

Answers

1.

The correct answer is B. In amyloidosis, the left ventricular wall appears speckled on the echocardiogram, and there is a restrictive cardiomyopathy. In such a condition, ventricular filling is impaired, and the cardiac silhouette may be mildly enlarged. An ECG may reveal a host of nonspecific arrhythmias. Primary cardiac amyloidosis usually develops into diastolic dysfunction.

Alcoholic cardiomyopathy (**choice A**) is typically the cause of a biventricular dilated cardiomyopathy, which leads to both right- and left-sided heart failure. An S3 will be heard. An echocardiogram will show enlarged left and right ventricles. The walls of the ventricles may appear very thin and stretched, consistent with volume overload.

Hemochromatosis (**choice C**) also may cause a restrictive cardiomyopathy, as seen in amyloidosis. However, the speckled pattern mentioned above would be absent. Other noncardiac features include bronzing of the skin and diabetes.

Tuberculosis (**choice D**) may cause a chronic tuberculous pericarditis that can manifest clinical symptoms similar to those seen in constrictive cardiomyopathy. The presentation is similar to that seen with restrictive features. However, patients tend to have normal ventricular wall thickness on echocardiogram, pericardial calcification, an absent S3, and S4.

Viral myocarditis (**choice E**), like alcohol, can lead to a dilated cardiomyopathy. Unfortunately, such conditions may progress to complete left and right ventricular failure, ultimately requiring cardiac transplantation in refractory cases.

2.

The correct answer is A. This patient will also likely have absent breath sounds in the right hemithorax. The question clearly describes the sudden hemodynamic collapse in a patient who has developed a tension pneumothorax while being given positive-end expiratory pressure (PEEP) on a respirator. The sudden hypotension and decreased oxygenation is consistent with a tension pneumothorax, which is compressing venous return to the right side of the heart and thus producing jugular venous distention. The treatment would be the immediate placement of either a needle or tube thoracostomy to allow rapid re-expansion of the right lung.

A high amplitude cardiac upstroke pulse (**choice B**) is usually seen in conditions such as aortic regurgitation, in which extra blood must be pushed into the aorta to compensate for backflow into the heart during diastole through an incompetent valve.

A pleural friction rub (**choice C**) suggests inflammation of the pleura, which can be seen either when an underlying lung process involves the lung tissue adjacent to the pleura or when infection or irritating substances are present in the pleural space. It would not be expected in the setting of a pneumothorax, in which the visceral pleura becomes separated from the parietal pleura by a large volume of air.

Pulsus alternans (**choice D**), also known as alternating pulse, is a pulse in which the beat is regular but alternate beats are weaker or stronger. It usually indicates serious myocardial disease.

Splenomegaly (**choice E**) is not usually a feature of tension pneumothorax because the

venous return from the abdomen enters the heart through the inferior vena cava, which is much less affected by the pneumothorax than is the superior vena cava.

3.

The correct answer is A. This patient has multiple risk factors for atherosclerotic coronary artery disease: hypertension, hypercholesterolemia, as well as a positive family history. A stress test is considered positive when there are ST depressions of greater than 1 mm that last longer than 0.08 seconds or when there is development of hypotension (10 mm Hg drop in systolic blood pressure) or the appearance of S3 or S4 heart sounds. Patients with positive stress tests require a coronary angiogram as the next step to assess the need for revascularization.

A Holter monitor (**choice B**) is used to detect intermittent myocardial ischemia or paroxysmal arrhythmias. The Holter monitor continuously monitors electrocardiac and rhythm changes for 24 hours during symptomatic periods. Because ECG changes are already documented on an exercise stress test, a Holter monitor is not required.

Not performing any further tests (**choice C**) is not appropriate because he is symptomatic and has a positive exercise stress test.

Performing a stress test with thallium (**choice D**) is a more sensitive way of detecting atherosclerotic heart disease. But because he already has a positive exercise stress test, repeating the test with thallium is not needed.

An upper gastrointestinal endoscopy (**choice E**) is performed when reflux or peptic ulcer disease is thought to be the cause of chest pain.

4.

The correct answer is B. This patient, with a long history of chronic obstructive pulmonary disease (COPD), has evidence of a community-acquired pneumonia. The common organisms causing pneumonias in patients with COPD are *Streptococcus pneumoniae*, *Haemophilus influenzae*, and *Moraxella catarrhalis*.

This patient has no other history suggestive of *Escherichia coli* (**choice A**) infection elsewhere (such as in the urinary tract), and primary *E. coli* pneumonia is rare.

Klebsiella pneumoniae (**choice C**) is typically found in alcoholic patients and it may cavitate.

There is no evidence of tuberculosis (**choice D**) by history. Tuberculosis usually presents with a more chronic presentation. Furthermore, it would generally be found as an upper lobe infiltrate, consistent with reactivation tuberculosis. Much less commonly, tuberculosis may present as a primary infection, but this is generally seen in patients with an underlying immunocompromised state. In the setting of primary tuberculosis, a lower lung field pneumonia is in fact possible.

Mycoplasma pneumoniae (**choice E**) does not present with a lobar consolidation and is generally a disease of younger people who present with fever, malaise of at least several days duration, and a nonproductive cough. The chest x-ray film in a patient with *Mycoplasma pneumoniae* would classically reveal faint bilateral interstitial infiltrates.

5.

The correct answer is A. Reinfarction in a hospitalized patient is evaluated best by creatinine kinase (CK). CK, total levels and specific MB fraction, are elevated as early as 3 hours after onset of chest pain, and, more importantly, have a duration of no more than 2 days, peaking within 18 to 24 hours. As such, CK levels are easier to track. A sudden increase in CK levels represents new infarction rather than a delayed effect from a

previous myocardial event.

Dynamic EKG changes (**choice B**) are not sensitive for infarction. Further, this patient has a left-bundle branch pattern on EKG, making interpretation of any EKG changes impossible.

Lactate dehydrogenase (LD) (**choice C**) and relative amounts of different LD isoforms are not as common now that more sensitive and specific serum markers are available. Further, LD remains elevated for 6 to 8 days. An elevated LD in this patient would be difficult to interpret, as it would not be clear whether the enzymes were still elevated from the first event.

Myoglobin (**choice D**) has many characteristics that make it seem an excellent serum marker for an acute event. It is the first enzyme elevated, often within an hour or two, peaks relatively early within 6 to 7 hours, and lasts no more than 1 day. It is notoriously nonspecific, however, and is often elevated in hospitalized patients for unrelated reasons.

Troponin levels (**choice E**) increase in 3 to 12 hours, peak in approximately 1 day, and gradually taper over the next 10 days.

6.

The correct answer is A. The combination of hematuria and hemoptysis should always raise the possibility of Goodpasture syndrome. Anti-glomerular basement membrane antibodies are pathognomonic for this diagnosis.

Anti-mitochondrial antibodies (**choice B**) are found in patients with primary biliary cirrhosis.

The anti-neutrophilic cytoplasmic antibodies (**choice C**) are found in patients with Wegener granulomatosis. Wegener granulomatosis may also present with pulmonary and renal involvement but will have associated upper respiratory tract findings, e.g, sinusitis and sinus abscesses.

Anti-parietal cell antibodies (**choice D**) are found in patients with the autoimmune disease known as pernicious anemia.

Anti-smooth muscle antibodies (**choice E**) are found in patients with autoimmune hepatitis.

7.

The correct answer is E. Pesticide exposure should prompt one to think of organophosphate poisoning. Organophosphate poisoning inhibits cholinesterase, resulting in an accumulation of acetylcholine. This results in cholinergic excess at muscarinic sites (salivation, arrhythmia, bronchoconstriction) and peripheral nicotinic sites (such as sweating and weakness). Pralidoxime activates acetylcholinesterase, reversing the effects of most, but not all, organophosphates. In contrast, atropine (**choice A**), which competes with acetylcholine only at muscarinic receptors, will not reverse the nicotinic effects of any organophosphates.

Charcoal (**choice B**) is inappropriate in this situation. Most pesticide exposure is topical, and thorough surface decontamination (skin, clothing, contacts), but not gastric decontamination, is warranted.

Glucagon (**choice C**) will reverse beta-blocker overdose and can be used to treat severe hypoglycemia.

Naloxone (**choice D**) is used to reverse the effects of opioids. It has no use in treating pesticide overdose.

8.

The correct answer is A. Since the patient can cough and breathe, he should be allowed to clear the foreign object spontaneously, if possible. In the management of foreign object obstruction, if the patient can cough and breathe, it is best to initially observe and allow spontaneous resolution, since intervention may actually be damaging. Often, blind finger sweeps (**choice B**) may remove the foreign object and resolve the symptoms. Also, this will need to be done if the patient is to be intubated. The next step would be performing back blows if the patient was less than 1 year of age (**choice C**).

If the patient were over 1 year old, abdominal thrusts would be the next management option (**choice D**).

An emergency tracheostomy (**choice E**) should be the last option and should be undertaken only by a physician trained to perform the procedure.

9.

The correct answer is C. The most likely diagnosis is a dystonic reaction to the droperidol. Droperidol causes its antiemetic effect by antagonizing dopaminergic receptors in the vomiting center (central chemoreceptor zone) of the brain. This antidopaminergic action can produce torticollis or other dystonias.

A cerebral vascular accident (**choice A**) is unlikely given that the patient is alert and oriented, has no detectable language deficit, and has an otherwise nonfocal neurologic examination.

A conversion disorder (**choice B**) is unlikely since the patient has no prior history of a psychiatric disorder and has a viable medical reason (dystonia from droperidol) for her neuromuscular deficit.

Munchausen syndrome (**choice D**) is also unlikely since the patient had valid medical reasons for her initial admission and your current visit. We are also not informed of any prior history of hospitalizations or seeking of medical attention without appropriate cause.

A seizure (**choice E**) is similarly unlikely since the patient has no history of a seizure disorder and is alert, oriented, and conversant.

10.

The correct answer is E. This patient has a classic presentation for variant angina, which is caused by coronary vasospasm that induces transient ischemia and ST-segment elevations. Vasospasm often is seen in the distribution of the right coronary artery, which can result in transient inferior ischemia. The ST-depressions in the anterior leads likely represent reciprocal changes rather than the primary pathology. Associated vascular phenomena, such as Raynaud phenomenon or migraines, are common clues to the diagnosis. ST-segment elevation that responds to nitroglycerin makes the diagnosis almost certain, because a transmural myocardial infarction, caused by plaque rupture and thrombus formation (**choice D**), does not have transient ST-segment elevations.

Diffuse intimal thickening and focal areas of atherosclerotic narrowing (**choice A**) may be seen in severe atherosclerotic disease and classic angina. Classic angina may present with ST-segment depressions but would not cause transiently increased ST-segment elevation.

Intermittent thrombus formation and lysis (**choices B and C**) is the pathophysiology underlying unstable angina. It is an extremely unlikely diagnosis in a young, healthy woman with no coronary risk factors. Variant angina, also known as Prinzmetal angina,

is a more likely diagnosis.

11.

The correct answer is A. Abdominal ultrasound (U/S) is the most cost-effective screening test for a suspected abdominal aortic aneurysm (AAA). A CT of the abdomen (**choice D**) with IV contrast is about twice as expensive as U/S, adds little if any benefit, and exposes the patient to unnecessary radiation.

The other choices, lumbosacral (L/S) spine film (**choice B**), CT of the spine (**choice C**), and spinal MRI (**choice E**) are imaging studies directed at evaluating spinal pathology, which is not suspected here. MRI provides the highest resolution and is useful for detecting abscesses or cord compression. CT is excellent for disk pathology and even bone pathology. Plain radiographs are useful for very gross visualization of the bony density and integrity.

12.

The correct answer is A. This patient should have antibiotic prophylaxis before undergoing dental work. The patient's physical examination is consistent with asymptomatic aortic insufficiency, as indicated by his lack of symptoms combined with a characteristic diastolic murmur. This has occurred as a result of his childhood rheumatic fever. Patients with any significant cardiac valvular disease should be instructed to have antibiotic prophylaxis before dental work to reduce the risk of subacute bacterial endocarditis.

Although this man is at increased risk for lung cancer given his long history of smoking, chest x-ray films (**choice B**) have never been proven effective as early detection.

Although he does have underlying valvular heart disease, there is no indication for an annual echocardiogram (**choice C**) unless specific symptoms develop and warrant evaluation.

A sigmoidoscopy (**choice D**) is one of several choices that are appropriate colorectal cancer screening examinations beginning at age 50.

Prostate specific antigen testing (**choice E**) remains controversial in asymptomatic adults and is certainly not recommended in asymptomatic men younger than 50.

13.

The correct answer is C. It would be most appropriate to add an IV steroidal agent, such as hydrocortisone, to augment the action of bronchodilators by reducing inflammation surrounding the airways.

Beclomethasone (**choice A**), a surface-acting steroid dispensed in aerosolized form, is used when the side effects of systemic steroids need to be avoided. Such agents, however, also have minimal utility in the acute treatment of bronchospasm.

Disodium cromoglycate (**choice B**) is a mast cell stabilizer that is used only for the prevention of bronchospasm due to asthma. Once bronchospasm is established, this type of agent has little utility.

