling and patient education. If the hemoglobin electrophoresis is normal, or near normal, then alpha thalassemia is the most likely cause.
3. Answer is C. The effects of Hemoglobin E are most significant when combined with beta thalassemia minor (see text), which is why the newborn's current hemoglobin (mostly fetal hemoglobin with no beta chains) is of the least concern. A CBC should be done at 9 or 12 months of age to screen for coexisting beta thalassemia.
 - 4a. Fe is indicated as a therapeutic trial. But if no improvement in the hemoglobin results, then a thalassemia is possible.
 - 4b. Fe is contraindicated since it will not improve the hemoglobin and it will add to the potential for iron toxicity.
 - 4c. Fe is contraindicated, since it will not improve his hemoglobin and it will add to the potential for iron toxicity.
 - 4d. Despite the presence of thalassemia, iron deficiency is documented by laboratory studies, so iron supplementation is indicated until iron deficiency resolves. Once iron deficiency is no longer present, iron supplements become contraindicated.

5. The four alpha genes are not inherited independently. They are inherited in pairs on each chromosome. Thus, a patient with alpha thal trait who has two defective alpha genes and two normal alpha genes could have this in one of two ways: 1) AX/AX, or 2) AA/XX, where "A" is a normal alpha gene and "X" is a defective alpha gene. Some ethnic groups have the genes arranged in the first form only, in which case, two parents with alpha thal trait would always pass AX to their child resulting in a child with AX/AX (alpha thal trait). Fetal hydrops (XX/XX) could never result from such a genetic arrangement. However, if both parents with alpha thal trait were AA/XX, then their children could either be: AA/AA, AA/XX, or XX/XX (fetal hydrops).

Chapter XI.3. Sickle Cell Disease

1.c. This fever is significant, thus there will be an increase in sickling, and the patient is at risk for vaso-occlusive events. Therefore, IV hydration is necessary. It is also prudent to start empiric antibiotics after blood cultures are obtained.

2.b. Appropriate initial management should include vigorous IV hydration, plus IV pain management to include both a continuous infusion and a PCA. One would not transfuse initially, because a transfusion of packed red blood cells will only increase the viscosity of the blood, causing more sickling. Also, one does not know at this point, what the baseline hemoglobin is. The hemoglobin of 7.9 g/dl may not be very different than baseline. If there is further hemolysis, and a transfusion is indicated, it should be done carefully after several hours of IV hydration. Also, remember that meperidine increases seizure activity in children with sickle cell anemia, and is contraindicated.

3. It has been shown that a proactive approach to sickle cell disease decreases morbidity and mortality. Therefore, by identifying all children with sickle cell disease at birth, before symptoms start (usually after 1 year of age), quality of life can be improved.

4. Only after 6 months of age is gamma globin chain production decreased and beta globin chain production sufficient to cause sickling.

5. No. Both beta and sickle anomalies are on the beta globin gene. The newborn screen will identify the sickle hemoglobin, but will not identify the abnormal beta globin genes. The newborn screen will therefore appear as that for sickle cell trait with Hemoglobins F,A, S.

Chapter XI.4. Bone Marrow Failure

1. Stem cell transplantation from a matched sibling donor or other compatible stem cell source.
2. Diepoxybutane induced chromosome breakage (increased in patient with Fanconi's anemia).
3. Diamond Blackfan anemia presents at an earlier age (<1 year) and may have associated physical anomalies. At diagnosis, MCV, hemoglobin F and i-antigen are increased. TEC presents at an older age (>1 year). Since it is acquired, there are no associated anomalies. MCV, hemoglobin F and i-antigen should be normal.

4. Skin hyperpigmentation, mucous membrane leukoplakia, dystrophic nails.

5. Cow's milk allergy and thrombocytopenia.

6. Viruses: hepatitis, HIV and parvovirus. Drugs: Chemotherapy, chloramphenicol.

7. The i-antigen is a marker found on immature red cells. It, along with fetal hemoglobin and macrocytosis are manifestations of the fetal-like hematopoiesis seen in the stressed bone marrows of patients with acquired aplastic anemia, Fanconi's anemia, and Diamond Blackfan anemia.

Chapter XI.5. Newborn Hematology

1.F

2.F

3.d

4.F

5.T

6.T

Chapter XI.6. Bleeding Disorders

1. ITP is an immune-mediated disorder in which circulating antiplatelet antibodies target epitopes on the platelet membrane. The antibody-coated platelets are subsequently destroyed by macrophages in the reticuloendothelial system.
2. Thrombocytopenia, microangiopathic hemolytic anemia, and uremia.
3. X-linked recessive.
4. Following trauma or injury, especially head injury; to treat spontaneous bleeding, such as hemarthrosis or deep muscle bleeding, and prior to procedures, including dental work.
5. von Willebrand factor is a cofactor for platelet adhesion and carrier protein for factor VIII.
6. Bleeding time, PTT, and ristocetin cofactor.
7. Patients who have blood group O have a lower normal range for von Willebrand studies.
8. Factors II, VII, IX, and X.
9. If the prolonged PTT is due to a factor deficiency, then the addition of factors from the "normal plasma", will correct the PTT. However, if the PTT is due to a circulating anticoagulant such as heparin or a lupus anticoagulant, the circulating antibody will inhibit the "normal" factors and the PTT will remain prolonged. Failure to normalize the PTT after the addition of normal plasma, implies the presence of a circulating anticoagulant.

Chapter XI.7. Transfusion Medicine

1. b. Once a unit is spiked (IV infusion begun from unit bag), any uninfused blood must be discarded after 4 hours. Thus, the most time allowed for 1 unit to run is over 4 hours. Therefore, a unit may not be transfused over 6 hours. Giving 390 ml would give this patient 15 ml/kg, but giving this over 4 hours would be slightly too fast with such a low and fast falling hemoglobin. Additionally, it would expose the patient to a second donor, and half of the second unit would be discarded (wasted). Giving 262 ml means giving 1 unit (about 250 ml), and about 10 ml from a second unit (discarding the rest). Giving this over 2 hours would also be too fast as noted.
2. b. For just a few hives, it is not necessary to check the crossmatch of the blood, since this will detect antibodies causing hemolysis. Urticaria is not a hemolytic reaction. Usually diphenhydramine alone can resolve the hives, and the same unit can be continued with the diphenhydramine in effect.
3. c. See text
4. d. Epinephrine has no known beneficial effect on the hemolytic process.
5. e. All of these children should probably receive a transfusion.
6. a. See text
7. d. A unit of PRBCs can be split in the blood bank (like neonatal units) so that only one part of this is out of the blood bank and infusing into the patient at a given time (which can infuse up to 4 hours). Additionally, a child with such an extremely low hemoglobin needs to be transfused very slowly, at least initially, so as not to push his already compromised heart into further failure. With severe anemia, the patient is already in high output congestive heart failure. Blood is a potent volume expander which can suddenly worsen the CHF. Thus, the transfusion must proceed very slowly under close hemodynamic monitoring.

Chapter XI.8. Neutrophil Disorders

- 1.b
- 2.c
- 3.e
- 4.d
- 5.e

Section XII. Oncology

Chapter XII.1. Oncology Treatment Principles

1. Common infections include candidiasis, aspergillosis, and *Pneumocystis carinii*. Prophylactic treatment with trimethoprim/sulfamethoxazole is indicated.
2. Hormones (prednisone), antimetabolites (methotrexate, 5-fluorouracil), plant alkaloids (etoposide, vincristine, paclitaxel), antibiotics (doxorubicin, bleomycin), anti-angiogenesis drugs.
3. Leukoencephalopathy
4. Most of these work by inhibiting some metabolic pathway or DNA synthesis, which ultimately leads to cytotoxicity
5. Bone marrow, peripheral blood, and sometimes even cord blood.

Chapter XII.2. Leukemia and Lymphoma

1. b. The fact that the child is short of breath in the supine position could be related to a mediastinal mass, which can be identified on a chest x-ray. A mediastinal mass could be a potential emergency situation, therefore a chest x-ray should be considered shortly after the history and physical exam are completed.
2. c. Live vaccines are contraindicated throughout the treatment course due to the immunocompromised status of the patient.
3. d. Delays in growth and development may occur as a result of chemotherapy and/or radiation therapy.
4. b. The chemotherapy may have induced tumor lysis causing hyperuricemia, which in turn may be affecting the kidneys.
5. b. As part of the differential diagnosis, you should consider ITP.

Chapter XII.3. Solid Tumor Childhood Malignancies

1. d
2. c
3. b
4. Growing pains (e) are ill-defined, but are supposedly very common, so from a numerical standpoint, this diagnosis is probably the most common. However, since this age group is one of the peak ages for osteosarcoma (b) and since this is a serious condition that should be diagnosed as early as possible, osteosarcoma is the most serious likely consideration.
5. a, b, and d are correct.

Chapter XII.4. Palliative Care

1. True
2. False
3. True
4. True
5. True

Section XIII. Nephrology/Urology

Chapter XIII.1. Nephritic Syndrome

1. C3 levels return to normal within a 6-8 week period in APSGN. Persistently low C3 levels suggest a cause other than APSGN.
2. The presence of red cell casts on urinalysis almost always indicates the presence of glomerulonephritis. They can also be seen after strenuous exercise and renal trauma.
3. The presence of white cell casts on urinalysis can be seen in APSGN, interstitial nephritis and pyelonephritis.
4. Gross hematuria resolves within days to weeks. Microhematuria may persist for months.
5. An uncertain diagnosis, significant hypertension, anticipated poor follow-up, cardiovascular or cerebrovascular compromise, etc.
6. Gross hematuria, oliguria, hypertension, edema (usually mild).
7. APSGN and Goodpasture's. Other causes of nephritis include SLE nephritis, MPGN, RPGN, Alport's, etc.
8. Convalescing APSGN.

Chapter XIII.2. Nephrotic Syndrome

1. d. Minimal change disease or "nil disease" accounts for 80-85% of cases of primary idiopathic nephrotic syndrome in childhood.
2. b and d. Infection, especially peritonitis and thrombosis account for the majority to nephrotic syndrome mortality.
3. false. The decision to perform a renal biopsy is usually deferred until the initial course of corticosteroid is initiated, unless there are specific risk factors such as age below one or above 10, hypertension on presentation or decreased complement on presentation.
4. d. Primary nephrotic syndrome is sporadic in nature. Congenital nephrotic syndrome is passed in an autosomal recessive manner.
5. b and e. Nephrotic syndrome in a child less than 1 year old may indicate congenital nephrotic syndrome and renal biopsy is often performed. In a patient with SLE, the nephrotic syndrome is likely secondary and a renal biopsy is indicated.