An IV steroid is preferred over an oral steroid, such as prednisone (**choice D**), since this patient has a history of recent nausea and vomiting and, therefore, may have impaired absorption of oral medications. In addition, the time course of onset of an oral agent is too long to have any effect in an acute situation.

Theophylline (**choice E**), a methylxanthine phosphodiesterase inhibitor, is useful for the chronic control of asthma, but it has lesser utility in the acute treatment of bronchospasm.

14.

The correct answer is C. This patient has sustained first-degree burns of the face and superficial second-degree burns of the scalp and both arms, for a total of 27% estimated body surface area involvement based on the "rule of nines." When evaluating a burn victim, estimation of body surface area of the burn should be performed. This can be done by using burn charts for children of different ages up to the age of 14 years, or by means of the "rule of nines" used in adults and in children 14 years of age or older. The "rule of nines" allows a quick estimate for preliminary calculations of the required volume of resuscitation fluid. The head and arms are counted as 9% each. The legs are estimated to be 18% each, and the trunk is 36% (18% for the front and 18% for the back). The "rule of palm" may be used for burns that involve less than 10% body surface area, in which the child's palm equals 1% of the child's body surface area. Based on the estimation of body surface area involved by burn injury by using the "rule of nines," 9% (**choice A**), 18% (**choice B**), 36% (**choice D**), and 45% (**choice E**) are incorrect.

15.

The correct answer is E. This patient most likely has acute bronchitis. Acute bronchitis in a healthy patient with no other medical conditions is often due to a viral infection that is usually self-limited. Given that this patient has only had 3 days of symptoms, an antibiotic is not necessary and is inappropriate. If the symptoms persist for longer than 1 week, a macrolide antibiotic may be given. A chest x-ray film and a sputum culture are not indicated.

Admission to the hospital for medical management (**choice A**) is inappropriate for a healthy patient with acute bronchitis.

A chest x-ray film (**choice B**) has no role in the diagnosis of acute bronchitis in a healthy patient.

A sputum culture (**choice C**) is used to identify organisms, but should only be used in elderly patients with chronic disease that fail antibiotic therapy.

Sending the patient home with antibiotic therapy (**choice D**) is appropriate management for acute bronchitis in an elderly patient with chronic disease. A macrolide is the treatment of choice

16.

The correct answer is E. This child has apnea of prematurity (AOP), a common problem that affects premature infants and, to a lesser degree, term infants. Apnea is the cessation of air flow/exchange for >20 seconds and is often associated with bradycardia and hypoxemia, as seen in this case. It occurs in approximately 50% of infants born at 30 to 31 weeks' gestation and appears to be due to immaturity of the infant's neurologic and respiratory systems. The first-line pharmacologic agents of choice for the management of AOP are methylxanthines (caffeine and theophylline) which act to stimulate respiratory neurons. AOP usually resolves by the time the infant is 34 to 36 weeks gestational age.

Bicarbonate (**choice A**) may be used to correct documented acidosis. Because routine laboratory tests are within normal limits, bicarbonate is not indicated.

Dextrose (**choice B**) is used for the correction of hypoglycemia, which can occur in premature infants. However, hypoglycemia typically does not manifest as apnea. Furthermore, the normal laboratory panel does not reveal hypoglycemia.

Epinephrine (**choice C**) is used in neonatal resuscitation when the heart rate stays below 80/min despite effective ventilation with 100% oxygen and chest compressions for at least 30 seconds. It is not the appropriate management for the bradycardia found in AOP.

17.

The correct answer is E. Total anomalous pulmonary venous return is characterized by the pulmonary veins forming a confluence behind the left atrium, and draining into the right atrium. Complete mixing takes place in the right atrium, with a right-to-left shunt through the foramen ovale to the left side of the heart. Often, no murmur is heard on cardiac examination, although a short systolic murmur is sometimes heard. ECG often reveals right atrial enlargement and right ventricular hypertrophy. The chest roentgenogram often shows a normal heart size with pulmonary edema. If there is obstruction to pulmonary venous return, as is almost always present with veins draining inferior to the diaphragm, cyanosis can be very prominent. Definitive treatment is surgical anastomosis of the pulmonary vein to the left atrium.

Atrial septal defect (ASD) (**choice A**) is a hole in the septum between the right and the left atria. It results in a left-to-right shunt and causes right ventricular volume overload and increased pulmonary blood flow. Approximately 3% to 5% of children with congenital heart disease have an ASD, making it the third most common congenital heart defect.

Hypoplastic left heart syndrome (**choice B**) is characterized by underdevelopment of the left ventricle and the ascending aorta. Typically, there is obstruction at the mitral valve, causing all pulmonary venous blood to shunt through either an ASD or a patent ductus arteriosus (PDA) into the right atrium. Total systemic blood flow is channelled through the ductus arteriosus from the pulmonary artery. As the ductus closes, these infants present with shock because systemic blood flow is significantly reduced.

PDA (**choice C**) causes symptoms of pulmonary congestion, dyspnea, widened pulse pressure, and bounding arterial pulsation because aortic blood flow is shunted from left to right.

18.

The correct answer is B. The diagnosis of pneumonia absolutely requires that an infiltrate of some sort (whether interstitial or parenchymal, lobar or diffuse) be present on a chest radiograph.

Hypoxemia on pulse oximetry (**choice A**) is one possible physical manifestation of severe pneumonia but is certainly not required for the diagnosis.

A sputum Gram's stain showing gram-positive diplococci (**choice C**) is suggestive of a pneumonia, but it could also easily be explained by pharyngeal colonization with this organism.

A sputum Gram's stain showing neutrophils (**choice D**) is also highly suggestive of some pulmonary inflammatory process, although not necessarily pneumonia.

A temperature to 38.6 (101.4 F) (**choice E**) is clearly a nonspecific sign and can be associated with a multitude of possible etiologies.

19.

The correct answer is E. This patient most likely has duodenal atresia that may have been detected with prenatal ultrasonography. Duodenal atresia is an obstruction resulting from a failure of recanalization of the duodenal lumen, resulting in complete intrinsic obstruction. It occurs in 1/10,000 live births. Twenty to thirty percent of patients

with duodenal atresia have trisomy 21. There is no predilection for sex or race. Duodenal atresia presents early, usually in the first hours or day of life. The neonate has bilious vomiting without abdominal distension. In approximately 15% of neonates with this anomaly, vomiting is nonbilious because the obstruction is located proximal to the ampulla of Vater. There may be a history of polyhydramnios (which is commonly associated with diabetes and twin pregnancy). Prenatal ultrasonography may reveal a dilated and fluid-filled stomach and duodenum in addition to other possible anomalies. On physical examination, the newborn may have a scaphoid abdomen and epigastric fullness from a dilated stomach and proximal duodenum. Nasogastric suction produces bilious fluid. Abdominal films reveal the characteristic "double bubble" of the stomach and proximal duodenum. Therapy involves surgical correction. The patient should be evaluated for associated anomalies.

Elevated alpha-fetoprotein levels (**choice A**) have been reported in fetomaternal hemorrhage, umbilical cord hemangioma, polycystic kidneys, cytomegalovirus, and parvovirus infections, as well as central nervous system anomalies.

20.

The correct answer is B. This patient has an endocardial cushion defect. Endocardial cushion defect occurs when an atrial septal defect and ventricular septal defect are present and contiguous, and the atrioventricular valves are also abnormal. Patients with trisomy 21 are at highest risk. Patients who have a large atrial septal defect tend to have heart failure early in infancy, with hepatomegaly and failure to thrive. Increased pulmonary blood flow over time leads to pulmonary vascular disease and Eisenmenger physiology. A systolic thrill is appreciable on physical examination. With increased pulmonary blood flow, S2 becomes widely split. A systolic ejection murmur at the upper left sternal border and a low-pitched diastolic murmur may be present, with the first heart sound accentuated. Chest radiographs show a markedly enlarged heart, reflecting enlargement of all chambers. The main pulmonary artery is usually prominent with increased pulmonary vascular markings. After pulmonary hypertension develops, a reduction in pulmonary vascular markings is observed. Electrocardiography shows left axis deviation, biventricular hypertrophy, right ventricular conduction delay, and tall P waves. The echocardiogram reveals right ventricular enlargement and a common atrioventricular valve. Color flow demonstrates shunting at the atrial and ventricular levels. The treatment is surgical, preferably early in infancy because of the high risk for early development of pulmonary hypertension. The defects are patched and the cleft mitral valve is repaired. Complications and prognosis depend on the size of the shunt and the amount of pulmonary vascular resistance. Without surgery, death from heart failure results.

21.

The correct answer is D. The tidal volume for a patient is generally estimated as 10 mL/kg of weight, which for this patient would be 850 mL/breath. Giving a lower tidal volume will yield hypoventilation and be insufficient to eliminate pCO₂. Providing a tidal volume greater than 10 mL/kg increases the risk of pneumothorax, particularly in a patient with longstanding emphysema who may have thin-walled alveoli.

A low tidal volume with risk of hypoventilation would be produced by **choice A** (500 mL/breath), **choice B** (600 mL/breath), and **choice C** (700 mL/breath).

A high tidal volume with risk of pneumothorax would be produced by **choice D** (1000

mL/breath).

22.

The correct answer is D. Barrett's esophagus may occur in a small number of patients who have gastroesophageal reflux disease (GERD). This condition is a metaplasia of the normal squamous mucosa of the esophagus to a columnar (glandular) type of epithelium, and is usually seen as a response to repeated acid exposure to the distal esophagus. Tobacco and alcohol use are also thought to contribute to the process. The significance of Barrett's esophagus is that it may lead to the development of low-grade dysplasia, high-grade dysplasia, or esophageal adenocarcinoma. However, this is a very infrequent occurrence when considering the large number of patients with GERD and even those with Barrett's esophagus. Barrett's esophagus usually does not resolve with either medical or surgical therapy. Endoscopic surveillance (with multiple small biopsies, since dysplasia cannot be reliably evaluated by endoscopic appearance alone) every 1-2 years has been often recommended, but some studies suggest that it may not be cost-effective. It is not factually true to inform the patient that his concerns are "ungrounded" (**choice A**), because there is in fact a small risk of adenocarcinoma.

It is never appropriate to belittle a patient's concerns and inform him that his worries are "foolish" (**choice B**).

Barrett's esophagus is a histologic change and, unless accompanied by a stricture, does not produce symptoms of mechanical dysphagia (**choice C**).

It is inappropriate to refer the patient to a cancer specialist (**choice E**) for the prevention of a very unlikely development of cancer; furthermore, preventive strategies should include instructions to avoid factors that exacerbate GERD.

23.

The correct answer is C. No further steps are necessary because the patient makes the medical decisions, not the physician. Competent, informed patients can refuse recommended interventions. In such cases, it is important for the physician to document in the patients' charts that they were informed of the potential risks of refusing treatment.

A court order (**choice A**) and the surgery chairman's decisions (**choice B**) are not indicated because this is a competent and informed patient who can refuse treatment.

Sending the patient to another primary care physician because of opposing views (**choice D**) is inappropriate because the patient should never be abandoned. If the physician provides the appropriate information and the competent patient makes an informed decision, there is no reason to try to send her to another physician because of differing beliefs.

One psychiatrist concluded that this patient is competent; there is no reason to doubt the evaluation and send her to another (**choice E**).

24.

The correct answer is C. It is generally accepted that the blood supply from 1978 to 1985 was likely to be tainted with HIV positive blood. For this reason, as well as the indolent nature of HIV infection, patients with a history of blood transfusions during these years, even if currently asymptomatic, should be screened.

Testing for hepatitis B (**choice A**) or hepatitis C (**choice B**) would not be incorrect, but there are no formal guidelines recommending this. The primary reason is that such diseases generally manifest in some way after a 20-year period, and the patient would not be asymptomatic.

PPD skin testing (**choice D**) is reserved for health care workers, alcoholics, IV drug abusers, diabetics, and end-stage renal patients. This does not change with a history of blood transfusion.

RPR, or rapid plasma reagin testing (**choice E**), is used to test for syphilis. It is recommended if the patient is pregnant, has a history of sexual transmitted diseases, or is a prostitute.

25.

The correct answer is D. This patient's history suggests pheochromocytoma. This rare (but often considered diagnostically) tumor is most often found in the adrenal medulla, although it can also be found in other tissues derived from neural crest cells. The tumor cells secrete catecholamine hormones or their precursors, which can cause either paroxysmal (as in this case) or persistent hypertension. Urinary metabolites of epinephrine and norepinephrine are vanillylmandelic acid (VMA) and homovanillic acid, so screening 24 hour urine collections for these substances can be helpful in establishing or excluding these diagnoses even in cases in which a physician does not observe one of the paroxysms and thus blood cannot be drawn for serum catecholamine levels at that time.

DHEA (**choice A**) is the adrenal androgen dehydroepiandrosterone (made by the adrenal cortex rather than the adrenal medulla), and is measured in serum in cases where adrenal virilism is suspected.

hCG (**choice B**) is human chorionic gonadotropin, and both serum and urine levels can increase in pregnancy or trophoblastic disease.

17-ketosteroids (**choice C**) are measured in urine during evaluation of congenital adrenal hyperplasia (a disorder of the adrenal cortex rather than medulla).

Zinc protoporphyrin (**choice E**) is measured in blood when evaluating possible porphyria.

26.

The correct answer is A. This patient has probable acute hepatitis. Features specifically suggesting acute hepatitis include his jaundice, itchiness leading to multiple excoriations, tender enlarged liver, and palpable spleen tip. He reports no risk factors for hepatitis A infection, such as drinking water in a foreign country with periodic epidemics of hepatitis A. He does have a risk factor for hepatitis B, which can be transmitted through blood products (now rare because of blood screening), contaminated syringes among drug users, and sexual contact (particularly when involving the rectum). The findings of a coagulopathy or of an encephalopathy confer the worst prognosis in patients with an acute viral hepatitis. Most patients with hepatitis B, even with severe laboratory abnormalities, may be followed conservatively so long as they do not develop a coagulopathy or exhibit signs of encephalopathy. These findings, in fact, suggest the possibility of fulminant hepatic failure.

Although the transaminases may rise to very high levels (**choices B and C**), they are not of prognostic value in viral hepatitis.

Similarly, severe jaundice (**choice D**) is of limited prognostic value, as is leukocytosis (**choice E**).

27.

The correct answer is D. The patient has rhinocerebral mucormycosis, which can be caused by fungal species including *Rhizopus*, *Rhizomucor*, *Absidia*, and *Basidiobolus*. Predisposing conditions include immunosuppression, uncontrolled diabetes mellitus, and patients using the iron-chelating drug desferrioxamine. While these infections

occasionally appear more or less incidentally, as in this case, they are very important to diagnose because they have a tendency to become fulminant. The lesions tend to be very locally destructive and can erode into the eye, palate, and central nervous system, often from an initial site in the sinuses. Fulminant infections are frequently fatal. Pulmonary infections can also occur. The organism can be difficult to culture. The appropriate antibiotic is intravenous amphotericin B, but surgical debridement should also be strongly considered, since penetration of antibiotic into necrotic tissues may be poor.

Aspergillus (**choice A**) can also cause sinusitis, but has narrow hyphae.

Blastomyces (**choice B**) usually involves the lung and occurs as a yeast form in the body.

Candida (**choice C**) can infect sinuses, but has narrow hyphae and yeast forms.

Sporothrix (**choice E**) usually infects the skin and subcutaneous tissues and occurs as a yeast.

28.

The correct answer is E. The infant in this clinical vignette has hemorrhagic disease of the newborn as a result of vitamin K deficiency. It was a major cause of bleeding in neonates in the past, but it is now uncommon because of the routine administration of vitamin K at birth. However, it is still encountered in situations in which babies are born outside the hospital. The normal newborn has a moderate deficiency of the vitamin K-dependent coagulation factors. The plasma levels of these factors fall even further during the first 2-5 days of life, rise again when the infant is 7-14 days old, and attain normal adult levels at about 3 months of age. This variation usually does not produce any bleeding or bruises. However, in hemorrhagic disease of the newborn, the initial fall is accentuated, and the restoration is delayed and incomplete. As a result, coagulation abnormalities become severe and bleeding may occur. All newborns should receive 0.5-1.0 mg of vitamin K intramuscularly within the first hour after birth.

Prematurity has been associated with hemorrhagic disease of the newborn. Delayed feeding, breast-feeding, vomiting, severe diarrhea, and antibiotics also delay the colonization of the gut by bacteria.

Bleeding is usually severe and occurs most commonly on the 2nd or 3rd day of life. The most common manifestations are melena, large cephalohematomas, and bleeding from the umbilical stump and after circumcision. Generalized ecchymoses, often without petechiae, intracranial bleeding, and large intramuscular hemorrhages, also may develop in severe cases. In infants with hemorrhagic disease of the newborn, the prothrombin time (PT) is always prolonged. The partial thromboplastin time (PTT) and the thrombin time are also prolonged. Specific factor assays reveal deficiencies of prothrombin; factors VII, IX, and X; and proteins C and S. The bleeding time and the platelet count usually are within normal limits. In the differential diagnosis of hemorrhagic disease of the newborn, virtually all causes of bleeding, particularly thrombocytopenia and disseminated intravascular coagulation (DIC), must be considered.

29.

The correct answer is C. Papillary muscle rupture occurs in less than 5% of patients with acute myocardial infarction. The patient usually presents with acute onset of congestive heart failure approximately 1-7 days after infarction. Rupture of an entire papillary muscle is usually rapidly fatal secondary to massive mitral regurgitation.

However, rupture of one head of a papillary muscle may be tolerated for a period of time. A loud systolic murmur is heard at the left sternal border, and echocardiogram shows a flail mitral valve. Surgical repair or replacement of the valve can be effective if the patient tolerates the procedure.

Aortic dissection (**choice A**) is associated with the sudden onset of severe chest pain radiating to the abdomen and back. Pain may radiate as the dissection progresses.

Auscultation is significant for murmur of aortic insufficiency. Pulses may be absent or asymmetric.

Cardiac rupture (**choice B**) is usually rapidly fatal. Rupture usually occurs approximately 5 days after transmural infarction. The patient's initial course may be uneventful, followed by abrupt occurrence of cardiogenic shock and rapid death.

Septal perforation (**choice D**) of the ventricle occurs in less than 1% of patients following myocardial infarction and results in marked biventricular failure. It is associated with a holosystolic murmur along the left sternal border, and a thrill is commonly appreciated. It is difficult to differentiate from acute mitral insufficiency.

Ventricular aneurysm formation (**choice E**) is a localized area of thin scarred myocardium that protrudes beyond and distorts the ventricular cavity. It may develop days following myocardial infarction and may gradually enlarge over several weeks.

30.

The correct answer is C. This patient has symptoms of exercise-induced asthma.

Unsurprisingly, he appears normal while in the office. The symptoms of exercise-induced asthma are due to mast cell release of histamines, which degranulate with the initiation of exercise. These symptoms can be prevented with the pre-exercise use of inhaled cromolyn, which will act to stabilize the mast cells. It is less effective once exercise has begun. Once bronchoconstriction has occurred, symptomatic therapy can be provided with a beta-agonist inhaler.

Eosinophils (**choice A**) are involved in allergen-induced asthma.

Lymphocytes (**choice B**), monocytes (**choice D**), and neutrophils (**choice E**) are involved with inflammation, but do not mediate the process of asthma.

31.

The correct answer is E. Gastric ulcers are associated with malignancy, and therefore biopsy should be performed when they are discovered. This association with malignancy is not found with duodenal ulcers. The increased risk for malignancy with gastric ulcers is a good reason to test patients for *H. pylori* and initiate treatment if it is found.

Gastric ulcers have a variable response to medications (**choice A**). In contrast, duodenal ulcers have a better and more reliable response to treatment with H₂ blockers and proton pump inhibitors.

As in our patient, gastric ulcers often present with significant gastrointestinal bleeding.

These bleeds can be serious and life threatening and sometimes require operative interventions. Although duodenal ulcers can perforate and bleed, they do so less commonly than gastric ulcers (**choice B**).

The gastric pH in patients with gastric ulcers is higher (less acidic) than the gastric pH in patients with duodenal ulcers (**choice C**). This might explain the discrepancy in response to acid suppressing medication in the two conditions.

Initially, it was believed that duodenal ulcers were more commonly associated with *H. pylori* than are gastric ulcers. Now it is believed that the two conditions are probably

equally associated with *H. pylori*. Therefore, **choice D** is incorrect.
32.

The correct answer is A. This patient has Bell palsy, a postinfectious allergic or immune demyelinating facial neuritis. Epstein-Barr virus is the preceding infection in approximately 20% of cases. 85% of patients have their symptoms resolve on their own over a period of several weeks. 10% retain mild facial weakness and 5% have permanent severe facial weakness. Therapeutic intervention should include daily and nocturnal eye lubricants to protect the cornea from drying.

Group A *Streptococcus* (**choice B**), HIV (**choice C**), and influenza (**choice D**) are not associated with Bell palsy. However, herpes simplex virus, Lyme disease caused by *Borrelia burgdorferi* and mumps have been associated with Bell palsy.

Measles (**choice E**) is associated with subacute sclerosing panencephalitis, a chronic encephalitis of the central nervous system manifested by progressively bizarre behavior and decline in cognitive function.

33.

The correct answer is D. The history of severe HIV-related immune compromise and evidence of destruction of myelin at multiple sites in the CNS points to a diagnosis of progressive multifocal leukoencephalopathy (PML). PML is caused by JC virus, a papovavirus that produces asymptomatic infections in immunocompetent hosts. Oligodendrocytes in active lesions contain characteristic intranuclear inclusions. Usually, biopsy of the lesions is not necessary, as a presumptive diagnosis of PML can be made on clinical grounds. PML is also seen in patients with lymphomas, or those receiving organ transplants.

In AIDS-dementia complex (**choice A**), there is no focal brain lesion. Sometimes, diffuse, but usually mild, cerebral atrophy can be identified by MRI. Patients present with progressive dementia, often associated with incontinence and disorientation.

Cerebral toxoplasmosis (**choice B**) typically manifests with a round, well-circumscribed lesion that shows a peripheral rim of contrast enhancement.

CMV encephalitis (**choice C**) has a predilection for the periventricular gray matter and ependyma, as well as the retina.

Multiple sclerosis (MS) (**choice E**) does not develop in the context of immune impairment. Demyelinating plaques of MS are typically well-demarcated and most commonly located in the periventricular regions.

34.

The correct answer is A. This patient had a head trauma with a resultant concussion. The fact that he has motor findings along with memory loss and headache suggest an epidural or subdural hematoma. This is a potentially dangerous situation and requires prompt diagnosis. The procedure of choice and the most appropriate next step in the workup of this patient would be a noncontrast CT scan of the head.

Intubation, hyperventilation, and mannitol (**choice B**) are the treatment for severe intracranial hemorrhage in preparation for surgery. Right now, our patient is alert, oriented, and hemodynamically stable and does not require such aggressive management.

Simple reassurance, acetaminophen, and observation (**choices C and D**) are not appropriate in this patient because the patient had a serious head trauma with a resultant motor deficit. He needs prompt diagnosis with a CT scan.

Skull x-rays (**choice E**) are not helpful in the diagnosis of intracranial hemorrhage. A normal x-ray does not exclude disease, and an abnormal x-ray does not confirm the presence of hemorrhage.

35.

The correct answer is A. Acetylcholine is known to modulate attention, novelty seeking, and memory by way of basal forebrain projections to cortex and limbic structures. In Alzheimer dementia, there is a deficit of acetylcholine, causing cognitive deficits.

Dopamine (**choice B**) affects several brain functions, mostly by modulating other systems. Dopaminergic projections from the ventral tegmental area to the cortex play a role in fine-tuning of attention and ability to screen out irrelevant stimuli. Dopamine counteracts acetylcholine and thus plays a role in Parkinson disease and related subcortical dementias.

Glutamate (**choice C**) has a crucial role in long-term potentiation and thus in formation of long-term memory. Excess stimulation of glutamatergic receptors is seen in stroke, seizures, and neuronal cell death.

Norepinephrine (**choice D**) modulates sleep cycles, mood, appetite, and cognition by targeting thalamus, limbic structures, and cortex. Locus ceruleus is crucial for fine tuning of the attentional tone of cortex. In Alzheimer dementia, only if depression is superimposed, one might want to enhance noradrenergic activity.

Serotonin (**choice E**) is linked to many functions because of widespread projections and a variety of receptors. It has not been shown to have a prominent role in Alzheimer type dementia, rather in mood disorders that could be associated with dementia.

36.

The correct answer is C. One of the most important tasks that physicians can perform in regard to the treatment of epilepsy is counseling caretakers about what to do when seizures occur. Freeman et al. (*Seizures and Epilepsy: A Guide for Parents*, 2nd edition, Johns Hopkins University Press, 1997) list a number of "do's" and "don't's." During the seizure episode, *don't*

Put any object into the patient's mouth
Call an ambulance unless seizing lasts more than 10 minutes
Try to restrain the patient

During a seizure episode, *do*

Place the patient on the side
Put a pillow or other soft object under the patient's head
Loosen tight clothing around the neck
Remove sharp objects from the surroundings

After the seizures, caretakers or parents should remain with the patient until he/she is fully alert and allow him/her to go back to the usual activities.

Calling an ambulance (**choice A**) is appropriate if the seizure episode lasts for more than 5-10 minutes.

Putting something in the patient's mouth at the onset of seizure (**choice B**) increases the risk of suffocation.

Trying to restrain the child during the seizure (**choice D**) is a useless measure.

Not allowing the child to return to his activities (**choice E**) is a mistake if the patient has become fully conscious and alert after recovery.

37.