Chapter XIII.3. Cystic Kidneys

1. ARPKD and ADPKD are inherited. MCDK is usually non-heritable. ADPKD is the most common inherited renal disease.
2. ARPKD: bilateral enlargement and microcysts on ultrasound. Hepatic fibrosis is also present in ARPKD. ADPKD: macrocysts and usually involve extrarenal cysts in the liver, pancreas, ovary, and/or spleen. ADPKD will also have a positive family history in a parent and the aunts/uncles on the affected parent's side of the family.
3. Unilateral MCDK has an excellent prognosis with most cases decreasing in size. ARPKD, since it is bilateral, eventually leads to end-stage renal disease.
4. Signs of portal hypertension: spider nevi, esophageal varices, hepatomegaly. There can also be signs of respiratory distress or abnormal feeding due to the compressive effects of enlarged kidneys.
5. No, extrarenal manifestations of ADPKD such as intracranial aneurysms and extrarenal cysts usually present in adulthood.

Chapter XIII.4. Dialysis

1. Renal failure with uremia; BUN over 150 mg/dl; creatinine over 10 mg/dL; severe hyperkalemia; severe acidosis; refractory fluid overload (CHF); certain inborn errors of metabolism; certain acute poisonings; tumor lysis syndrome.
2. In hemodynamically unstable patients.
3. Can be done at home; no complex machinery; no vascular access.
4. Hypotension, seizures, hypothermia.
5. Anemia; acidosis; hypertension; growth retardation, renal osteodystrophy, platelet dysfunction.

Chapter XIII.5. Hemolytic Uremic Syndrome

1. E. coli O157:H7
2. Microangiopathic hemolytic anemia, thrombocytopenia, and renal failure.
3. d. schistocytes
4. c. serum BUN >100
5. False
6. Crampy abdominal pain (due to colitis), crying with puffy eyes (due to abdominal cramps, fluid retention due to renal failure causing puffy eyes), currant jelly diarrhea (actually bloody diarrhea due to E. coli O157:H7), pallor (due to hemolytic anemia), dehydration (due to diarrhea), oliguria (due to renal failure).

Chapter XIII.6. Urinary Tract Infection

1. Empiric treatment for UTI should not be initiated without first obtaining an adequate specimen for culture. The only pediatric exception would be a child so severely ill (in septic shock and/or anuric) that waiting to obtain a urine sample could be life threatening. One might consider empiric treatment without culture in an uncomplicated older teen, however, such patients are rarely "uncomplicated" when considering issues such as recurrence, sexually transmitted diseases, etc.
2. The method of obtaining a urine specimen is affected by the patient's age, severity of illness, state of cooperation, toileting abilities, and whether or not antibiotics are to be started empirically. The colony count considered positive varies with the collection method: any growth with suprapubic aspiration; greater than or equal to 10,000 CFU for a catheterized specimen; and greater than or equal to 100,000 CFU for a clean catch specimen. Bag specimens are only definitive when culture result is negative (and therefore should not be used if empiric therapy is to be initiated).
3. Host factors contributing to development of UTI include uncircumcised male, labial adhesions, poor hygiene, constipation, urinary tract obstruction, dysfunctional voiding patterns, and neurogenic bladder. Pathogens are those commonly found in the vicinity of the urethra: skin and GI organisms, as well as blood-born organisms in the neonate. The strains of E. coli which commonly cause UTI show increased adherence to uroepithelial cells.
4. Classical signs of pyelonephritis include CVA tenderness, fever, and signs of systemic illness, while lower tract disease is milder and may present with only urinary urgency, frequency, or dysuria. Abnormal DMSA scan or elevated CRP results support the diagnosis of pyelonephritis.
5. The commonest presentation of UTI in the child under two years of age is fever. Associated signs and symptoms may include vomiting, diarrhea, irritability, poor feeding, malodorous urine, oliguria, constipation, or jaundice.
6. Empiric parenteral therapy and/or hospitalization should be considered when suspected UTI is associated with signs of urosepsis, severe clinical illness, dehydration, immunologic compromise, or urologic abnormality. Vomiting, poor oral intake, or concerns for poor compliance are also reasons to use parenteral therapy.
7. "Clean catch mid-stream" urine sample means that the urethral meatus and surrounding area should be clean, and that the urine collected should be from the middle of the stream: i.e., the first few drops of urine should not be collected. For girls, cleaning involves separating the labia and cleaning the area (usually with a series of 3 pre-moistened antiseptic towelettes). For circumcised boys, the glans of the penis should be similarly cleansed. For uncircumcised boys the foreskin is gently retracted prior to cleaning. After cleaning, the child voids over the toilet, with the parent "catching" the urine in a clean specimen cup after the first few drops are passed. In girls this is often more easily accomplished by having the child sit facing backwards on the toilet, so the parent can easily catch the urine stream from behind the child.

8a. Transurethral catheterization is an invasive procedure and is performed using standard sterile technique, including povidone/iodine wash of the periurethral and perineal areas, sterile field, sterile gloves, and sterile catheter and specimen cup.

8b. Infant feeding tubes in #5 or #8 french size are adequate for most infants and toddlers. It is not necessary or advisable to use a Foley catheter, as there is no need for a balloon. The catheter is removed as soon as the sample is obtained.

8c. The catheter is introduced into the urethral meatus, and advanced gently until there is return of urine. This is done with the infant in the supine, "frog-leg" position. The catheter tip may be lubricated with sterile lubricant or sterile water. In circumcised boys the urethral meatus is easily seen. In uncircumcised boys it is usually revealed by gentle retraction of the foreskin (if not, the foreskin is retracted as far as is easily possible and the catheter introduced with gentle probing until the meatus is located). The urethral meatus may be less easy to see in infant girls. It is helpful to remember that it lies anterior to the vaginal introitus, and to be familiar with the often fleshy appearance of the infant hymen. Separation of the labia, adequate light, and familiarity with the appearance of the genitalia facilitate locating the urethral meatus. A frequent error is introduction of the catheter into the vagina (recognized by the absence of urine return and by resistance to gentle advancement of the catheter beyond a couple of centimeters). Some practitioners opt in this situation to leave the misdirected catheter in place while a second catheter is introduced into the urethra (using the first catheter to "block" or mark the vaginal introitus). Whether or not the first catheter is left in place, a new sterile catheter must be used for the second attempt, to avoid contamination with vaginal flora.

8d. Complications of urethral catheterization include doubling back of the catheter (either in the urethra or in the vagina), trauma to the urethral meatus or mucosa, and possible introduction of infection. There can be subsequent stricture formation. Familiarity with the anatomy and avoidance of any forceful catheter advancement can minimize the risk of complications. A lubricated catheter of appropriate size should advance easily through the urethra. Any resistance should be taken as a sign to retract the catheter rather than to advance it more forcefully. The risk of introduction of infection is minimized by careful adherence to sterile technique.

Chapter XIII.7. Hydronephrosis and Reflux

1. Hydronephrosis represents 50% of all abnormalities detected with prenatal US.
2. A renal and bladder US should be obtained on day 2 of life. US done earlier may yield a false negative (no hydronephrosis) due to low urine output not distending the collecting systems. If it is normal, then the US should be repeated at 1 month of age, and be normal before considering the hydronephrosis to have resolved.
3. A VCUG should be obtained to evaluate for posterior urethral valves. If PUV are present, the VCUG will show a prominent bladder neck, a dilated posterior urethra, with a bulging membrane at the distal aspect of the verumontanum. The bladder may be thickened. Reflux may be present. The treatment is centered on securing adequate drainage of the urinary tract; initially by placement of a urinary catheter, and later by transurethral ablation of the valves. A vesicostomy (surgical formation of a cutaneous bladder stoma) may be done as a temporizing measure if the infant cannot undergo transurethral ablation of the valves.
4. Ureteropelvic junction (UPJ) obstruction is the most common cause, with ureterovesical junction (UVJ) obstruction being the second most common cause of congenital hydronephrosis. They are distinguished by the fact that with UPJ obstruction, the ureter is not dilated, whereas the ureter is dilated with UVJ obstruction.
5. The infant should be placed on antibiotic prophylaxis (with penicillin) and a VCUG and diuretic renal scan done at 4 to 6 weeks of age.
6. In infants noted to have good (35 to 40% or greater) split function on the renal scan, then serial ultrasound and diuretic renal scans (at 3 to 6 months of age, then at 12 month of age) may be used to follow the patient nonsurgically, on antibiotic prophylaxis. If there is renal function deterioration, breakthrough UTIs, or symptoms of renal colic, then surgery (pyeloplasty in UPJ obstruction, and ureteral reimplant in UVJ obstructions) is indicated. Only 25% of children with UPJ obstructions will require conversion to surgical management.
7. A ureterocele is a cystic dilation of the distal ureter at the level of the ureteral orifice. A ureterocele which has prolapsed into the urethra is the most common cause of congenital bladder outlet obstruction in females. Transurethral incision of the ureterocele is a minimally invasive treatment for symptomatic ureteroceles.

8. The ectopic insertion of the ureter into the bladder wall laterally results in a short intravesical ureter (a short submucosal bladder tunnel), which acts as an incompetent valve during urination, allowing urine to reflux back up into the ureter.

9. The antibiotic prophylaxis sterilizes the urine, and thus prevents bacteria ascending up the refluxing ureters, from causing pyelonephritis and renal scarring/damage. This allows time for normal growth and development of the ureter and bladder to occur. With growth, lengthening of the submucosal bladder tunnel/intravesical ureter results in the resolution of reflux over time, particularly in those with lower grades of reflux. Observation includes serial cystograms (usually nuclear scintigraphy) every 12 to 18 months.

10. The failure of medical management (and thus the need for ureteroneocystostomy) is indicated by breakthrough UTIs, the development of new renal scars, or the failure of reflux to resolve over time. Non compliance or allergic reactions to the prescribed antibiotics may also lead to the failure of medical management.