The correct answer is E. Headache of sudden onset ("thunderclap" headache), rapid deterioration of mental status and blood in the CSF are virtually diagnostic of ruptured

berry aneurysms. Note the characteristic hyperdensity on CT of the suprasellar cistern, indicating blood in the subarachnoid space. Rupture of a berry aneurysm is the most common cause of subarachnoid bleeding. Berry aneurysms develop as a result of congenital weakness at branching points of the arteries in the circle of Willis. These outpouchings tend to expand progressively, but in most cases they remain asymptomatic. Hypertension facilitates development and rupture of berry aneurysm. One third of patients recover, one third die, and one third develop re-bleeding. Rapid onset of coma is an ominous sign.

Amyloid angiopathy-related hemorrhage (**choice A**) would manifest as a cortical-based hematoma in a lobar distribution. It is due to accumulation of A β amyloid in blood vessel walls.

Cavernous sinus thrombosis (**choice B**) is a rare complication of conditions leading to coagulation abnormalities, such as sepsis, antiphospholipid antibody syndrome, and leukemias. It leads to hemorrhagic infarction of large areas of hemispheric gray and white matter.

Hemorrhagic infarction (**choice C**) usually develops as a result of embolic occlusion of an intraparenchymal artery. It gives rise to a hyperdense wedge-shaped area in a cortical field corresponding to a specific vascular territory.

Pituitary apoplexy (**choice D**) refers to hemorrhage in the pituitary gland. It may occur in the setting of a large pituitary adenoma or in pregnancy. It manifests with rapid onset of panhypopituitarism.

38.

The correct answer is D. Trazodone is an older antidepressant medication that is commonly used in lower doses for the treatment of insomnia associated with depression. Due to its significant alpha-adrenergic blocking properties, it has been associated with priapism. Priapism is a serious adverse event defined as a persistent, painful penile erection. Erectile dysfunction occurs in approximately 50% of men suffering an episode of priapism. This patient has a history of a prior episode of priapism associated with an antihypertensive (most likely one with significant alpha-adrenergic antagonism, such as prazosin). Given his history of priapism (and due to the fact that he is likely predisposed to subsequent episodes of priapism) trazodone should be avoided in this patient.

Bupropion (**choice A**) is a newer antidepressant that is not associated with the development of priapism. It is also marketed for smoking cessation treatment.

Clonazepam (**choice B**) is a long-acting benzodiazepine that is often used in the treatment of anxiety associated with major depression, during the initiation of antidepressant treatment. It is not associated with priapism and would not be contraindicated in this patient.

Paroxetine (**choice C**) is a selective serotonin reuptake inhibitor (SSRI) that is used for the treatment of depression and a variety of primary anxiety disorders (obsessive-compulsive disorder, panic disorder, social phobia). It is not associated with priapism.

Zolpidem (**choice E**) is a non-benzodiazepine hypnotic agent that acts at the gamma-aminobutyric acid (GABA)-benzodiazepine complex. It is used in the treatment of insomnia. It is not associated with priapism.

39.

The correct answer is E. The child described in this case description has the clinical

features characteristic of Rett syndrome. During the first five months after birth, the infant has age-appropriate motor skills, head circumference, growth, and social interactions. At six to 30 months, the child has progressive encephalopathy with decline in previously developed motor and social skills. Associated features include seizures in up to 75% of affected children and irregular respiratory patterns. Long-term receptive and expressive communication and socialization abilities remain at a developmental level of less than one year. Rett syndrome occurs almost exclusively in females.

Asperger disorder (**choice A**) is characterized by at least two of the following indications of social impairment: markedly abnormal nonverbal communicative gestures, failure to develop peer relationships, lack of social or emotional reciprocity, and an inability to express pleasure in other people's happiness. Restricted interests and patterns of behavior are also present.

Attention deficit/hyperactivity disorder (**choice B**) is characterized developmentally by an age-inappropriate poor attention span, age-inappropriate features of hyperactivity and impulsivity, or both.

Autistic disorder (**choice C**) is characterized by impairments in social interactions and communication, and restricted repetitive and stereotyped patterns of behavior, interests, and activities. Onset is prior to the age of three years.

Pervasive developmental disorder, not otherwise specified (**choice D**) is a diagnostic category that would be used when a child manifests a qualitative impairment in the development of reciprocal social interaction and communication but does not meet the criteria for other pervasive developmental disorders.

40.

The correct answer is A. Exposure therapy, a type of behavior therapy, is the most commonly used treatment of specific phobia. The therapist usually desensitizes the patient by a gradual exposure to the phobic stimulus. Relaxation and breathing control are important parts of the treatment.

Hypnosis (**choice B**) is used to enhance the therapist's suggestions that the phobic object is not dangerous. At times, self-hypnosis can be taught so that the patient uses it as a method of relaxation when confronted with the phobic stimulus.

Insight-oriented psychotherapy (**choice C**) was initially used to treat phobias, but analyzing unconscious conflicts didn't resolve phobic symptoms. It does help the patient understand the origins of the phobia and how to deal with anxiety-provoking stimuli.

Medication (**choice D**) is used in the treatment of a specific phobia only if it is associated with panic attacks and generalized anxiety. The pharmacologic treatment is then directed toward the panic attacks.

Supportive therapy (**choice E**) may be used in helping the patient actively confront the phobic stimulus during treatment. It is usually used in addition to an ongoing treatment.

41.

The correct answer is C. *N*-acetylcysteine should be administered immediately when ingestion is suspected, as the longer the delay in treatment particularly if longer than 6 to 8 hours the worse the clinical outcome. Acetaminophen levels may not be immediately detectable and absorption may be delayed by codeine or other opioids in combined painkillers. The efficacy of gastric lavage diminishes after 2 to 4 hours, the time for complete gastric emptying. However, the codeine may delay gut motility, making this emergency room physician's choice to initiate lavage quite reasonable.

Admission to a monitored unit (**choice A**) may be reasonable. However, the first step is not to transfer the patient but to give *N*-acetylcysteine. Cardiac monitoring, however, is not essential. These patients are at risk for liver failure, not arrhythmias.

Hemodialysis (**choice B**) does not work on acetaminophen overdose, although it is quite effective for treating aspirin overdose, another common over-the-counter medication used in suicide attempts.

The acetaminophen level should be checked in 2 hours (**choice D**). Treatment, however, should be started now.

Naloxone (**choice E**) should be given as needed. If this patient were stuporous, with pinpoint pupils and a depressed respiratory drive, naloxone would be appropriate.

42.

The correct answer is A. Influenza pneumonia during pregnancy can be a severe illness. Normally "the flu" is a self-limited illness that lasts 3-4 days and produces few major sequelae. However, patients with influenza pneumonia during pregnancy can develop high fever, malaise, cough, and headache. In some cases a bacterial superinfection will occur (often with *Staphylococcus aureus*), which can lead to peribronchial infiltrates, cavitation, and a pleural effusion. Current recommendations are that pregnant women who will be in the second or third trimester during the flu epidemic season should be given the influenza vaccination. Also, pregnant women with significant medical problems should be given the vaccination before the influenza season, regardless of trimester.

The measles (**choice B**), mumps (**choice C**), and rubella (**choice D**) vaccines are live attenuated vaccines. Their use during pregnancy is contraindicated.

The varicella (**choice E**) vaccination is used to prevent chickenpox. It is a live-virus vaccine; therefore, its use during pregnancy is also contraindicated.

43.

The correct answer is B. Once patients reach 42 completed weeks of gestation, many physicians will induce labor for post-term pregnancy. This is done to avoid the uncommon but catastrophic outcome of fetal demise and the higher rates of placental insufficiency that develop as patients get further post-term. Prostaglandin (PGE₂) gel is an effective agent to use for labor induction. It has been shown to improve the Bishop's score, to shorten the length of labor and delivery, to decrease the amount of oxytocin needed, and to decrease the cesarean delivery rate. The main complication from its use is uterine hyperstimulation. This hyperstimulation is defined as an increased frequency of contractions (greater than 5 every 10 minutes) or an increased length of each contraction (greater than 2 minutes) with evidence of fetal distress. When this hyperstimulation occurs, the patient may be treated with IV or subcutaneous terbutaline. This medication usually has a rapid onset of action in resolving hyperstimulation. IV magnesium sulfate can also be used.

To administer general anesthesia (**choice A**) would be incorrect. There are occasions in which the fetal heart rate tracing rapidly deteriorates and emergency cesarean delivery is needed. On these occasions, it may be necessary to administer general anesthesia to the mother during the cesarean. In this case, however, more conservative measures should be tried prior to cesarean delivery.

To perform amnioinfusion (**choice C**) would be incorrect. Amnioinfusion can be used when a patient has ruptured membranes and decelerations of the fetal heart rate or

thickened meconium. It is not used with intact membranes.

To start oxytocin (**choice D**) would be contraindicated. Oxytocin is known to cause uterine hyperstimulation, as is prostaglandin (PGE₂) gel. Oxytocin would not be given to a patient in the midst of uterine hyperstimulation.

44.

The correct answer is A. Hyperprolactinemia is the cause in approximately 10 to 20% of cases of amenorrhea. It is known that elevated prolactin levels alter the hypothalamic-pituitary-ovarian axis such that ovulation is suppressed and menses do not occur. This patient has amenorrhea, galactorrhea (i.e., a milky discharge from the breasts), and an elevated prolactin level. All of these findings are consistent with hyperprolactinemia, likely coming from a pituitary microadenoma. The fact that no mass is seen on the head MRI is also consistent with a pituitary microadenoma, as small microadenomas may not be visualized. The treatment of choice for this patient is with bromocriptine. Bromocriptine is a dopamine agonist that has been shown to decrease prolactin levels and bring about a return of ovulation and menses. The re-establishment of ovulation is especially important for this patient who wishes to conceive.

Dicloxacillin (**choice B**) is often used to treat a breast infection, which can occur in a nursing mother. This patient, however, does not have findings consistent with breast infection. Rather, the nipple discharge is secondary to the patient's elevated prolactin levels.

Magnesium sulfate (**choice C**) is used in obstetrics to prevent seizures in patients with pre-eclampsia and to stop the uterus from contracting in patients with preterm labor. It is not indicated for the treatment of hyperprolactinemia.

The oral contraceptive pill (**choice D**) would not be appropriate as this is a young woman who wishes to become pregnant. If she did not desire pregnancy, the oral contraceptive pill would be appropriate therapy. One of the major concerns in young women with microadenomas is that decreased levels of estrogen will lead to bone loss and the eventual development of osteoporosis.

45.

The correct answer is E. This patient has a presentation and findings that are most consistent with a benign cystic teratoma (dermoid). Dermoids are a type of ovarian germ cell tumor. Germ cell tumors are the most common type of ovarian neoplasm in females under the age of 20 and dermoids are the most common benign ovarian neoplasm. Dermoids can range in size from small masses that are noted incidentally on ultrasound and cause no symptoms, to large cysts that cause pain and pressure, as this patient has. Laparotomy is the most appropriate next step in the management of this patient because, as adnexal masses enlarge--especially when they become greater than 5 cm--the risk of ovarian torsion increases. Thus, laparotomy with removal of the dermoid is indicated to prevent torsion. Also, this patient's mass is causing her symptoms of pain and pressure and, on that basis, should be removed. Finally, while the mass most likely is a dermoid, this is not certain without pathologic diagnosis and, therefore, the cyst should be removed and evaluated by a pathologist. At the time of surgery, close examination should be made of the other ovary because dermoids may be found bilaterally in more than 10% of cases.

To repeat pelvic examination in 1 year (**choice A**) would not be correct management. This patient is symptomatic with a 6 cm ovarian mass that is at risk for torsion. She

should, therefore, be managed surgically.

To repeat pelvic ultrasound in 6 weeks **choice B** is appropriate for some adnexal masses. For example, in a young woman with a small complex cyst that appears consistent with a corpus luteum, it may be most prudent to recheck an ultrasound in 6 weeks to see if the cyst has resolved. This patient, however, is symptomatic with a 6 cm cyst that appears to be a dermoid, which will not resolve spontaneously. She, therefore, requires surgery.

46.

The correct answer is A. Pelvic floor dysfunction should be suspected in multiparous women. This patient has pelvic floor dysfunction, which can cause urinary incontinence, fecal incontinence, or constipation, depending on the nature of the anatomic or physiologic lesion. In pelvic floor dyssynergia, the puborectalis and external anal sphincter inappropriately contract or fail to relax as rectal pressure increases. This can be confirmed by anorectal manometry.

Autonomic neuropathy (**choice B**) is common in patients with diabetes and can manifest as diabetic gastroparesis. This condition can present with nausea, severe gastrointestinal reflux symptoms, or constipation. Gestational diabetes places patients at risk for diabetes, but it is unlikely that this patient has autonomic neuropathy, which develops after decades of diabetes.

Functional or idiopathic constipation (**choice C**), such as irritable bowel syndrome, should be diagnosed only after other reasonable diagnoses have been excluded. The recent worsening of her symptoms, which correlate with the birth of her last child, and her occasional urinary incontinence suggest pelvic floor dysfunction.

Colon cancer, either left- or right-sided (**choices D and E**), is unlikely in a woman this age. The uncle who had colon cancer at age 65 years does not place this woman at increased risk for colon cancer in her thirties.

47.

The correct answer is D. In both men and women, growth of axillary and pubic hair is stimulated by androgens. Androgens are also responsible for libido, which is absent in this girl. The rest of the presentation suggests the presence of a tumor in the pituitary area, from which some degree of hypopituitarism has ensued.

Excessive production of androgens (**choice A**) in a female would have resulted in virilization.

Excessive production of estrogens (**choice B**) would produce feminization in a male (gynecomastia, for instance).

High levels of prolactin (**choice C**) would produce galactorrhea and amenorrhea in a young lady.

Insufficient production of estrogens (**choice E**) would have inhibited the development of her breasts.

48.

The correct answer is A. In this instance, an extremely stressful event has been followed by localized loss of memory or amnesia of circumstances surrounding the event, making the diagnosis of dissociative amnesia the most likely diagnosis.

Dissociative amnesia is often accompanied by nightmares and anxiety concerning the event, both of which this patient also has.

Dissociative fugue (**choice B**) is a disturbance of identity that requires a sudden,

unexpected travel away from home or one's place of work, with inability to recall one's past.

Dissociative identity disorder (**choice C**) is also a disturbance of identity. It requires the presence of two or more distinct identities or personality states, which recurrently take control of the person's behavior. This is popularly known as multiple personality disorder.

Factitious disorder (**choice D**) is a diagnosis requiring intentional production of symptoms and gratification from assuming the sick role.

Transient global hypoxia (**choice E**) is not a likely diagnosis given this patient's lack of altered consciousness following this event.

49.

The correct answer is C. Tamoxifen is a nonsteroidal agent with both pro- and antiestrogenic properties. It was first approved in 1977 by the U.S. Food and Drug Administration for use in postmenopausal women with advanced breast cancer. Since that time, it has been approved for many other uses related to breast cancer: as adjuvant therapy in postmenopausal women with resected node-positive disease, in postmenopausal women with metastatic breast cancer, and as adjuvant therapy in women (pre- and postmenopausal) with resected node-negative disease. Recently, much attention has been focused on its use for breast cancer prevention. There is evidence that women at high risk for the development of breast cancer may reduce their risk by taking tamoxifen. However, although tamoxifen appears to be antiestrogenic at the level of the breast, it appears to act in a proestrogenic fashion at the level of the endometrium. Many women on tamoxifen will develop endometrial changes, including polyp formation, hyperplasia, and frank invasive carcinoma. Thus, women on tamoxifen need to be followed carefully, and prompt evaluation of abnormal vaginal bleeding should be conducted.

Tamoxifen is used to prevent breast cancer (**choice A**).

Tamoxifen, like estrogen, has been shown to lower blood levels of LDL cholesterol (**choice B**).

Women on tamoxifen appear to be at no greater risk, and may be at a lower risk, for the development of myocardial infarction (**choice D**).

Tamoxifen, like estrogen, has been shown to increase bone density and to reduce the likelihood of development of osteoporosis (**choice E**).

50.

The correct answer is A. This boy most likely has familial short stature (FSS). Children with FSS usually have a normal birth weight and length. At the age of 2-3 years, however, their growth begins to decelerate and drops to around the 5th percentile. The onset and progression of puberty in children with FSS are normal. Bone age is typically consistent with the chronologic age.

A decreased complement C3 level (**choice B**) may suggest chronic inflammatory disorders. But, the lack of any signs or symptoms makes any chronic inflammatory disorder unlikely.

A decreased serum albumin concentration (**choice C**) can be secondary to a variety of conditions, such as nephrotic syndrome and malnutrition. But, the lack of supportive history and physical examination data makes these conditions unlikely.

Decreased thyroid stimulating hormone (**choice D**) suggests hyperthyroidism as the etiology of the boy's short stature, but it is highly unlikely in this case.

An increased serum creatinine level (**choice E**) indicates renal failure, but this is inconsistent with the child's history and physical examination.

51.

The correct answer is A. All of the listed choices are appropriate interventions.

However, in this patient, the first treatment should be to administer epinephrine (**choice A**), as the patient shows signs of anaphylactic shock. Epinephrine can reverse both the hypotension and the bronchospasm in this patient. Among patients with anaphylaxis, fatality rates are highest among patients whose treatment with epinephrine is delayed.

Fluid boluses (**choice B**) are likely to be needed. However, first give epinephrine, as the major pathophysiologic problem is poor vascular tone. Additionally, the patient should not be bolused with hypotonic fluids, such as half-normal saline, but should receive normal saline or lactated Ringer's solution.

Endotracheal intubation (**choice C**) is the first priority if there is significant angioedema or stridor. However, this patient does not need to be intubated at this time, although close monitoring is warranted. First give epinephrine, which will reverse her bronchospasm and treat her wheezing.

Diphenhydramine and cimetidine (**choice D**) will block histamine, a key mediator in anaphylaxis. However, both have a delayed effect. The first priority is making sure this patient receives epinephrine.

Intravenous corticosteroids (**choice E**) will prevent the late phase reaction from IgE-mediated histamine release. However, this is a delayed effect. Although the steroids should be provided in an expedient manner, the first treatment should be epinephrine.

52.

The correct answer is D. As part of the widely accepted University of Pittsburgh criteria for brain death, the presence of either posturing or brain stem function (e.g., pupillary reflexes or corneal reflexes), as are present in this case, violates the brain criteria for the formal definition of brain death. That said, the fact that the patient has no purposeful activity one week after an anoxic brain injury bodes poorly for a meaningful neurological recovery. There are published studies that stratify long-term prognosis of such patients based upon neurological examinations made in the first 48 hours after injury.

While the passing of time often aids in the prognosticating of likely neurological recovery, the diagnosis of brain death can be made at any time and is not time-dependent (**choice A**).

Neurologists are often asked to help predict neurologic recovery and diagnose brain death (**choice B**), but any physician (generally two are required) may do so within current accepted diagnostic guidelines.

MRI (**choice C**) may help assess the extent of brain injury but has no role in the formal diagnosis of brain death.

The EEG, even when suggestive of minimal or no cortical function (**choice E**), does not exclude brainstem activity and therefore can not be used in isolation to make the diagnosis of brain death.

53.

The correct answer is C. This patient has an acute exacerbation of her chronic obstructive pulmonary disease (COPD). On the basis of the history and physical examination, she would be expected to have a compensated respiratory acidosis because of CO₂ retention. Furthermore, she would be expected to have evidence of mild

hypoxia.

With respirations of 24/min, which is high but not extremely high, she would not be expected to show the severe levels of acidosis and CO₂ retention illustrated in **choice A**. The patient is sufficiently symptomatic that the near normal pO₂ of 94 seen in **choice B** would be unlikely.

Although asthmatics may present during an acute exacerbation with a respiratory alkalosis (**choices D and E**), in a patient with underlying COPD, there is usually a baseline respiratory acidosis.

54.

The correct answer is C. AIDS and other profoundly immunosuppressed patients are vulnerable to infectious esophagitis. Endoscopy with biopsy is usually used to identify the causative agents, which are typically *Candida*, Herpes simplex, or cytomegalovirus. Careful review of the biopsy material is warranted, since these patients may actually be infected by more than one agent. In the case of cytomegalovirus infection, the distinctive histological finding is the presence of small numbers of cells with markedly enlarged nuclei, which on careful observation may show both cytoplasmic and nuclear viral inclusions. Cytomegalovirus infection can be treated with ganciclovir.

Acid reflux (**choice A**) could produce inflammation, but would not alter nuclear morphology.

Candida (**choice B**) would have hyphal and yeast forms.

Herpes simplex (**choice D**) causes multinucleated cells with nuclear viral inclusions.

Herpes zoster (**choice E**) infection would resemble herpes simplex infection, but it much less commonly involves the esophagus.

55.

The correct answer is B. Pigmented, mole-like lesions are very common in the general population, and virtually every physician needs to develop some skill in distinguishing obviously benign lesions from potentially malignant ones. Dysplastic nevi are an intermediate category between obviously benign nevi and malignant melanoma. While not considered cancerous themselves, they do have an increased rate of progression to melanoma. Isolated dysplastic nevi are often excised to remove the melanoma risk. More problematic are cases like this one, in which large numbers of dysplastic nevi are present. In this situation, careful monitoring with serial photographs can identify any changing lesions which may be undergoing malignant transformation.

Compound nevi (**choice A**) are usually dark, typically elevated, 3 to 6 mm lesions with a very regular shape; most people have about 10 of these lesions.

Halo nevi (**choice C**) are flesh-colored or dark nodules, usually 3 to 5 mm, surrounded by a ring of depigmented skin.

Lentigos (**choice D**) are flat, sharply marginated, uniformly pigmented, 2 to 4 mm diameter skin lesions.

Malignant melanomas (**choice E**) are often quite asymmetrical in shape and the color varies more widely than in dysplastic nevi, potentially showing tan, brown, black, blue, red, or clear areas.

56.

The correct answer is D. The most likely diagnosis is primary (essential) thrombocythemia. The condition is due to a clonal abnormality of a multipotent hematopoietic cell that produces megakaryocytic hyperplasia with resultant increased

platelet count. Since the platelets are often abnormal, either a thrombotic or a hemorrhagic tendency may be seen. The platelet count may be as low as 500,000/ μ L or greater than 1,000,000/ μ L. The clinical presentation and laboratory findings illustrated in the question stem are typical. The other choices listed commonly must be excluded before a diagnosis of primary thrombocythemia is confirmed.

Chronic myelogenous leukemia (**choice A**) can be a cause of increased platelet count, but the absence of either a Philadelphia chromosome or a markedly increased white count argues against this possibility.

Myelofibrosis (**choice B**) can also cause thrombocythemia, but would likely show some abnormally shaped (often tear drops) red cells.

Polycythemia vera (**choice C**) can also cause thrombocythemia, but would be associated with an increased red cell mass.

Secondary thrombocythemia (**choice E**) is a reactive process that may occur in a variety of settings including chronic inflammatory disorders, acute infection, hemorrhage or hemolysis, tumors, iron deficiency, or splenectomy. Abnormal platelet forms are not usually seen on smears from these patients and platelet function tests are usually normal.

57.

The correct answer is B. Hirschsprung disease or congenital aganglionic megacolon is caused by a congenital absence of the ganglion cells of both the Meissner and Auerbach plexuses. It is the most common cause of lower intestinal obstruction in the neonatal period. In early childhood it may present as chronic constipation with intermittent fecal soiling. It occurs predominantly in males and there is an increased family incidence. Surgical treatment is indicated, but the diagnosis is confirmed by a suction biopsy that can be easily performed without general anesthesia. The biopsy would reveal an absence of ganglion cells in the submucosal and myenteric plexuses.

A stool culture (**choice A**) would be performed if one was entertaining a bacterial cause of gastroenteritis, especially in a hospitalized patient. However vomiting, diarrhea, and abdominal distention in a newborn are unlikely to be caused by gastroenteritis, especially in this case with a tight sphincter noted on rectal examination.

A barium enema (**choice C**) may be indicated in suspected cases of Hirschsprung disease, but it is the biopsy that makes the diagnosis. The barium enema in this case of Hirschsprung disease revealed a dilated proximal bowel with evidence of a contracted distal rectum.

An alpha₁-antitrypsin level (**choice D**) would not be indicated in this case. It is obtained when one suspects an alpha₁-antitrypsin deficiency. Affected infants would present with jaundice, acholic stools, and hepatomegaly.

A serum TSH (**choice E**) would be performed if a newborn infant were thought to have hypothyroidism. In the U.S., most states have mandatory newborn screening for thyroid disease. Frequently, congenital hypothyroidism is asymptomatic, but it may present with symptoms of constipation, lethargy, poor feeding, mottling, and prolonged jaundice. The typical features in this case are more suggestive of Hirschsprung disease.

58.

The correct answer is B. Cimetidine is the only drug listed known to cause psychiatric effects of clinical significance, including delusions and psychosis. The exact mechanism is unknown, but is thought to be related to the effects of cimetidine on the H-2 histamine

receptor in the brain. The treatment of cimetidine-induced psychosis is to reduce the dosage of the medication, and discontinue it if an alternative therapy is available. Aspirin (**choice A**) has not been shown to induce any clinically significant psychiatric changes.

Digoxin (**choice C**) has been known to cause delirium in toxic levels, but does not cause the long standing delusion that the patient above demonstrates.

Docusate sodium (**choice D**) and ibuprofen (**choice E**) are medications not known to cause demonstrable psychiatric effects.

59.

The correct answer is B. This patient has an obstetrical history that is consistent with abnormal cervical competence. This diagnosis may be made when the patient has a history of painless cervical dilation in the second trimester. Cervical incompetence is a cause of second-trimester pregnancy loss and preterm delivery. Cervical incompetence may be congenital and/or acquired. Women who have had previous trauma to the cervix (e.g. dilation of the cervix, cervical conization, or obstetric trauma) and women with mullerian anomalies, or a history of in-utero exposure to diethylstilbestrol may be at increased risk. This patient, given her history, was offered a cerclage. Cerclage is a procedure in which a suture is placed at the level of the internal os after bladder dissection (Shirodkar) or as high up on the cervix as possible (McDonald). A prophylactic cerclage is placed between 12 and 16 weeks' gestation. Once the cerclage is placed, the patient should not engage in sexual intercourse, prolonged standing, or heavy lifting. This patient, however, refused to have a cerclage placed. Given her history, however, she needs to be followed closely to ensure that any signs of cervical incompetence are detected as soon as possible. Regular examinations of the cervix, either digitally or with ultrasound, should begin at 16 weeks because cervical incompetence becomes a concern during the second trimester.