Chapter XIII.8. Circumcision

1. Penile cancer, balanitis, phimosis, urinary tract infection, reduced risk of HIV.
2. Gomco clamp, the Bronstein (Mogen) clamp, and the Plastibell.
3. Bleeding and infection.
4. Hypospadias, chordee, epispadias, penile torsion, micropenis, significant prematurity, blood dyscrasia, or family history a bleeding disorder.
5. Yes or no; see reasons associated with each answer. Yes, because it protects against penile cancer, etc., (see #1). No, because of the risks of complications of infection, bleeding, concealed penis, penile adhesions, meatitis, fistula formation, penile amputation and penile necrosis.

Chapter XIII.9. Enuresis

1. Typically at age 5 or 6 years.
2. False.
3. Imipramine.
4. Urinalysis with specific gravity, glucose, protein, blood and white cells.
5. Most adults have a bladder capacity between 250-400 ml, but the average bladder capacity in children can be approximated by the formula: volume (oz.) = 2 + age in years.
6. The abdominal exam should assess for masses secondary to enlarged urinary organs (bladder, kidney) and for evidence of palpable stool in the colon suggesting fecal impaction.
7. True.

Chapter XIII.10. Acute Scrotum

1.	Acute testicular torsion	Epididymitis
Onset	Acute	More gradual
Fever	Absent	May be present
Cremasteric reflex	Absent	Usually present
Scrotal lie of testicle	Cephalad/transverse	Lower in scrotum
Prehn's sign	No change in pain	Decrease in pain
Pyuria	Absent	May be present
Dysuria	Absent	May be present

2. Blood flow to the testicles can be evaluated rapidly and the testicular anatomy can be assessed. Normal or increased blood flow is seen in epididymitis, while absent blood flow is indicative of torsion. Testicular rupture as in trauma, can also be identified.

3. Cremasteric reflex: Gently stroking the medial thigh elicits spermatic cord cremasteric muscle contraction and testicular movement. Prehn's sign: elevation of the affected testicle may improve the pain in epididymitis. Blue dot sign: a torsed ischemic testicular appendage may appear as a blue dot through the scrotal skin. Bell clapper deformity: incomplete investment of the tunica vaginalis onto the testicle and epididymis, with the testicle being predisposed to rotate, and torse, more easily than if the tunica vaginalis were present.

4. Detorsion within 6 hours of the onset of the torsion.
5. Acute scrotal exploration and testicular detorsion with bilateral testicular fixation (if the testicle was detorsed and salvageable).
6. Antibiotics for acute epididymitis.

Chapter XIII.11. Ambiguous Genitalia

1. Bilateral non-palpable testes in a full term infant. Hypospadias associated with separation of the scrotal sacs. Undescended testes with hypospadias.
2. Clitoral hypertrophy. Foreshortened vagina with single opening. Inguinal hernia containing a gonad.
3. Amenorrhea, inappropriate breast development, virilization, or the onset of "cyclic hematuria".
4. Congenital adrenal hyperplasia and mixed gonadal dysgenesis.
5. Chromosomal karyotype, pelvic ultrasound, genitogram, cystovaginoscopy, gonadal inspection and biopsy, and biochemical studies as necessary (i.e., in an infant with symmetrical masculinization and non-palpable gonads, serum 17-hydroxyprogesterone, deoxycorticosterone, electrolytes, and glucose would be checked because of suspected congenital adrenal hyperplasia).
6. Fertility potential, capacity for normal sexual function, endocrine function, potential for malignant change in a gonad, and psychosexual factors.
7. Male reconstruction may require hypospadias repair, orchiopexy, and removal of inappropriate gonads and internal Mullerian structures. Female reconstruction may require a feminizing genitoplasty (clitoral reduction and vaginoplasty), as well as the removal of inappropriate gonadal tissue.

Chapter XIII.12. Hypospadias

1. 1 in 300 newborn males.
2. Nonpalpable gonads and hypospadias (especially severe proximal hypospadias) is associated with an increased risk of the presence of an intersex state (about 27%) (4).
3. Cryptorchidism and inguinal hernias.
4. A normal appearing circumcised penis with the meatus at the glans tip. The erect penis should be straight.
5. Urethral fistula, urethral stricture and recurrent penile chordee.
6. Glanular, coronal, distal penile shaft, midshaft, penoscrotal, scrotal, perineal.
7. Mild, moderate, severe.

Section XIV. Critical Care and Emergency Medicine

Chapter XIV.1. Pulmocardiac Resuscitation

- 1.e
- 2.c
- 3.e
- 4.a
- 5.d
- 6.c

Chapter XIV.2. Shock

1. c,a,b,d,f,e
2. c
3. c
4. d
5. b
6. c
7. c. This represents a case of cardiomyopathy with four classic findings of congestive heart failure. Note that the patient's condition worsened with fluid administration. Dopamine would be the first agent to try. Epinephrine may be used later in desperation since its alpha effect may have detrimental consequences on overall circulation.

Chapter XIV.3. Respiratory Failure

- 1.false
- 2.f
- 3.c
- 4.c
- 5.d
- 6.e
- 7.true
- 8.d

Chapter XIV.4. Intubation

- 1.false
- 2.a
- 3.false
- 4.true
- 5.false
- 6.b

Chapter XIV.5. Mechanical Ventilation

1. a
2. Coughing or gag intact. NPO. Minimized sedation. Adequate oxygenation on 40% FiO₂ with CPAP less than or equal to 4. Availability of personnel to reintubate if necessary. Availability of equipment to reintubate if necessary.
3. False
4. False
5. tidal volume
6. a
7. d
8. b
9. d

Chapter XIV.6. Submersion Injuries

1. c
2. False. The AAP recommends against swimming lessons below the age of 4 years.
3. b
4. d
5. d. Hyponatremia may occur in a salt water submersion victim, but it is not considered clinically important in most instances and it is not considered to be a "complication".

Chapter XIV.7. Pneumothorax and Other Air Leaks

1. False. A patient with this type of body habitus should have a work-up that includes looking for a connective tissue disorder such as Marfan's syndrome.
2. d. It is the second or third interspace in the midclavicular line or the fourth or fifth interspace in the midaxillary line.
3. Categories: Spontaneous and traumatic. Subcategories: Primary and Secondary. Classifications: Simple, Communicating, and Tension.
4. a & e. Tension pneumothorax is most likely to occur on ventilator patients and those with penetrating chest trauma. A stab wound to the lateral mid thorax is very likely to have entered the lower thorax.
5. False. Treatment depends on the classification of pneumothorax.
6. d.

Chapter XIV.8. Trauma

- 1.b
- 2.d
- 3.d
- 4.b
- 5.c
- 6.e
- 7.a
- 8.c

Chapter XIV.9. Toxicology

- 1.b
- 2.d
- 3.e
- 4.e
- 5.a
- 6.c
- 7.b

Chapter XIV.10. Acetaminophen Overdose

- 1.c
- 2.False. Acute ingestion of acetaminophen does not cause altered mental status.
- 3.d
- 4.True
- 5. 8 hours
- 6.b
- 7. d

Chapter XIV.11. Iron Overdose

- 1.False
- 2.d
- 3.b
- 4. a & d
- 5.b
- 6.False
- 7.b
- 8.a

Chapter XIV.12. Child Abuse

1. This child should be admitted to the hospital for his initial management and evaluation of potential child abuse. The hospital can offer the necessary diagnostic studies necessary to determine the presence and extent of other injuries. In addition the hospital environment offers and opportunity to observe child and family interactions by trained staff. It is the obligation of those caring for this child to insure that he be returned to a safe environment (16).

2. It is a unique form of child abuse where the child's caregiver inflicts or fabricates illness on the child.

3. When a child's weight is plotted on a growth curve and is found to be below the 5th percentile for their chronological age.

4. One of the major keys in determining the difference between accidental injuries and abusive ones is that the description of the incidents does not match the injury.

5. False. Many variables can affect the progression of a bruise. Bruises do tend to follow different stages progressing from red to green, yellow, brown and then clearing. An exact time frame cannot be established when the injury occurred, only that some bruises are older than others.

Section XV. Endocrinology

Chapter XV.1. Diabetes Mellitus

1. Infants: 100-200. Children: 80-180. Teenagers: 70-120.
2. Type 1
3. 50 %
4. Anti islet cell, anti insulin, and anti GAD antibodies
5. HgA1C is the combination of hemoglobin and glucose. It is elevated when the glucose levels are high and it is a good marker for diabetes control.
6. Postprandial.

Chapter XV.2. Thyroid Disorders

- 1.False
- 2.False
- 3.True
- 4.False
- 5.False
- 6.True

Chapter XV.3. Short Stature

1. 177.5 cm (5'10")
2. A) supine B) standing
3. No, random serum growth hormone levels are generally unhelpful in the work-up of short stature.
4. hemiskeleton
5. constitutional delay of growth and adolescence

Chapter XV.4. Adrenal Disorders

- 1.b
- 2.b
- 3.a
- 4.c
- 5.c
- 6.True
- 7.c
- 8.d
- 9.d
- 10.b

Chapter XV.5. Antidiuretic Hormone

1. The main biologic actions of ADH are to reduce the rate of urine flow by increasing the reabsorption of solute-free water from the filtrate in the distal tubules and collecting ducts of nephrons. This occurs via V2 receptors. When ADH acts on V1 receptors it causes vasoconstriction and contraction of smooth muscle elements.
2. Besides polyuria and polydipsia, physical exam and lab studies are typically within normal limits. However, in severe cases, signs and symptoms of hypernatremia and dehydration may be present.
3. Vasopressin challenge test: polyuria and polydipsia are corrected in central diabetes insipidus, but not corrected with standard doses in nephrogenic diabetes insipidus.
4. Yes and No. There are 4 types, only one is regulated by osmolality; however, the osmostat is reset to a lower osmolality.
5. Small cell lung cancer
6. The urine sodium is the test that should be done next. If the urine sodium is low, then the hyponatremia is due to total body sodium depletion. If the urine sodium is high, then the hyponatremia is due to SIADH, Addisonian crisis, diuretics, or salt losing nephropathy.
7. False. Hypertonic saline infusion is dangerous.