Starting regular examinations at 10 weeks (**choice A**) is unlikely to be helpful. Cervical incompetence most often manifests itself in the second or third trimester.

Starting regular examinations at 22 weeks (**choice C**) or 28 weeks (**choice D**) would not be correct, as these gestational ages may be too late to detect cervical changes. This patient lost her last pregnancy at 22 weeks, which means that her cervix may have started changing several weeks earlier. To wait until 22 or 28 weeks would risk missing cervical changes and the possibility of instituting changes (e.g., bed rest, hospitalization, or cerclage placement) to help prevent pregnancy loss.

60.

The correct answer is A. This 64-year-old man has a minimal smoking history and yet has very severe emphysema. He also has a history of elevated transaminases. The condition described here, alpha-1-antitrypsin deficiency, is the only disease among the choices that will affect both organ systems. They do not need to coexist in the same patient, but there will typically be a family history of either emphysema in nonsmokers, or liver function test abnormalities without other obvious cause. There is also a family history present of precocious emphysema in his younger brother, and this suggests a familial disease pattern.

Primary hemochromatosis (**choice B**) is a genetically transmitted disease with an autosomal recessive pattern that leaves iron deposition in the liver, heart, and pancreas. It leads to a bronze hyperpigmentation of the skin.

Primary sclerosing cholangitis (**choice C**) occurs in patients with ulcerative colitis and does not affect the lungs.

Secondary hemochromatosis (**choice D**) occurs in patients who have received massive amounts of blood transfusion over the years.

Wilson disease (**choice E**) is a familial genetic liver disease that also involves the eye (Kayser-Fleischer ring) and causes neuropsychiatric disorders. There is no lung involvement in Wilson disease.

61.

The correct answer is B. This is a patient of Mediterranean descent who has a microcytic anemia with an appropriate reticulocyte count, normal iron studies and a hemoglobin electrophoresis with an increased alpha chain component of hemoglobin. The thalassemias are a heterogeneous group of inherited disorders characterized by the underproduction of either the alpha or the beta chains of the hemoglobin molecule. In beta thalassemia, a reduced production of beta chains occurs with normal amounts of alpha production.

Alpha-thalassemia (**choice A**) is not supported by the presence of alpha chains on the electrophoretic pattern.

Iron deficiency anemia (**choice C**) although a microcytic anemia, is not supported by the normal iron studies.

Megaloblastic anemia (**choice D**) is not supported by the low MCV.

62.

The correct answer is D. This patient has developed gonococcal conjunctivitis and requires treatment with systemic ceftriaxone. Conjunctivitis is an inflammatory response of the conjunctival vessels to a variety of insults. Chemical conjunctivitis caused by instillation of silver nitrate drops is the most common cause of conjunctivitis in the first 24 hours of life. *Neisseria gonorrhoea* and *Chlamydia trachomatis* are common infectious causes acquired by passage through the birth canal. Chlamydia is the most common cause of infectious ophthalmia neonatorum. Bacteria, such as *Haemophilus*, *Streptococcus*, *Staphylococcus*, or *Pneumococcus*, usually cause acute purulent conjunctivitis. Viruses (adenovirus, enterovirus) may cause an isolated conjunctivitis or one associated with an exanthem. Allergic conjunctivitis usually develops after the neonatal period. Clinically, tearing, conjunctival injection, lid edema, and discharge are characteristic. Pain, photophobia, and decreased vision are rare and indicate corneal disease. Gonococcal conjunctivitis is purulent and can occur at birth or after 5 days of age if the patient has received topical antibiotic prophylaxis at birth. Chlamydial conjunctivitis typically appears about 5 to 23 days after birth. As for the diagnosis, Gram stain and culture may help identify an infectious cause. The treatment of choice for gonococcal conjunctivitis is systemic ceftriaxone, 30 to 50 mg/kg/day in divided doses intravenously, or intramuscularly up to a maximum of 125 mg/day. Neonates with gonococcal ophthalmia should have their eyes frequently irrigated with saline to eliminate the discharge.

63.

The correct answer is D. The rhythm described on the ECG is multifocal atrial tachycardia. This is characterized by variable P wave morphology and PR and RR intervals. The control of this tachycardia comes with improved ventilation and oxygenation. This condition is associated with severe pulmonary disease.

Digitalis (**choice A**) enhances myocardial contractility primarily by inhibiting membrane sodium-potassium ATPase. It is useful in the management of heart failure and rate control in atrial fibrillation.

Warfarin (**choice B**) is used in the management of chronic atrial fibrillation to reduce the risk of an embolic stroke. In the acute setting, however, it has little utility in rate control or anticoagulation.

Electrical cardioversion (**choice C**) is of little benefit in rhythm or rate control in multifocal atrial tachycardia. It is indicated in the conversion of atrial fibrillation to a normal sinus rhythm. In those situations, the patient must first be adequately anticoagulated.

Defibrillation (**choice E**) is indicated in patients with arrhythmia who are hemodynamically unstable. Typically, it is indicated in ventricular fibrillation and tachycardia. If a patient has syncope as a result of such tachyarrhythmias, then an internal defibrillator must be installed.

64.

The correct answer is B. This patient has *Legionella* pneumonia, which is caused by *L. pneumophila*, a gram-negative bacillus. It is often acquired from a contaminated water supply (air conditioning systems) and can lead to outbreaks. The major risk factors include advanced age, immunosuppression, chronic lung disease, and cigarette smoking. Clinical features include a cough, chest pain, diarrhea, high fever, malaise, fatigue, and anorexia. A chest x-ray film reveals infiltrates and later, consolidation. Gram's stain reveals leukocytes, but no organisms. Cultures on BCYE agar grow the organism. The treatment is erythromycin.

Chlamydia trachomatis (**choice A**) is an obligate intracellular parasite that has features similar to gram-negative bacteria. Pneumonia develops in neonates born to mothers infected with the bacteria.

Moraxella catarrhalis (**choice C**) is a gram-negative coccus that causes pneumonia in elderly patients with COPD. Clinical features include low-grade fever, chills, chest pain, and malaise. Gram's stain reveals the organism. Penicillin/clavulanic acid is the treatment.

Mycoplasma pneumoniae (**choice D**) causes a community-acquired pneumonia that occurs in young adults. The features are a sore throat, nonproductive cough, and headache. Chest x-ray films show interstitial infiltrates. Treatment is with erythromycin.

Pneumocystis carinii (**choice E**) is an opportunistic pathogen that causes pneumonia in immunocompromised hosts. Diffuse interstitial infiltrates are found on chest x-ray films. Diagnosis is made by examination of sputum or bronchial washings. The treatment is trimethoprim-sulfamethoxazole.

65.

The correct answer is C. This patient has gout. Pain in the metatarsal phalangeal joint of his first toe is a classic symptom and is known as podagra. Common comorbid conditions include diabetes, hypertension, coronary artery disease, hyperlipidemia, and alcohol abuse. The first-line treatment for gout is nonsteroidal antiinflammatory drugs. Indomethacin is commonly used, although any nonsteroidal antiinflammatory drug is appropriate.

Allopurinol (**choice A**) is a medicine used to prevent gout recurrences by decreasing uric acid levels. It should be used in patients who have recurrent gouty attacks. The important learning point about allopurinol is that it has no role in the patient with an acute

gouty flare because it can actually increase gouty pain by mobilizing uric acid in an already inflamed joint.

Dietary modifications (**choice B**) are important for the long-term prevention of recurrences but do not have a role in the treatment of the acute gouty flare. Our first responsibility in this patient is to control his severe pain. Excessive dietary purine intake together with alcohol may contribute to elevated uric acid levels seen in gout. Patients with gout should be encouraged to modify their diets to minimize secondary causes of hyperuricemia.

Prednisone (**choice D**) is an effective medication in the treatment of the acute gouty flare, but because of its side effects it is not used as first-line therapy. It is useful in patients who cannot tolerate nonsteroidal antiinflammatory drugs because of gastrointestinal upset or renal failure. Intra-articular steroids are another good option for patients who cannot tolerate nonsteroidal antiinflammatory drugs, and they have the advantage of not subjecting the patient to the systemic effects of steroids.

66.

The correct answer is D. Tangier disease is a rare familial disorder characterized by alpha-lipoprotein deficiency, which leads to very low high-density lipoprotein (HDL), recurrent polyneuropathy, lymphadenopathy, and hepatosplenomegaly due to storage of cholesterol esters in reticuloendothelial cells. Although you may never see this disease, the association of orange-yellow tonsillar hyperplasia (due to the cholesterol ester deposition there as well) with Tangier disease is a sufficiently distinctive clue in physical diagnosis to be worth remembering. (One rule of thumb in medicine is that although you will probably never see most of the very rare diseases, you will almost certainly see some of them.)

The presence of multiple angiokeratomas (**choice A**) on the lower half of the body suggests Fabry disease.

Grey-brown pigmentation of the forehead, hands, and pre-tibial region (**choice B**) suggests Gaucher disease.

Irregular black deposits of clumped pigment in the peripheral retina (**choice C**) are characteristic of retinitis pigmentosa, which may occur in association with abetalipoproteinemia and Refsum disease.

Pingueculae (**choice E**) could suggest Gaucher disease, but this could also occur in normal adults.

67.

The correct answer is B. Shaken infant syndrome was defined by pediatric radiologist John Caffey in 1972. Approximately 1-2 million cases of child abuse and neglect are reported every year, and nearly 2000 children die from abuse and neglect annually. A normal platelet count and coagulation studies eliminate the misdiagnosis of bleeding abnormalities.

An ammonia level (**choice A**), lipid panel (**choice C**), thyroid studies (**choice D**) or urine electrolytes (**choice E**) are not indicated.

68.

The correct answer is C. The patient is experiencing a normal grief reaction. Note that this can include symptoms such as feeling as if one is hearing his or her name called by the departed. Additionally, the patient appears to be improving with respect to neurovegetative symptoms. Beyond grief counseling at his own request, there is no

further intervention that is absolutely necessary at this time.

The patient's hearing his name called in the context of the passing of his wife is not suspicious for seizure activity. An electroencephalogram (**choice A**) would be of little value at this time.

The patient's hearing his name called by his wife is not to be taken as a symptom of a psychotic episode in the context of the improvement in grief symptoms that the patient is experiencing. The patient does not require antipsychotic treatment (**choice B**) at this time.

The patient's straightforward presentation and lack of imminent need for extensive mental health services make neuropsychologic testing (**choice D**) unnecessary at this time.

The patient's symptoms constitute a normal grief reaction. There is no evidence of a decline in function, suicide risk, or real psychotic symptoms. Therefore, an emergency evaluation (**choice E**) is not necessary.

69.

The correct answer is B. Corticosteroids are known to lead to more difficult glucose control in diabetic women. To ensure that these patients do not develop diabetic ketoacidosis, blood glucose levels should be checked regularly, and elevated values treated with insulin. This will often require hospitalization, which is usually required by the condition for which they received the corticosteroids in the first place (e.g., preterm labor or preterm premature rupture of membranes). In patients who do not have diabetes, the hyperglycemic effect will last 2-3 days.

Studies have been performed to determine whether antenatal treatment with corticosteroids leads to decreased childhood intelligence (**choice A**). There is no evidence that this relationship exists.

Because of the immunosuppressive properties of corticosteroids, there has been concern that their use may increase rates of maternal infection (**choice C**) or neonatal infection (**choice E**). There is no definitive proof that corticosteroid use leads to higher rates of infection in either the mother or fetus. And, although there may be some instances of maternal or neonatal infection in some cases of corticosteroid administration, the increased maternal insulin requirement occurs almost without exception.

Neonatal adrenal suppression (**choice D**) has not been proven to result from antenatal corticosteroid administration.

70.

The correct answer is D. Uterine atony is the most common cause of postpartum bleeding. Prolonged labor and large children (or twins) increases the likelihood of uterine atony. Treatment is fundal massage and, if ineffective, initiation of uterotonic agents, such as oxytocin, intramuscular Methergine, carboprost tromethamine, and misoprostol. Inversion of the uterine fundus (**choice A**) is a rare cause of uterine bleeding. Given this patient's risk factors and knowing that such conditions are common, uterine atony is a much more likely finding.

Mucosal bleeding and bruising (**choice B**) might indicate disseminated intravascular coagulation, such as can occur from retained placental products. There is no reason to suspect such a scenario.

Uterine rupture (**choice C**) is rare and usually is seen in patients who have had a

previous cesarean section.

Cervical or vaginal lacerations (**choice E**) are common but less likely in a patient with previous deliveries. Given this patient's response to oxytocin and fundal massage, uterine atony is a more likely finding.

71.

The correct answer is D. Throwing a ball overhead is most consistent with the motor development of a 24-month-old child. As the cerebral cortex develops, the child's hands and arms are more able to perform more skillful and delicate motor functions.

Building a tower (**choice A**) is a good way to assess a child's motor function. A child should be able to build a tower of two cubes by 14 months of age. By 24 months, he or she should be able to build a tower of at least 6 cubes.

A 24-month-old child will not usually be able to copy a circle (**choice B**). This ability emerges around 36 months of age.

Imitative scribbling (**choice C**) appears around 16 months; spontaneous scribbling appears around 18 months.

A child should be able to walk backward (**choice E**) by 18 months. Climbing stairs or going down stairs with support are more consistently seen at 24 months.

Two other milestones that are consistent with 24 months of age are jumping up and kicking a ball forward.

72.

The correct answer is C. This is a patient who has 3 clear risk factors for coronary artery disease (tobacco, family history and age) and based on his physical examination, likely has severe peripheral vascular disease. He has, by definition, typical chest pain, so called "new onset angina". He is a prime patient to have significant coronary disease, and thus we suspect ischemia as a cause for his pain. As a surrogate for coronary angiography, which actually shows anatomy, an exercise treadmill test allows us to detect ECG changes of ischemia with activity and thus stratify this patient as requiring intervention (such as percutaneous transluminal coronary angioplasty, or coronary artery bypass grafts), or perhaps angiography to better evaluate his anatomy.

A resting ECG (**choice A**) is appropriate, but not the most appropriate, given that he is pain-free at present and one would not expect to see any ECG changes associated with ischemia.

A cardiac echocardiogram (**choice B**) will likely be performed, given his dyspnea on exertion, but is not an appropriate test in the triaging of suspected ischemic chest pain.

In some centers a "stress-echo", specifically a dobutamine echocardiogram, is used to evaluate ischemic potential.

A non-urgent coronary angiography (**choice D**) is also inappropriate since angiography is an invasive procedure reserved for people that have had equivocal results from less invasive diagnostic procedures, or are having signs of crescendo angina. This patient has new angina, but it is "typical" angina in that it is exertional.

73.

The correct answer is B. The risk of a smoker with asbestos exposure developing lung cancer is about *75 times* greater than the regular population. He needs to be advised that he is at increased risk for lung cancer and would greatly benefit from smoking cessation.

Asbestosis (**choice A**) can lead to an interstitial lung disease in patients with or without a

smoking history. Even nonsmokers may present with dyspnea on exertion, cough, chest wall pain, and ultimately, end-stage lung disease.

Patients with asbestosis are at increased risk for pleural or peritoneal mesotheliomas (**choice C**) compared to patients without asbestos exposure. Despite this increased risk, this is not the most common cancer found in smokers with asbestos exposure. Rather, adenocarcinoma and squamous-cell carcinoma are the most common cancers seen in these patients. Small-cell cancer (**choice D**) is no more common than adenocarcinoma or squamous-cell carcinoma.

Steroids (**choice E**) have no proven beneficial role in the treatment of asbestos-associated lung disease.

74.

The correct answer is E. This patient has known hepatitis C. Features indicating that he has advanced signs of cirrhosis and portal hypertension include ascites, splenomegaly, jaundice with elevated bilirubin, hypoalbuminemia with leg edema, increased prothrombin time probably secondary to inadequate synthesis of clotting factors by the liver, and esophageal variceal bleeding. In the setting of disease this advanced, antiviral treatment for hepatitis C is without value and the patient should undergo evaluation for a liver transplant.

As discussed above, all anti-viral therapy (**choices A, B, and C**) would be futile given the advanced stage of his cirrhosis at this point.

A mesocaval shunt (**choice D**) is a surgical procedure whereby portal flow is diverted from the superior mesenteric vein into the inferior vena cava to reduce portal pressures. It is often complicated by encephalopathy and does not improve the underlying liver dysfunction.

75.

The correct answer is A. This girl has the typical clinical picture of impetigo contagiosa, a staphylococcal, streptococcal, or combined infection characterized by discrete, thin-walled vesicles that rapidly become pustular and then rupture. Impetigo occurs more frequently on the exposed parts of the body-the face, hands, neck, and extremities-although it may appear at sites of friction as well. Over 50% of cases are due to *Staphylococcus aureus*, with the remainder being due to *Streptococcus pyogenes* or a combination of the two bacteria. Group B streptococci are associated with newborn impetigo. Impetigo is most commonly seen in early childhood and during hot, humid summers in temperate climates.

76.

The correct answer is C. This patient has the acute onset of podagra and ankle involvement as well, secondary to gout. These attacks often follow the ingestion of red meats and/or alcohol since both of these lead to transient hyperuricemia. Note that this patient has a normal serum uric acid, which may occur during an acute attack; however, all patients with gout have some history of hyperuricemia. The appropriate evaluation of an edematous joint is an arthrocentesis. Examination of the fluid under polarizing light microscopy in a case such as this will reveal the typical uric acid crystals, which appear as negatively birefringent needle-shaped crystals (compare with **choice E**) under polarizing light microscopy.

Although it is always important to consider the possibility of an infectious arthritis, this patient has no risk factors for either gram-negative bacteremias or arthritis (**choice A**).

The gram-positive cocci in pairs and chains (**choice B**) are describing *Streptococcus*, but this organism does not cause joint involvement.

Rhomboidal crystals (**choice D**), when positively birefringent, are due to calcium pyrophosphate (pseudogout crystals), and the typical history of recent ingestion of alcohol or red meat would not usually be elicited.

77.

The correct answer is E. Perimenopausal symptoms can often be vague. The first signs of estrogen deficiency may be interrupted sleep and fatigue, both of which can cause irritability and mood swings. Hot flashes often will develop after sleep disturbances, and menstrual cycles will become irregular before ceasing. Signs and symptoms that are consistent with estrogen deficiency in a woman of this age should prompt one to think of perimenopause rather than an underlying illness.

Adjustment disorder (**choice A**) is unlikely. Although there was a major change in her life recently (her son leaving home), her symptoms precede this event.

Anemia (**choice B**) should always be considered when patients are suffering from a brisk menstrual flow. However, anemia will not explain the mood swings.

There is no indication that this patient is suffering from cyclothymia (**choice C**), which can be thought of as a mild, bipolar-like condition of depression and hypomania.

This patient does not meet the diagnostic criteria for depression (**choice D**). There is no anhedonia, change in eating habits, or loss of interest in activities and friends, etc.

78.

The correct answer is B. This patient has achalasia, which is a neurogenic esophageal disorder thought to be related to a malfunction of the myenteric plexus of the esophagus.

The result is that the esophagus behaves as if it has lost the normal peristaltic mechanism. There is also an accompanying failure to relax the lower esophageal sphincter (LES) when food reaches the distal esophagus. Because of these problems, patients experience difficulty with swallowing both solids and liquids. This is in contrast to masses of the esophagus that cause luminal narrowing, in which the swallowing of liquids is at least initially relatively preserved. Manometry of patients with achalasia typically shows decreased peristalsis in the body of the esophagus with increased resting LES pressure. These patients often have nocturnal coughing because of aspiration of retained food contents within the esophagus.

Choices A and C are not typical of any condition you need to remember.

Choice D is typical of severe esophageal disease in scleroderma.

Choice E is typical of symptomatic diffuse esophageal spasm.

79.

The correct answer is B. The child's story is worrisome for shaken baby syndrome, in which the symptoms may not correlate with the physical findings. This diagnosis should be considered in any infant presenting with a dissonant history suspicious of child abuse. The child's fontanelles are full, indicative of increased intracranial pressure. A retinoscopic examination will indicate if this is indeed the case, since blurred fundi would suggest increased pressure.

A retinoscopic examination can be done faster than a head CT (**choice A**). The patient may ultimately need a head CT but the eye examination should be done first.

Ammonia levels (**choice C**) should be checked if hepatic encephalopathy is a consideration. This is a possibility if Reye syndrome is on the differential. The increased

fontanelle pressure leads to a diagnosis of trauma.

Benzodiazepines may be needed (**choice D**) if the child has a new seizure, but it is important to first determine the cause of the prior seizure.

Because the patient has increased intracranial pressure (suggested by the full fontanelles), a lumbar puncture may cause uncal herniation and should be avoided (**choice E**).

80.

The correct answer is E. Major depression with melancholic features is characterized by a depressed mood most of the time and a lack of reactivity to pleasurable stimuli during episodes. In addition, three or more of the following criteria must be met: distinct quality of depressed mood, worse depression in the morning, early morning awakening, psychomotor retardation or agitation, weight loss, and inappropriate guilt.

Catatonia (**choice A**) can be applied to any of the episodes of major depressive or bipolar disorder. In the clinical picture, the most dominant feature is motoric immobility, as evidenced by waxy flexibility, excessive motor activity, extreme negativism, peculiar voluntary movements, and echolalia or echopraxia.

Cotard's syndrome (**choice B**), or nihilistic delusional disorder, is diagnosed when patients complain of not only having lost their possessions, status, or strength but also their inner organs. It can be seen in schizophrenic or depressive episodes.

Dysthymia (**choice C**) involves depressed mood for most of the time in the past 2 years and the presence of two or more of the following: changes in appetite, changes in sleep, fatigue, low self-esteem, poor concentration, and feelings of hopelessness. The symptoms do not meet the criteria for major depressive episode.

Major depression with atypical features (**choice D**) can be applied when, in the most recent 2 weeks, there is mood reactivity (the patient brightens to positive events) and two or more of the following are present: weight gain or increased appetite, hypersomnia, leaden paralysis, and sensitivity to rejection from others.

81.

The correct answer is D. This patient is having a hypertensive emergency, given the presence of end-organ damage in the setting of hypertension. Hypertensive encephalopathy counts as an emergency. Immediate therapy before the laboratory results are known is needed. This requires immediate, but not precipitous, lowering of the blood pressure over a period of minutes to hours. Sodium nitroprusside is used since it allows titratable blood pressure reduction.

Observation (**choice A**) is not appropriate for this patient, since he has an emergent medical condition that needs to be addressed.

Diabetic ketoacidosis or sepsis (**choice B**) may present with mental status changes and an anion gap. However, the patient most likely has hypertensive encephalopathy, and this needs to be addressed first.

A beta blocker (**choice C**) may reduce the pulse but will not cause much vasodilation or arterial dilation, which is needed to immediately lower the blood pressure. It may be used as an adjunct therapy. IV labetalol may prove to be an acceptable alternative, however.

The patient has mental status changes, and meningitis is on the differential. However, management of the blood pressure should not be delayed to perform a lumbar puncture (**choice E**). Furthermore, he is afebrile, placing meningitis lower on the differential.

82.

The correct answer is A. The age, symptoms, and imaging findings are classical. Craniopharyngiomas (**choice B**) occur in the suprasellar region, and produce endocrine symptoms. Ependymomas (**choice C**) arise in the floor of the fourth ventricle, and grow to occupy its lumen. Medulloblastomas (**choice D**) arise in the cerebellar vermis and grow down into the fourth ventricle. Meningiomas (**choice E**) are extrinsic tumors that arise from the arachnoid cap cells. They are very common in adults, but are rare in children.

83.

The correct answer is C. The Swan-Ganz tracing indicates that the patient has an elevated right-sided pressure and a low-filling pressure. Cardiac output is decreased as a result of insufficient left heart filling pressures. This is due to the right ventricular infarct, which causes backing up of venous blood and decreased forward flow, producing a decrease in left ventricular filling, as indicated by the low wedge pressure. The treatment for this patient is aggressive fluid administration.

The patient had a right ventricular infarct, presumably from involvement of the right coronary artery. Balloon angioplasty (**choice A**) may ultimately be needed to correct the underlying cause of the infarction. However, the acute event has passed, and the patient must first be stabilized. She will ultimately need a cardiac catheterization.

Digoxin (**choice B**) is not needed in this patient, since she is not in left sided heart failure. Positive inotropy is not needed as much as fluid resuscitation. Also, the patient is not in atrial fibrillation.

If the patient had sustained a left ventricular infarct, she may have needed afterload reduction in the form of intraaortic balloon counterpulsation (**choice D**). Since the patient sustained a right ventricular infarct, however, afterload reduction is not as important as is preload repletion.

The patient is hypotensive. If the hypotension does not resolve with fluid repletion, then vasopressors such as norepinephrine may be needed (**choice E**). However, fluid resuscitation must be continued.

84.

The correct answer is B. This patient, who has just returned from a cruise, has developed symptoms of toxicity in association with confusion, pulmonary findings, gastrointestinal complaints, and liver function test abnormalities. This should suggest the diagnosis of Legionnaires pneumonia, which he acquired through the ventilation system on the ship. These patients may appear quite toxic, and immediate initiation of therapy is essential since diagnosis using direct fluorescent antibody assays may take several days. Sputum Gram's stain in these patients is usually unrevealing of the *Legionella pneumophila* organism.

Ceftazidime (**choice A**) and gentamicin (**choice C**) are antibiotics effective against gram-negative organisms but are not used in treatment of *Legionella*.

Vancomycin (**choice E**) is used in patients with staphylococcal infections that are resistant to the nafcillin (**choice D**) family of antibiotics. These drugs have no role in the management of Legionnaires disease.

85.