Chapter XV.6. Calcium Disorders

1. True.
2. False. Calcitonin lowers serum calcium levels.
3. False. Breast milk contains low levels of vitamin D. Vitamin D supplementation will prevent rickets.
4. False. A compensatory increase in PTH in response to hypocalcemia (such as with rickets) will usually result in low or normal calcium levels. High PTH levels should result in hypercalcemia; however, pseudohypoparathyroidism is an end-organ resistance to PTH, so despite an elevated PTH, patients have hypocalcemia.
5. False. The mainstay of therapy in hypophosphatemic rickets is oral phosphate replacement. Calcitriol is used to decrease the amount of phosphate needed (increases intestinal phosphate absorption), and prevent hypocalcemia and secondary hyperparathyroidism.

Section XVI. Rheumatology

Chapter XVI.1. Systemic Lupus Erythematosus

1. Malar rash, discoid rash, photosensitivity, oral ulcers, arthritis, serositis, renal disorder, neurologic disorder, hematologic disorder, immunologic disorder, antinuclear antibody
2. Corticosteroids, NSAIDs, hydroxychloroquine, cyclophosphamide, azathioprine, methotrexate, cyclosporine, and mycophenolate mofetil
- 3.c. Retinal toxicity may be a complication of hydroxychloroquine therapy. Long-termed corticosteroid therapy may be complicated by cataracts, glaucoma, and increased intra-ocular pressure. Rare cases of orbital/ocular vasculitis may occur.
4. The answer is true, depending on your interpretation. The ANA test is non-specific in that a positive ANA test by itself is not diagnostic of SLE. The ANA is frequently positive in normal individuals. However, a screening test is not a diagnostic test, but it is just used to screen. A negative ANA suggests that the patient does not have SLE. The answer to this question true, but it should be noted that the ANA is frequently ordered excessively and inappropriately.
5. False. It is true that patients with a lupus anticoagulant have a prolonged PTT. However, the lupus anticoagulant paradoxically is associated with and increased risk of thrombosis, rather than hemorrhage.

Chapter XVI.2. Juvenile Rheumatoid Arthritis

1. None of these answers are correct. None of these tests have a high positive predictive value for JRA.
2. True.
3. True. Iridocyclitis may be difficult to diagnose by a non-ophthalmologist.
4. Polyarticular, pauci-articular, systemic JRA. Refer to the chapter for how they are different.
5. NSAIDs, Cox-2 inhibitors, hydroxychloroquine, oral gold, D-penicillamine, methotrexate, sulfasalazine, glucocorticoids, TNF-alpha antagonists, IV gammaglobulin.
6. In JRA, NSAIDs inhibit an inflammatory reaction which is pathological and destructive. The inflammation which occurs in an ankle sprain is largely a repair process. The benefit of using NSAIDs to inhibit this type of inflammation is less clear.

Chapter XVI.3. Vasculitis

1. IgA.
2. purpura, arthritis, abdominal pain and glomerulonephritis.
3. leukocytoclastic vasculitis.
4. Kawasaki disease.
5. JRA, SLE, dermatomyositis, scleroderma and Behcet disease.

Section XVII. Ophthalmology

Chapter XVII.1. Neonatal Conjunctivitis and Eye Prophylaxis

1. Late presentation, presence of pseudomembranes and accompanying pneumonia.
2. The patient's mother and her sexual contacts should seek medical attention and treatment for urogenital chlamydia and other sexually transmitted diseases.
3. Chemical irritants, *Neisseria gonorrhoeae*, and *Chlamydia trachomatis* are the most common causes. However, *Staphylococcus aureus*, group A or B streptococcus, *S. pneumoniae*, *Haemophilus influenzae*, *Pseudomonas aeruginosa* and Herpes simplex virus should also be remembered as potential pathogens.
4. None.
5. Infants who develop chlamydial conjunctivitis with or without pneumonia should be treated with oral erythromycin (50mg/kg/day in 4 divided doses) for 14 days. For nondisseminated *N. gonorrhoeae* ophthalmia neonatorum, infants should receive ceftriaxone (25-50 mg/kg IV or IM) once. Alternatively, 100 mg/kg of cefotaxime (IV or IM) can also be given.
6. Without prompt treatment, *N. gonorrhoeae* ocular infection may result in corneal ulceration and perforation, iridocyclitis, anterior synechiae, and panophthalmitis leading to permanent vision loss and blindness. Left untreated, chlamydia conjunctivitis will subside within 2-3 weeks, but chronic infection is common. Chlamydia pneumonia is the most serious consequence of neonatal *C. trachomatis* infection. The pneumonia does not appear to be life threatening; however, the disease can lead to chronic cough and long-term pulmonary impairment.
7. Infantile hypertrophic pyloric stenosis.

Chapter XVII.2. Primary Care Eye Examination

1. Cataracts, retinal detachment, and other pathology that is obscuring the vitreous or aqueous clarity.
2. Retinoblastoma.
3. Performing a Cover Test and corneal light reflex test.
4. About an adult's arm length.
5. To the midline is 1 month, past the midline is 2 months, and 180 degrees is 5-6 months.
6. Two methods are to spin the child and turning his head, both of which use the vestibular systems.
7. By being patient, looking at the child's red reflex in all four quadrants in a stationary position from about 12 inches away, and as you move closer, viewing the optic disk as it passes by, and lastly the fovea by telling the child to look directly at your "magic light."

Chapter XVII.3. Strabismus and Amblyopia

1. Infantile esotropia.
2. 6 months.
3. Patching.
4. Possible answers: pseudoesotropia, accommodative esotropia, sixth nerve palsy, sensory deprivation esotropia, nystagmus-blockage syndrome, Duane syndrome, Mobius syndrome.
5. 1 year of age.
6. The child will have a permanent reduction in visual function. This can range from reduction of stereopsis to total blindness in one eye. It is possible for stereopsis to be lost even if visual acuity is preserved (i.e., measured visual acuity is 20/20) since stereopsis is dependent on vision, plus integration and processing of the images by the brain.

Chapter XVII.4. Eye Infections and Conjunctivitis

1. The answer is d. Herpes simplex conjunctivitis can present with all of the above.
2. The answer is all of the above. Although a skin laceration is easily diagnosed, a sinusitis needs to be confirmed with a CT scan. A chalazion is usually diagnosed by history or a fluctuant skin mass in the eyelid. A dental infection involving the upper teeth can easily spread itself into the orbit.
3. Topical corticosteroid is the only choice that is not appropriate for a primary care physician to prescribe. The rest of the choices are appropriate, although most chalazia do not require oral antibiotics.
4. Topical erythromycin for two weeks and oral erythromycin for two weeks for the patient AND oral erythromycin for two weeks for her sexual partner.
5. The baby is probably developing an allergic reaction to the long-term use of topical sulfacetamide. The eyedrops should be discontinued right away and patient can be treated with tear duct massage and another antibiotic eyedrop on an as-needed basis.

Chapter XVII.5. Corneal Abrasions

1. Choice d is the correct answer. A corneal abrasion which is at significant risk for infection should not be patched. Choices a, b, and c are all at higher risk for infection.
2. Choices a and b are all reasonable answers. Choice d would be too slow for an office or emergency department, but it would be reasonable if one is willing to wait for it to take effect. Choice c is incorrect because topical ophthalmic agents should not be sent home with patients. Prolonged corneal anesthetic use often results in corneal complications because this blocks the eye's natural protection reflexes to minimize further corneal injury.
3. The differential diagnosis consists of corneal foreign body, conjunctival foreign body, early conjunctivitis. The eyelids should be flipped to look for small foreign bodies. If possible, the cornea should be inspected again with some magnifying glasses to look for a foreign body as well.
4. Whenever the cornea has white lesions, one should always suspect corneal ulcers or infiltrates. Overnight contact lens wear is the most significant contributor to the development of corneal ulcers in a contact lens wearer. The patient should be referred to an ophthalmologist as soon as possible and the patient should be advised to discontinue contact lens wear until treatment is completed.
5. The patient should have an ophthalmology consult as soon as possible. A metal shield should be placed on the eye, NOT a gauze eye patch (which can press on the eyeball), to decrease further chance of injuring the eye. He probably should be admitted to the hospital for bedrest and observation to decrease the chance of re-bleed.

Section XVIII. Neurology

Chapter XVIII.1. Neurologic Examination

1. Examination of the skull, cranial nerves, strength, cerebellar function, sensory, and reflexes.
2. Ventral suspension, horizontal suspension (Landau reflex), Moro reflex, tonic neck response (fencer's stance), palmar and plantar grasp reflexes, parachute response, reflex placing and stepping responses.
3. Lateral rectus and superior oblique muscles, respectively.
4. Signifies that cortical vision is intact, in addition to showing the integrity of the frontal and parietal lobes, and visual fields. It can be performed at about 4 to 6 months of age.
5. When the arms are lifted, a positive sign is when an arm is hyperpronated with the elbow flexed. It tests for strength of the upper extremities, and a positive sign signifies weakness.
6. In what two instances can a positive Babinski's sign be seen in normal patients? In newborns up to 2-1/2 years of age and sometimes in patients just after a febrile seizure.

Chapter XVIII.2. Cerebral Palsy

- 1.b
- 2.a
- 3.a
- 4.d
- 5.false
- 6.true

Chapter XVIII.3. Febrile Seizures

1. 6 months to 5 years. It occurs in 2-5% of all children and is the most common reason for convulsions in children less than 5 years of age.
2. 33%.
3. Simple seizures are characterized by being less than 15 minutes duration and generalized. Complex febrile seizures are greater than 15 minutes duration, multiple within 24 hours, and focal. Simple febrile seizures have a higher risk for febrile seizures. Complex febrile seizures have a higher risk for epilepsy. One should have a lower threshold for performing tests and hospitalization in cases of complex febrile seizures.
4. Meningitis, encephalitis, Shigella gastroenteritis, medications and toxins, hypoglycemia, electrolyte abnormalities, shaken baby syndrome, accidental head trauma, and epilepsy.
5. Infants less than 12 months of age.
6. Unstable clinical situation, possibility for meningitis, and parents unreliable or unable to cope with the child developing another seizure.

7. Disadvantages include lethargy, drowsiness, ataxia, and masking of a CNS infection.
8. 1) Seizure will not cause brain damage and the risk of a child developing epilepsy is small. 2) Possibility that it can happen again, especially in the first 24 hours. One third of children will have at least another febrile seizure with most occurring within one year of the episode. 3) If seizure occurs again, child should be kept on his or her side. If seizure does not stop within 3 minutes, then emergency medical services should be contacted.

Chapter XVIII.4. Epilepsy

1. Partial simple (also called "partial elementary" or "focal motor"), partial complex, generalized tonic-clonic, generalized absence.
2. Focal motor seizures (partial simple) because only one part of the body exhibits tonic clonic seizures. "Jacksonian seizures" describe focal motor seizures, while "Jacksonian march" describes a partial simple with secondary generalization because of gradual spread of motor activity. Temporal lobe epilepsy (partial complex) due to lesions in the temporal lobe. Psychomotor seizures (partial complex) because they display behavioral changes in addition to focal motor abnormalities, such as twitching and grimacing. Grand mal (generalized tonic-clonic) because they exhibit grand abnormalities as manifested by generalized jerking. Petit mal (generalized absence) because they exhibit smaller abnormalities limited to the eyes and face in most instances, and also because these patients are generally in elementary school and thus petit in size.
3. Partial complex seizures. She has experienced an aura (burning rubber smell). The witnesses suggest mostly facial motor symptoms. She lost consciousness. The temporary expressive aphasia suggests a temporal lobe origin which is confirmed on CT scan which identifies a lesion in the left temporal lobe (which is why this used to be called temporal lobe seizures). Students will often confuse this presentation with generalized absence seizures, which usually occurs in elementary school aged children who have just a few seconds of impaired/loss of consciousness. This is not a partial simple seizure because there are motor, aura, aphasia and olfactory symptoms, in addition to loss of consciousness.
4. No, petit mal refers to generalized absence seizures. Jerking of one arm (even if they are small jerks) are partial simple seizures (focal motor), not generalized absence (petit mal).
5. Electrolytes, glucose, toxicology, AED levels, CT of the head, lumbar puncture would be a basic set of initial tests. An eventual EEG would be in order if no obvious precipitating factors were found.
6. The MRI has better resolution for smaller and isodense lesions (e.g., low-grade gliomas). MRI may also provide better images in some areas (e.g., posterior fossa) and for some lesions types (e.g., neuronal migration disorders, lesions of the neurocutaneous syndromes, AVM).
7. No, a negative EEG does not rule out epilepsy. False negative results can be associated with epileptogenic foci that are deep to the cerebral surface, discharges that are orthogonal to the cerebral surface, excessive muscular artifact, and simply due to limitations in monitoring duration in comparison with the frequency of epileptiform EEG activity. Of all the seizure types, partial complex seizure foci are the most difficult to reliably identify on EEG.
8. Generalized absence seizures typically have a generalized 3 per second (Hertz) spike and slow wave EEG pattern, often provoked by hyperventilation. Infantile spasms have a hypsarrhythmia pattern on EEG which has an asymmetric disorganized mixture of spikes and slow waves.
9. Ethosuximide and valproic acid are used to treat generalized absence seizures.
10. 60-70% of children with epilepsy eventually have good seizure control on AEDs and enter into long-term remission, but 30% will never become seizure free on AEDs.

Chapter XVIII.5. Status Epilepticus

1. Benzodiazepines.
2. Phenytoin (or fosphenytoin), phenobarbital, valproic acid.
3. IV and rectal.
4. Lorazepam.
5. Phenytoin (or fosphenytoin).
6. Hypoxic ischemic brain injury, death.
7. Epilepsy, encephalitis, neoplasm, drug overdose, metabolic derangement, cerebrovascular accident, trauma, etc.

Chapter XVIII.6. Infant Botulism

1. It is recommended to not give honey to any infant under 12 months of age.
2. Botulinum toxin is released by bacteria within the infant's GI tract. From here, the toxin is absorbed and carried by the blood stream to peripheral cholinergic receptors where it binds irreversibly. Clinically, the most important of the peripheral cholinergic receptors is the neuromuscular junction. Here the toxin's action results in flaccid paralysis and hypotonia, which are the classic clinical signs of infant botulism.
3. Initially, infected infants often present with a history of poor feeding, decreased activity and constipation. The diagnosis may not be considered initially because signs of an evolving bulbar palsy, flaccid paralysis and hypotonia may be subtle. Additionally, the infant may be worked up for sepsis if he appears toxic or "lethargic", or for constipation until the "classic" manifestations of infant botulism become apparent. The classic age distribution for infant botulism is 3 weeks to 6 months of age.
4. Isolation of the clostridium botulinum organism in stool can be accomplished in the early stages of disease, it is rarely isolated in blood. The most common method for proving infection is to isolate botulinum toxin in blood or stool samples. Toxin can be detected in the stool of infected infants for up to 4 months. Electrophysiological testing, specifically electromyography, can aid in ruling out other neurologic disorders such as Guillain-Barre syndrome, congenital myopathies, and myasthenic conditions.
5. The use of antibiotics in infant botulism should be reserved only for proven secondary infections such as pneumonia, otitis media or urinary tract infections. Aminoglycosides should be avoided as they are weak pharmacologic neuromuscular blocking agents which may potentiate paralysis acutely or cause respiratory failure in an unsuspected infant with botulism being treated for sepsis.
6. Human botulinum immunoglobulin (BIG), which acts by interrupting the blockade of nerve receptors by botulinum toxin, has been shown to reduce the need for mechanical ventilatory support and shorten overall duration of hospitalization.
7. If recognized early and given appropriate supportive care minimizing complications, full recovery and a normal neurologic function can be expected.
8. Classic "botulism" is a food borne disease in which high levels of toxin can be ingested in spoiled food. It often occurs in outbreaks linked to a particular source, and typically afflicts older children and adults. Wound botulism is rare, but is seen disproportionately in adolescents and children. Infant botulism has a more gradual onset. All types of botulism produce disease through a similar pathogenesis.

Chapter XVIII.7. Guillain-Barre Syndrome

1. Campylobacter jejuni enteritis.
2. Lack of cellular response (normal WBC count) in the CSF despite an elevated protein level. In the clinical setting of progressive flaccid paralysis, this is diagnostic of Guillain-Barre syndrome.
3. False. Improvement in strength occurs in reverse order (bulbar muscle strength returns first and lower extremity strength returns last).
4. IVIG does not require central venous access and does not decrease blood volume.
5. A child should be intubated if she/he has a rapidly decreasing vital capacity, dyspnea, fatigue, or deterioration of arterial blood gases. Dysphagia, shoulder weakness, and cardiovascular instability are also indications that mechanical ventilation may be necessary.

Chapter XVIII.8. Multiple Sclerosis

1. True
2. d
3. True
4. False
5. True

Chapter XVIII.9. Hydrocephalus

1. Hydrocephalus refers to pathological enlargement of the cerebral ventricles secondary to a mismatch between the amount of production of CSF and its drainage. Macrocephaly is a general term for any head circumference greater than two standard deviations from the mean. Megalencephaly refers to increased volume of the brain parenchyma.

2. Hydrocephalus is divided into two types: communicating and non-communicating. Communicating hydrocephalus is used if CSF flows freely throughout the ventricular system. Non-communicating hydrocephalus indicates that obstruction of CSF occurs somewhere within the ventricular system, including the outlet foramina of Luschka and Magendie. Communicating hydrocephalus may occur from scarring of the leptomeninges after viral or bacterial meningitis, or after a hemorrhagic brain event where the breakdown products of blood lead to diffuse fibrosis of the meninges. Non-communicating hydrocephalus occurs in cases of discrete obstruction within the ventricular system, such as occurs with aqueductal stenosis, the Chiari Malformations, the Dandy-Walker malformation, or mass effect from brain tumors or other mass lesions.

3. The most common causes of congenital hydrocephalus are Chiari malformations and aqueductal stenosis.

4. X-linked hydrocephalus is a form of aqueductal stenosis in which there is a mutation on the X-linked recessive L1 gene, which produces a family of abnormal neuronal cell adhesion molecules that leads to narrowing and obstruction at the level of the cerebral aqueduct.

5. False. The Dandy-Walker malformation, although present at birth, is responsible for less than 5% of cases of congenital hydrocephalus. Approximately 80% of cases will eventually be detected by one year of age.

6. When present, intraventricular bleeding in the very low birth weight infant usually occurs within the first 72 hours of life. Because up to 50% of these events will occur without immediate clinical symptomatology, it is recommended that routine screening be performed between days 4 to 7 of life.

7. False. MRI is the preferred imaging method for the diagnosis of hydrocephalus after the neonatal period as it will also elucidate more precisely than CT the specific etiology of the hydrocephalus. However, in an emergency situation, CT is preferred because it can be done rapidly.

8. Shunt malfunction is a fairly common occurrence with a one-year failure rate of 30 to 40%.

9. The rate of infection after shunt insertion varies among different institutions, and has been reported from 1 to 10%. The most likely etiologic agent is *Staphylococcus epidermidis*.

10. False. The overall outcome and prognosis of hydrocephalus is highly dependent on multiple factors, including age of onset, etiology, the rate of ventricular expansion, and the extent of neurologic damage prior to shunt placement or other corrective intervention. In one study that looked at 129 children 10 years after shunt placement (shunts were placed prior to age two), approximately 60% had IQs over 70, which is generally considered the cutoff for one of the diagnostic criteria for mental retardation.

Chapter XVIII.10. Neural Tube Defects

1. True. Folate supplementation prior to pregnancy and in early pregnancy reduces the risk of neural tube defects.

2. True, in that nearly all patients with myelodysplasia have bladder/bowel dysfunction; however, patients with spinal bifida occulta may only have a vertebral anomaly, without myelodysplasia, in which case, their bladder function will be normal.

3. Controversial question, but probably false. The hydrocephalus is usually due to a Arnold Chiari malformation in the brain (the other end of the neural tube) which results in hydrocephalus. It is probably not cord tethering which causes the hydrocephalus.

4. True.

5. True.

Chapter XVIII.11. Neurofibromatosis

1. 6 spots. >5 mm in prepubertal and >15 mm in postpubertal patients.

2. Vestibular schwannoma (acoustic neuroma). It is manifested by hearing problems.

3. Blood pressure measurements and eye examinations.

4. Deep nodular neurofibromas and diffuse plexiform neurofibromas.

5. Cafe au lait spots in newborn period. Discrete cutaneous and subcutaneous neurofibromas, axillary freckling in the older patient.

6. Autosomal dominant. 50% occur without a family history.

7. Lisch nodules.

Chapter XVIII.12. Tuberous Sclerosis Complex

1. Autosomal dominant.
2. 50%.
3. Ash leaf spots (birth), adenoma sebaceum or facial angiofibroma (5 years old), shagreen patch (after 10 years old).
4. ACTH.
5. Hypsarrhythmia.
6. Echocardiogram and renal ultrasound.

Chapter XVIII.13. Head Trauma and Hemorrhage

1. False. Epidural hematomas are a neurosurgical emergency and have a lenticular (lens or football shaped, also called biconvex) shape on CT scan.
2. False. Only 20% of epidural hematomas are produced by venous blood in children.
3. False. Acute subdural hematoma is associated with substantial brain parenchymal injury so its prognosis is poor compared to epidural hematoma.
4. False. Epidural hematoma is a neurosurgical emergency because its prognosis is dramatically better with early evacuation, while subdural hematoma is less of an emergency because the prognosis is already poor even with hematoma evacuation.
5. False. Infants are at higher risk for sustaining serious head injury. Anatomical considerations that predispose the younger child to head injuries are a large head to body ratio, a relatively weak neck, a thinner skull, and a larger subarachnoid space in which the brain can move freely.
6. True.
7. True.
8. False. Hyponatremia occurs with SIADH. Free-water is retained in the collecting tubules due to anti-diuretic hormone causing a dilutional effect of the serum sodium. Hypernatremia is usually caused by the use of hyperosmotic agents such as mannitol or diabetes insipidus.
9. False. In well appearing children 2-18 years of age with no loss of consciousness and a normal neurological exam, no imaging studies are required. Close observation and parental education is all that is needed (6).
10. True. GCS=9: motor=4, verbal=3, and eye opening=2.

Chapter XVIII.14. Muscular Dystrophy

1. Both Duchenne and Becker muscular dystrophy are X-linked recessive.
2. Dystrophin.
3. The Gowers' maneuver is seen when a child climbs up on his thighs with his hands when going from a sitting to standing position. This is due to weakness in the knee and hip extensors.
4. Delay in walking, waddling gait, walking unsteadily with frequent falling, walking on toes, and difficulty at climbing stairs, Gowers' maneuver, and pseudohypertrophy of the calves.
5. 20 to 30 years. They die from pulmonary or cardiac problems. They lose ambulation before 13 years old.
6. 5 years.
7. Pulmonary, cardiac, and neurological (CNS).
8. Corticosteroids.

Chapter XVIII.15. Myopathy and Myositis

1. True. Sun exposure can exacerbate JDM dermatologic lesions and myositis, even during therapy. It is important to advise patients with JDM about appropriate sun protection - large rimmed hat, clothing over majority of body in sun, sunblock with a SPF of 15 or higher.
2. Unknown, but thought to be low. The risk of recurrence of JDM is highest in the first year after diagnosis, therefore maintenance prednisone is usually continued for 2 years.
3. c.
4. a
5. a, b and f are correct. Pain is typically greater in viral myositis than in JDM. Acute viral myositis has a propensity to affect the calf region, rather than the biceps. Both viral myositis and JDM have elevated CPK and ESR so these tests do not distinguish the two.

Chapter XVIII.16. Developmental Brain Anomalies

1. Type II (Arnold-Chiari malformation).
2. Cerebellum.
3. Lissencephaly/pachygyria, polymicrogyria, heterotopia.
4. Smooth surface of the cerebral cortex.
5. Myelomeningocele and hydrocephalus.

Chapter XVIII.17. Reye Syndrome

1. True
2. c
3. b
4. c
5. d
6. a

Chapter XVIII.18. Brain Tumors

1. e
2. b
3. f
4. d
5. e

Chapter XVIII.19. Arteriovenous Malformations

1. True
2. D
3. True
4. True
5. True
6. False

Section XIX. Orthopedics

Chapter XIX.1. Fractures

1. Fractures in children heal more rapidly than those in adults because the pediatric bone has a thicker periosteum and more efficient remodeling.
2. A fracture is described by its anatomic location, configuration, relationship of the fracture fragments to each other, and relationship of the fracture fragments to the surrounding tissue. Physeal fractures can be described according to the Salter-Harris system.
3. External fixation refers to fixation of bones by splints, casts or transfixion pins. A cast is sometimes considered merely external support, rather than external fixation. Internal (or intraosseous) fixation is stabilization of the bone fragments by direct fixation to one another with surgical wires, screws, pins, rods, or plates.
4. The clavicle is the most frequently fractured bone in the pediatric population.
5. A toddler's fracture is a subtle non-displaced spiral fracture resulting from a rotational injury while running or playing.
6. A toddler's fracture is subtle and non-displaced, while a large distal tibia fracture is more likely to be associated with severe trauma (not just falling while walking) or child abuse.
7. Radial head dislocation (the Monteggia injury).
8. a) Non-displaced Salter-Harris type I fracture of the distal radius, b) scaphoid fracture, c) radial head fracture, d) Non-displaced Salter-Harris type I fracture of the distal fibula (lateral malleolus).

Chapter XIX.2. Splinting

1. Splints are generally used to temporarily immobilize fractures, subluxations, or soft tissue injuries such as ankle sprains.
2. Splints immobilize the extremity, reducing damage to the nerves, vasculature, muscle, and skin. This will minimize edema and pain. Splints also stabilize fractures and prevent further displacement of subluxations.
3. If the splint is too tight it will compress the swollen extremity causing decreased sensation, paresthesia, and pain. The patient should be educated to check for brisk capillary refill, mobility of distal anatomy, numbness, tingling, burning, and increased pain. The immobility of the joint may cause contractures. Mobility of the distal anatomy should be evaluated. Stiffness of the immobilized joint should be expected. Wrinkles in the splinting material may cause pressure sores and skin breakdown, especially over bony prominences. Skin breakdown often starts with burning or itching, and may progress to ulceration.
4. Conservative treatment involves splinting of the extremity. The general rule is, when in doubt, splint. Splinting is indicated with sprains overlaying an open physis, because of the similar presentation to a Salter-Harris type 1 fracture. However, many sprain injuries (ankle sprain is the best studied example), will improve faster with gentle activity compared to total rest or immobilization.
5. Plaster is inexpensive and it allows for anatomic molding. However, it is relatively heavy and it can take longer to set and cure. Fiberglass is a more expensive, prepackaged, strong and light splint that cures quickly, but does not allow exact anatomic molding. For example, for an ankle fracture, plaster splinting results in a heavy splint, compared to a fiberglass splint which is stronger and lighter.
6. Complicated fractures include open fractures, fractures with any neurovascular compromise, fractures that are too deformed/angulated/displaced to adequately splint, and any dislocation which cannot be reduced in the ED.
7. The strip should be approximately 50% of the circumference of the extremity.
8. Cold water slows the curing process in both plaster and fiberglass, but ROOM TEMPERATURE water rather than cold water should be used. Warm water is best avoided since it will add further heat to the exothermic reaction.
9. Inspection. Wounds must be cleaned and dressed. Neurovascular compromise should be ruled out, and documented.
10. Casting forms a rigid cylinder over the extremity. In the first 24 hours following a fracture, swelling within the cylinder may result in vascular compromise (i.e., compartment syndrome). Splinting initially, then casting later is associated with fewer complications compared to early casting. Additionally, if the extremity is already swollen and a cast is applied, the fit of the cast will be loose once the swelling resolves. Casts are generally applied by orthopedic surgeons who are not always available for minor fractures. Splints provide an immediate means of immobilizing the extremity and do not require the immediate presence of an orthopedic surgeon.

Chapter XIX.3. Scoliosis

1. Clinical - side-to-side (sagittal) curvature of the spine. Radiographic - curvature of the spine whose curvature is greater than or equal to 10 degrees.
2. Females are affected more commonly than males.
3. Congenital, neuromuscular, traumatic, infectious, neoplastic, inflammatory, syndromic and degenerative causes.
4. Side-to-side curvature of the spine, rib hump, shoulder elevation, chest wall deformity, prominence of the scapula on one side.
5. Asymmetry of the rib hump
6. Risk of progression - skeletal maturity and magnitude of curvature.
7. Observation, brace, and surgery
8. Curvature less than 30 degrees - asymptomatic, non-progressive. Curvature greater than 50 degrees - progression in adulthood (1-2 degrees/year).

Chapter XIX.4. Osteomyelitis

1. False. *S. aureus* is the most common. Group A strep is the second most common.
2. True
3. False. Typically the course is for 6-8 weeks, always starting with IV antibiotics and finishing with PO antibiotics if possible.
4. True
5. False. Plain films usually begin to show acute changes 5-7 days into the course of the disease process.
6. False. The femur is the most commonly involved bone. The tibia is the second most commonly involved.
7. False. The metaphysis is the most common site.
8. False. The rate of methicillin resistant *S. aureus* is too high to use oxacillin/methicillin as empiric therapy. Vancomycin should be initially started.

Chapter XIX.5. Septic Arthritis

1. False. It is a condition that usually affects younger children early in the first decade of life.
2. True
3. False. The hip joint is deep and has a significant amount of surrounding tissue, thus inflammation may not be easily detected on physical exam. Exam findings may be subtle, such as asymmetry or loss of function. Decreased and painful range of motion is the best way to detect an effusion by physical exam.
4. False. They also can present with fever. This is why differentiating between toxic synovitis and septic arthritis can be a difficult clinical problem.
5. No definite answer here. Low ESR and CRP values make septic arthritis unlikely. Very high ESR and CRP values make septic arthritis more likely. Intermediate ESR and CRP values are not very helpful in distinguishing toxic synovitis from early septic arthritis.
6. True
7. True
8. False. In larger joints surgical intervention is almost always performed. However in cases of septic arthritis of smaller joints, medical management can be carried out with good results. Orthopedic surgical consult should always be obtained expeditiously whenever the diagnosis is considered.

Chapter XIX.6. Hip Conditions

1. Is it painful? How old is the child? What is the duration of symptoms? These three questions will help to narrow the differential diagnostic possibilities.
2. Toddler: transient synovitis, infection, trauma. Juvenile: trauma, transient synovitis/infection, Legg-Calve-Perthes disease. Adolescent: slipped epiphysis, trauma, neoplasia
3. Hip instability - Ortolani sign. Asymmetric gluteal skin folds.
4. Leg length discrepancy, Galeazzi sign (apparent thigh length difference), waddling gait.
5. Concentric reduction of femoral head, stability, remodeling and growth of acetabular cartilage.
6. Although the etiology is unknown (commonly stated as idiopathic), most current theories involve vascular compromise of the femoral epiphysis. Two episodes of infarction are thought necessary to cause the changes consistent with LCP disease in humans. Increased blood viscosity, thrombophilia, and intraosseous venous hypertension have been proposed as mechanisms for vascular compromise.
7. 1. Initial, 2. Fragmentation, 3. Reossification, 4. Residual.
8. Age of patient at onset and proportion of femoral head involvement. Children who have LCP disease before age eight have a better prognosis over children greater than eight years of age at time of onset. The proportion of head involvement forms the foundation for several classification schemes. Maintenance of the height of the lateral column of the femoral epiphysis appears to have the most prognostic significance in children in any age group. Whole head involvement or collapse of the lateral column by more than fifty percent carries a poor prognosis.
9. Obese, delayed maturation of skeletal age.
10. Trendelenburg gait or antalgic limp, obligate external rotation of the hip with flexion, limited internal rotation of the hip.
11. Good to excellent. High activity level. Slow degenerative process of the hip with few cases requiring prosthetic hip replacement.

Chapter XIX.7. Common Sprains and Dislocations

1. Combination of plantar flexion and inversion.
2. Anterior talofibular ligament.
3. Rest, Ice, Compression, Elevation.
4. Traction injury resulting from being lifted or pulled by the hand or arm.
5. Supination or hyperpronation of the forearm at the elbow.
6. Anterior drawer test assesses ACL laxity. Physician should assess for other structural abnormalities in the affected knee, as multiple ligaments and/or menisci may be injured.
7. Axillary nerve injury. Injury to the axillary nerve can result in transient loss of sensation, tingling and numbness to the lateral aspect of the deltoid.
8. Stimson technique or external rotation method. See text.

Chapter XIX.8. Sports Injuries

1. Traction apophysitis of the tibial tuberosity.
2. Osgood-Schlatter: Usually older children or adolescents with male: female of 3:2. Those who do forceful contraction of the quadriceps (jumping sports such as basketball and volleyball). Sever's: Athletes who play with cleats (excess grip in the ground) who push hard while running; also soccer and basketball players.
3. It is more common on the left, however, about 25% have it bilaterally.
4. The term "little league elbow" is used to describe a group of pathologic entities in and around the elbow joint in young throwers secondary to overhead throwing. Valgus stress results in lateral compression and medial traction on the elbow leading to the many types of injuries described in the text.
5. Tennis serving, football quarterbacks, javelin throwers, volleyball spikers.
6. A direct blow to the bony rim causing enough of an increase in intraorbital pressures to fracture the thin interior bones (usually the orbital floor).
7. CT scan, with special coronal views.
8. Topical beta blockers, cycloplegics, osmotic diuretics, carbonic anhydrase inhibitors.
9. The most severe complication is a rebleed. Limiting physical activity in children within the first 72 hours is important. This includes bedrest, no television or videogames, and bilateral eye patching.

Section XX. Adolescent Medicine

Chapter XX.1. Puberty

1. Enlargement of the testes measuring greater than 2.5 cm in length and scrotal changes are the first signs of puberty in the male. The appearance of breast buds in the female indicate the onset of puberty.
2. There is approximately 6 months difference in the age of onset of sexual maturation in the female vs. the male.
3. Puberty is delayed when there is no sign of pubertal development by age 13 years in girls and 14 years in boys. Precocious puberty is secondary sexual development occurring before age 9 years in boys or 8 years in girls.
4. Height age is the age at which the height of the individual is equal to the height of 50 percent of a reference standard population by age and gender. This is done by taking the patient's height and finding the age, at which this height is the 50th percentile on an appropriate height grid.
5. The best indicator of the biological age of the individual is the skeletal age (bone age).

Chapter XX.2. Anabolic Steroids

1. True. One of the reasons it is difficult to dissuade competitive athletes from using anabolic steroids is that it can, in fact, result in increased lean body mass, muscle strength, and aggressiveness. These may, in fact, contribute to enhanced athletic performance.
2. Anabolic steroids may be taken orally or injected intramuscularly. Oral steroids are more hepatotoxic.
3. An adolescent in early puberty who uses steroids risks premature epiphyseal closure with resultant shorter stature than otherwise would be predicted.
4. Anabolic steroid use should be considered and addressed with all adolescent patients, male or female, athlete or non-athlete. Particular attention should be paid to those adolescents who have greater than expected muscle-mass development or in females with signs of masculinization.

5. On an individual level, pediatricians should, without lecturing, initiate an honest discussion of the risks and benefits of steroid use. They should ask all adolescents, and especially those with signs and symptoms of steroid use, about the possibility of using steroids. They also have a role in educating parents, teachers and coaches about the prevalence and dangers of anabolic steroid use.

Chapter XX.3. Substance Abuse

- 1.c
- 2.False
- 3.True
- 4.True
- 5.a-v; b-iv; c-i; d-ii; e-vi; f-iii

Chapter XX.4. Adolescent Suicide and Violence

1. True
2. False
3. True
4. False. Compared with adults, children and adolescents presenting with a major depressive episode are at relatively higher risk of actually having a bipolar disorder. Significant caution must therefore be exercised in prescribing an antidepressant, which may precipitate mania or hypomania. The author advises that child and adolescent psychiatric consultation be sought.
5. True.
6. False. Often, working with the family is a key component of treatment.
7. False. Currently, there are only 6300 child and adolescent psychiatrists in the United States, where the estimated need is for up to 30,000. The population of children is expected to grow 40% in the next 50 years. Pediatricians will likely play a very significant role in insuring the psychosocial health of children.

Chapter XX.5. Eating Disorders

1. Suicide is the leading cause of death in anorexia nervosa. The second highest cause of death is cardiac arrest.
2. Patients who self induce vomiting are most likely to develop a hypochloremic hypokalemic metabolic alkalosis.
3. Three indications for hospitalization of a patient with anorexia nervosa include: a) electrolyte abnormalities (hypokalemia, hyponatremia), b) cardiovascular abnormality (bradycardia, arrhythmia, hypotension), c) inability or refusal to engage in outpatient treatment.
4. The most likely diagnosis is anorexia nervosa. The point is that the most likely cause of significant weight loss in an adolescent female is an eating disorder, even if DSM-IV criteria are not completely met.
5. Disorders other than anorexia nervosa in the differential diagnosis of excessive weight loss in an adolescent include malignancy, diabetes mellitus, hyperthyroidism, malabsorption syndromes, systemic lupus erythematosus, inflammatory bowel disease, depression and substance use.
6. Bulimia nervosa is more likely to present with a normal physical exam. By definition, anorexia nervosa must show weight loss or a failure to gain weight appropriately during puberty.

Chapter XX.6. Adolescent Sexuality

1. False. The incidence of adolescent sexual activity, at least among in-school youth, appears to be declining. In addition, sexually active adolescents report fewer sexual partners and are more likely to use condoms than teenagers in the early 1990s.
2. a. Same-sex attraction is considered a normal part of adolescent and adult sexual experience. It may or may not reflect a bisexual or homosexual orientation, either of which, like heterosexuality, is believed to be established in early childhood and represents a normal developmental outcome.
3. True. The onset of sexual activity in younger adolescents is more likely to be associated with a history of negative life experiences and high-risk behaviors such as sexual abuse, substance use, parent-teen conflict and school problems. In older adolescents, the onset of sexual activity is often a more normative process.
4. True. Pediatrics as a discipline recognizes that sexual experimentation, with oneself and others, is a normal part of adolescent development. More controversial are the issues of age of initiation of sexual activity and the nature of those activities. There is a wide spectrum of viewpoints within pediatrics, reflecting broader societal views, on these latter issues.

5. b. Sexual coercion is a form of violence and, therefore, pathologic. Masturbation, homosexual orientation, and sexual fantasies and experimentation are considered a part of the spectrum of normal adolescent sexual development.

Chapter XX.7. Adolescent Gynecology

1. In Hawaii, a minor who is at least 14 years of age may consent to receive contraceptive services, prenatal care, and STD/HIV/AIDS services. The physician may notify parents (with the consent of the patient), but parental consent or notification is not required. In fact, if an adolescent demands confidentiality, it becomes a difficult situation since it might not be permissible for the physician to release information, even to parents. The wording of the statute is, "left up to the treating physician's discretion in consultation with the minor who received medical treatment", but the statute later states that the minor, "shall have the same legal capacity to act" as an adult, making their demand for confidentiality no different than that of an adult. Most insurance companies provide itemized claim information to the subscriber of the insurance policy (usually the parent). It is not possible to circumvent this in most instances. Thus, adolescents should be counseled that once they have used their parent's medical insurance, their parents will receive such information. They must consent to this release of information, or they must remove the medical insurance information so that an insurance claim is not submitted. They should also understand that they will receive a bill for all medical services, although their ability to pay it should not impede the delivery of medical services. In most instances, it may be appropriate to counsel the adolescent to share this information with their parents, and in many instances, they will consent once they understand all the issues. This requires provision of factual information to the adolescent and patience.

2. 10 to 16 year old, average 12.7.

3. NSAIDs are the treatment of choice in adolescents. Oral contraceptives may also be used.

4. Adolescents should be provided with information about their diagnosis, contraception, breast self-exam, STDs and AIDS. Instruction should be provided on how to track menses. Condom use should be encouraged in those who are sexually active.

5. 21-35 days between menses, 20-60 mL blood loss (avg 35 mL), 3-7 days of menstruation.

6. Irregular bleeding.

7. PCR or LCR (DNA methods) for chlamydia and gonorrhea assayed from a urine sample or vaginal fluid sample.

Section XXI. Skin

Chapter XXI.1. Eczematous Dermatitis (Atopic Dermatitis and Seborrhea)

1.False

2.True

3.True

4.c

5.a

Chapter XXI.2. Acne

1.c

2.a

3.False

4.False

5.True

Chapter XXI.3. Hemangiomas, Vascular Malformations and Nevi

1.False

2.True

3.b

4.False

5.False

6.True

Chapter XXI.4. Burns

1. Antibiotic ointments such as silver sulfadiazine and bacitracin are indicated for all burns except superficial burns.
2. Infants 6 months old or younger are more prone to fluid overload because of their reduced glomerular filtration rates. Additionally, they are more susceptible to hypothermia because they are unable to generate heat by shivering.
3. A patient should be sent to a burn unit if they have serious burns that are beyond the scope of care in the local institution. Examples of this include, second degree burns of 20% TBSA, third-degree burns of 5% TBSA, major burns to the hands, face, feet, perineum, or electrical burns.
4. % TBSA can be estimated by using the rule of nines, the Lund and Browder chart, or by designating the child's palm as 1% of the TBSA. The most accurate method is the Lund and Browder chart.
5. The Parkland formula is used to estimate the amount of fluid appropriate for administration in the first 24 hours.
6. The slow urine output indicates hypovolemia. The fluid infusion rate should be increased to improve the urine output.

Chapter XXI.5. Bites and Stings

1. b
2. d
3. True, anaphylaxis can occur from any repeated insect bite or sting in which re-exposure to an antigen occurs.
4. false
5. Southern black widow and Brown violin spider.
6. true
7. All except b.

Chapter XXI.6. Common Skin Conditions

1. The three layers of skin are the epidermis, dermis, and subcutaneous tissue. The skin serves as a barrier against the environment, protection against desiccation, and plays a role in immune surveillance.
2. Human papilloma virus is organism responsible for the development of warts.
3. Staph aureus is responsible for most infections in acute paronychia.
4. Pediculosis is treated with a shampoo such as 0.5% malathion rinse, permethrin 1% creme rinse, 1% lindane shampoo, or pyrethrin. After the shampoo is rinsed, the hair is combed with a fine toothed comb to remove dead nits. Clothes and bedding must be washed in hot water.
5. Varicella immunoglobulin should be given to immunocompromised individuals, neonates whose mothers develop chickenpox within five days prior to or two days following delivery, and premature neonates born less than 30 weeks gestation who have been exposed to chickenpox.
6. Suspicion of malignant transformation of nevi should arise upon observation of irregular borders, variegated color (multiple colors), size greater than 5-15 mm, and any change in texture including crusting, ulceration, or induration.

Section XXII. Reviewing the Medical Literature

Chapter XXII.1. Statistics

1. Descriptive statistics are the rates of bicycle helmet use in the injured group and in the control group. The proper inferential statistical test to use is a chi-square test.

2. The rate of bicycle helmet use in the injured group is significantly different from that in the control group. It might be tempting to say that bicycle helmets prevent significant head injuries from this study, but such a study is not good enough to conclude this.

3. Bicycle helmet use rates in the two groups are the same.

4a. Categorical.

4b. Continuous.

4c. Continuous.

4d. Continuous.

4e. Categorical.

4f. This could be both depending on what we mean by this. This would be a continuous variable if we are referring to the CSF WBC count, the RBC count, the CSF glucose, or the CSF protein. This would be a categorical variable if we are considering the CSF to be normal or abnormal, or if we are considering the gram stain result (organisms versus no organisms).

4g. Categorical. The patient either has it or they don't.

5. The basic descriptive statistic is the mean oxygen saturations in each group. Other commonly cited descriptive statistics are the standard deviations and the ranges for each group, which would describe the spread of the data. The inferential statistical test would be a T-test or ANOVA.

6. This difference is statistically significant, but it is not very clinically important because the difference between 95.6% and 94.5% is only about 1%. Continuous pulse oximetry readings will frequently fluctuate by 2 to 4 percentage points on the same patient without any clinical changes occurring.

7. The oxygen saturation (like most biomedical measurements) is not normally distributed. Most biomedical measurements have a theoretical limit on their values. Oxygen saturation values cannot exceed 100%. Thus, if one creates a distribution of oxygen saturation measurements, it will show a few points below 80%, a few more points between 80% and 90%, a fair number of points between 90% and 95%, a large number of points between 95% and 100%, and no points about 100%. This is not bell shaped. Other examples of theoretical limits are: glucose values cannot go below zero, respiratory rates will not go below 10, etc.

8a. These groups are not significantly different. The mean plus or minus two standard deviations should contain approximately 95% of the area under the bell shaped curve. Thus, the shapes of these curves are wide with substantial overlap. It is not likely that these groups will be shown to be significantly different.

8b. These standard deviations are small, so the bell shaped curves are very narrow and they do not overlap each other. Thus, it is likely that these groups will be shown to be significantly different from each other.

8c. This one is not easily determined. A T-test would have to be run to calculate the p value. The two means are fairly close to each other, but the standard deviation is also small.

Chapter XXII.2. Evidence-Based Medicine

1. I) Identify the clinical question. II) Search for sources of information. III) Identify the source(s) found. IV) Determine whether the results are valid. V) Determine what the results are. VI) Determine whether the results will help you in caring for your patients. VII) Resolve the clinical question.

2. Randomization ensures that both known and unknown factors are evenly distributed between the treatment and control groups, making it more likely that any difference in outcome between the two groups is due to the treatment effect alone.

3. This means that during the analysis of the study results, patients remain in the groups to which they were randomized in the beginning of the study, even if they are unable or unwilling to complete the treatment.

4. Relative risk reduction (RRR) = $1 - Y/X$. Absolute risk reduction (ARR) = $X - Y$. Number needed to treat (NNT) = $1/ARR$. See Table 3.

5. The "95% CI," which means that the exact RRR lies within the range of the confidence interval 95% of the time. The CI speaks to the power of a study, and the factor that has the most impact on a study's power is its sample size.

6. It is well known that if a patient or worker knows that a patient is receiving the study medication, this will bias their assessment of the patient's outcome.

7. Sensitivity = $a/(a+c)$. Specificity = $d/(b+d)$. Positive predictive value (PPV) = $a/(a+b)$. Negative predictive value (NPV) = $d/(c+d)$. See Table 5.

8. LR for a positive test result (+LR) = $[a/(a+c)]/[b/(b+d)]$ = sensitivity/(1-specificity). LR for a negative test result (-LR) = $[c/(a+c)]/[d/(b+d)]$ = (1-sensitivity)/specificity. LRs are different from sensitivity and specificity because they take into account each individual patient, using the pretest and posttest probabilities.

9. The pretest probability is the clinician's "gestalt" about the chances that a patient has a particular condition based on clinical information such as symptoms, risk factors, and physical examination. The LR then determines how a diagnostic test will affect the pretest probability, making a disease more or less likely, the outcome of which is called the posttest probability.

10. a) Improves the uniformity and standardization of care so that all patients receive optimal care; b) Helps providers make better use of limited resources by seeking the most effective treatments; c) Prevents harmful side effects or outcomes; and d) Makes the literature accessible to all, thereby helping clinicians make the most informed decisions possible.

Chapter XXII.3. Epidemiology and Research Methodology

1. No. Since the Acme emergency department has a hospitalization rate 6%, we know that 94% are not hospitalized. By just stating that all patients do not require hospitalization, I have a 94% chance of predicting this correctly. Therefore, no test at all is better than the 93% predictive value of the Acme physicians' tests. Although 93% sounds like a good number, it is actually a poor number in this case.

2. The more studies you see in the literature on a topic, the more controversial it must be. If the answer were clear-cut, no further publications on the topic are necessary. However, in controversial subject areas, multiple publications are often present in the literature, attempting to clarify the controversy. Thus, the correct conclusion should be that the efficacy of midazolam for pediatric sedation is controversial.

3. No matter what country you live in, we all eventually die. The mortality rate in all countries is 100%.

4. It could be that since PP is so poor, they don't have an organized health department which keeps accurate health statistics. When the World Health Organizations asks PP to submit their age adjusted mortality rate, the health minister in PP just writes down any number and sends it in. This random number just happens to be lower than the accurately determined age adjusted mortality rate submitted by WW. Another explanation is that since PP has such a poor health care system, any patient who is very ill, is illegally smuggled over the border into WW where the patient shows up in an emergency room. The ethical staff in WW hospitals take care of these very ill patients who frequently die. These death statistics are registered in the health statistics of WW. Thus, many deaths that should have been attributed to PP, actually show up in the age adjusted mortality rate of WW instead. Such a phenomenon could make it appear that many people in PP never die.

5. Prevalence is better for diabetes. Chronic diseases are best described with prevalence while acute diseases are best described with incidence. These numbers may not be very accurate. They may come from disease condition registries or from health department statistics. These systems require that hospitals and/or physicians send in report cards diagnosing the patient's condition so that the statistic can be kept. However, such reports are frequently not made even in "reportable" diseases which the law requires to be reported. Diabetes is not a reportable illness. Another source may be health insurance claims information which contain diagnostic codes.

6. Sensitivity = $TP/(TP+FN)$ = the fraction of all true positives that are caught by the test. A very sensitive test identifies most of the true positives. However, there may still be a substantial number of false positives in a highly sensitive test. Specificity = $TN/(TN+FP)$ = the fraction of negatives that are true negatives. A very specific test correctly identifies most of the true negatives. However, there may still be a substantial number of false negatives in a highly specific test. PPV = $TP/(TP+FP)$ = the likelihood of having a disease if the test is positive. NPV = $TN/(TN+FN)$ = the likelihood of not having a disease if the test is negative. The NPV frequently has a deceptively high value, such as >90% if the disease condition is infrequent.

7. It is possible. Most of these tests are gold standards since they are nearly perfect and they actually define the disease entity. But generally if a study publishes only two out of these four values, it is likely that are publishing the two best values and the authors have suppressed the other two values which do not appear as good. A test which is has nearly perfect sensitivity, specificity, PPV and NPV is the pregnancy test. Lumbar puncture for meningitis is also quite good. Radiographic images for certain types of fractures which are obvious (e.g., forearm fractures) are also quite good.