The correct answer is A. Suturing a laceration is one of the most common situations in

which sedation may be required in a child. Sedation may be classified as *conscious* or *deep*. By definition, during conscious sedation the patient is able to maintain airway patency, protective airway reflexes, and responses to physical stimuli. This level of sedation is indicated for children (or adult patients) who have not fasted prior to the procedure, or patients who do not require a deep level of sedation. Nevertheless, conscious sedation should be performed by appropriately trained personnel, and only when equipment for resuscitation measures is readily available, should the need arise. For minor surgical procedures such as suturing uncomplicated linear lacerations, administration of a short-acting or long-acting benzodiazepine (midazolam or diazepam, respectively) by the oral or rectal route provides sufficient sedation. Intravenous access is not required. Intravenous midazolam or diazepam can be used for procedures that produce more intense pain or discomfort, such as repair of complex lacerations, bone marrow aspiration, and reduction of fractures. Concomitant opioid and benzodiazepine administration (**choice B**) is used to achieve not only sedation, but also an adequate level of analgesia. The synergistic action of opioids and benzodiazepines increases the risk of respiratory depression. Intravenous propofol (**choice C**) provides rapid onset of sedation that resolves quickly once infusion is discontinued. This drug is used for procedures requiring deeper levels of sedation in appropriately fasted and stable children. Intravenous ketamine (**choice D**) is an appropriate alternative to propofol. Its most common side effect is the production of visual and auditory hallucinations (about 10% of cases).

86.

The correct answer is C. Nortriptyline is a tricyclic antidepressant, which is lethal in overdose and is the leading cause of overdose-related deaths in the psychiatric population. Overdose is associated with prolongation of the QT interval, leading to cardiac arrhythmia and death. Given this patient's history of previous overdoses, a tricyclic antidepressant would be contraindicated.

Buspirone (**choice A**) is an antidepressant medication whose pharmacologic action is not well understood, but which is not known to be lethal in high doses.

Fluoxetine (**choice B**), paroxetine (**choice D**), and sertraline (**choice E**) are all selective serotonergic reuptake inhibitors (SSRIs). These antidepressant medications are generally safe and effective and are not lethal except in extremely high doses.

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88.

The correct answer is B. This patient is demonstrating an abnormal labor pattern with

arrest of dilation. The normal pattern of labor is one of continued progression. Whether a patient is in the latent phase or the active phase, there should be a gradual progression with an increase in the amount of cervical dilation. This patient, however, has stopped dilating and has had her contractions space out considerably. An arrest of labor like this can be caused by several reasons: contractions may not be adequate; the fetus may have a malpresentation; or the maternal pelvis may not be able to accommodate the fetus. In this case it appears that the contractions are not adequate, so at this point, it would be reasonable to give intravenous oxytocin in an effort to re-establish a contraction pattern that can effect a vaginal delivery.

Expectant management (**choice A**) would not be the most appropriate next step. The patient is clearly demonstrating a dysfunctional labor pattern at this point. To "watch and wait" in the face of insufficient uterine contractions is to place the patient at risk of an even longer labor and the correspondingly higher risk of infection.

Cesarean delivery (**choice C**) would not be the most appropriate next step in management. This patient may very well need a cesarean delivery if she is truly unable to progress in labor. However, it is worth attempting a vaginal delivery in this multiparous patient who has already had two vaginal deliveries.

To attempt a forceps-assisted vaginal delivery (**choice D**) or a vacuum-assisted vaginal delivery (**choice E**) would be contraindicated. This patient's cervix is only 5 centimeters dilated. Forceps and vacuum cannot be attempted in patients unless they are fully dilated and at +2 station or lower.

89.

The correct answer is B. Acute diverticulitis is one of the very few inflammatory processes that gives acute abdominal pain in the left lower quadrant. Patients are middle-aged or older; there is fever, leukocytosis, physical findings of peritoneal irritation in the left lower quadrant, and sometimes a palpable mass. CT scan is diagnostic. Initial treatment is NPO, IV fluids, and antibiotics. Once the patient "cools down," they often require sigmoid resection.

Acute appendicitis occurs on the right side of the abdomen. Other than that, the symptoms may be similar to those of this patient (**choice A**).

Acute pancreatitis (**choice C**) should be suspected in the alcoholic patient who develops an upper acute abdomen. The classic picture has rapid onset and the pain is constant, epigastric, radiating straight through to the back, with nausea, vomiting, and retching.

Pelvic inflammatory disease (**choice D**) is seen in women of reproductive age with lower-abdominal pain. Pelvic examination usually reveals extreme pelvic tenderness and increased pain with cervical motion. Hysterectomy is a common procedure in the United States and the cervix may have been removed at that time.

Patients with renal colic typically have intermittent pain and move around a lot because they cannot get comfortable. They have costovertebral angle tenderness and may have urinary frequency and urgency (**choice E**).

90.

The correct answer is D. Henoch- Schönlein Purpura (HSP) is the most likely diagnosis. This boy has abdominal pain with guaiac-positive stools, but also has a prominent rash, mostly on his lower extremities. Other characteristic findings of HSP include hematuria and joint pains. The illness may follow an upper respiratory infection or strep throat. The rash starts out as an urticarial rash and progresses to become

petechial and purpuric. There may be a history of migratory joint pain and arthritis. Affected joints include ankles, knees, wrists, and elbows.

If the abdominal pain were described as epigastric with radiation to the back, pancreatitis (**choice A**) might have been the likely diagnosis. In children, pancreatitis is frequently associated with viral illnesses (e.g., mumps), drugs (e.g., sulfonamides), or underlying systemic disease (e.g., lupus). Although pancreatitis has been reported in association with HSP, it is not the most likely diagnosis.

Rocky Mountain spotted fever (**choice B**) is one of the most common tick-borne diseases. The typical rash of RMSF appears within a week of the tick bite. It begins on the palms, soles, and extremities and spreads centrally. Severe headache and photophobia are common complaints.

This child did not have the typical findings of nephrotic syndrome (**choice C**) including: proteinuria, edema, and oliguria. Nephrotic syndrome frequently follows an infectious illness.

91.

The correct answer is D. In a patient with cardiac complications, a selective serotonin reuptake inhibitor (SSRI), such as sertraline, has been demonstrated to be the safest and most effective medication for the treatment of clinical depression.

Methylphenidate (**choice A**) is a stimulant medication that would have the potential to increase heart rate and sympathetic tone, which would not be advantageous in the post-myocardial infarction period.

Nortriptyline (**choice B**) is a tricyclic antidepressant that has the potential for producing cardiac arrhythmias in overdose.

Phenelzine (**choice C**) is a monoamine oxidase inhibitor, which would be contraindicated with a history of recent myocardial infarction, as the ingestion of tyramine with an MAO inhibitor could induce a hypertensive crisis.

Thioridazine (**choice E**) is a low-potency dopamine antagonist antipsychotic medication that is not indicated for the treatment of depression.

92.

The correct answer is A. The decision of whether to have a termination of pregnancy is a deeply personal one. This patient has just been notified that she is pregnant with a 10-week fetus. She is unsure whether she wants to keep her pregnancy or terminate it. In this setting, the most appropriate next step is to counsel the patient regarding her options or refer the patient for counseling. In a balanced way, the patient should be fully informed of all of her options including raising the child herself, placing the child up for adoption, and abortion.

To notify the patient's parents (**choice B**) is not appropriate. Such an act would violate the patient's confidentiality. A 34-year-old woman is an adult and issues of parental notification do not apply.

To notify the patient's partner (**choice C**) is not appropriate. This notification would also violate confidentiality.

To schedule a termination of pregnancy (**choice D**) would not be appropriate. This patient has just informed the physician that she is unsure what she wants to do. To just go ahead and schedule the termination without proper counseling of the patient would not be a balanced or proper approach for the patient.

To tell the patient that she is likely to have a miscarriage (**choice E**) is inappropriate.

This patient may have a miscarriage, as might any patient with a first-trimester pregnancy. However, once an intrauterine pregnancy with fetal cardiac activity is identified, the risk of miscarriage is approximately 10%. Therefore, she is most likely not to have a miscarriage.

93.

The correct answer is C. The clear aspirate at a time when he is actively bleeding indicates a source distal to the ligament of Treitz. Colon is the most likely site, but it could be small bowel. Tagged red-cell study should provide an idea of the region where the blood is pooling and set the stage for an arteriogram if active extravasation is demonstrated.

Colonoscopy (**choice A**) might appeal to some, but as a rule endoscopy of the colon at the time of active bleeding is unrewarding because the blood comes from higher up, obscuring the view as the instrument is advanced.

Barium studies, either from above (**choice D**) or below (**choice B**), are contraindicated at the time of active bleeding. Barium gets in the way of other, more productive studies. The upper gastrointestinal is not the source of bleeding, and thus endoscopy of that area (**choice E**) will not help.

94.

The correct answer is B. Beta blockers have been shown in multiple studies to improve survival after myocardial infarction (MI) by decreasing both oxygen demand and the incidence of ventricular arrhythmia.

Angiotensin-converting enzyme (ACE) inhibitors (**choice A**), such as enalapril, have been shown to improve survival in post-MI patients who have ejection fractions less than 40%.

Digoxin (**choice C**) would be relatively contraindicated in this instance in that it would increase myocardial oxygen demand by increasing inotropy. This patient does not need positive inotropic support, as there is only minimal decrease in ejection fraction (normal 55% to 77%).

Loop diuretics (**choice D**) may help control this patient's hypertension, but they have not been shown to improve survival.

Warfarin (**choice E**) is beneficial in large anterior wall infarcts when there is a severely low ejection fraction. The drug decreases the risk of thromboembolic phenomena.

95.

The correct answer is A. The constellation of thymic hypoplasia, hypocalcemia (with tetany), abnormal facies, and congenital cardiac anomalies defines the condition known as *DiGeorge syndrome*. This results from a developmental failure of third and fourth pharyngeal pouches, which gives rise to congenital absence or anomalies of the parathyroid, thymus, lower face, and cardiac structures. Immune deficiency results from failure of T-lymphocytes to mature in the thymus. Thus, fungal and viral organisms, which are normally controlled by T-mediated mechanisms, become frequent causes of opportunistic infections. The underlying gene defect is related to 22q11 deletion, which results in two partially overlapping conditions, i.e. *DiGeorge syndrome* and *velocardiofacial syndrome*. These conditions are collectively referred to as *chromosome 22q11 deletion syndrome*.

In utero infection by human immunodeficiency virus (HIV) (**choice B**) would result in T-cell deficiency but would not be associated with congenital abnormalities.

Mutations of the autosomal gene encoding adenosine deaminase (**choice C**) represent the most common cause of the recessive form of *severe combined immunodeficiency disease* (SCID), encompassing a heterogeneous group of conditions characterized by deficiency of both T- and B-cell mechanisms. SCID may be autosomal dominant, autosomal recessive, or X-linked.

Mutations of the X-linked gene coding for a cytokine receptor subunit (**choice D**) represent the most common cause of the autosomal dominant form of SCID.

Mutation of an X-linked gene coding for a tyrosine kinase (**choice E**) is the underlying molecular mechanism leading to *X-linked agammaglobulinemia of Bruton*, a syndrome characterized by inability of pre-B cell precursors to mature into B-lymphocytes. Humoral immune deficiency thus manifests.

96.

The correct answer is E. Judgment is the ability of a patient to evaluate a particular course of action in order to determine if it is the most appropriate one within the patient's particular value system. If the patient were to say he would put the letter back in the mail or give it back to the postman, this would indicate appropriate judgment.

Abstract thinking (**choice A**) is the ability to think or perform symbolically, or to evaluate various aspects of a situation and shift between alternatives.

Cognition (**choice B**) is tested by evaluating the intellectual and perceptual levels of mental functioning.

Insight (**choice C**) is the ability to recognize the objective reality of a situation.

Intelligence (**choice D**) is the ability to learn and apply learned information to a given situation.

97.

The correct answer is A. Trisomy 21, also known as Down syndrome, occurs primarily as the result of meiotic nondisjunction. This phenomenon of meiotic nondisjunction increases with maternal age. A 33-year-old woman has a risk for having a newborn with Down syndrome of approximately 1 in 625. This risk increases to 1 in 106 at age 40 years and 1 in 30 by age 45 years. Down syndrome is characterized by mental retardation, muscular hypotonia, short stature, flat nasal bridge, oblique orbital fissures, and other distinctive characteristics. Also, congenital heart disease, especially endocardial cushion defects, and duodenal atresia are more common among newborns with Down syndrome.

Mosaicism (**choice B**) describes the condition in which one individual has two or more distinct cell lines. This is not the most likely cause of Down syndrome.

Polyploidy (**choice C**) refers to the condition in which cells have extra sets of chromosomes. The most common polyploidy is triploidy, in which the cells have 69 chromosomes rather than the usual 46. Polyploidy is not the cause of Down syndrome.

A translocation refers to the condition in which there is breakage and removal of a large segment of DNA from one chromosome, followed by the segment's attachment to a different chromosome. A Robertsonian translocation (**choice D**) is a particular type of translocation involving the acrocentric chromosomes. Down syndrome is rarely caused by a Robertsonian translocation (approximately 3–4% of cases).

Turner syndrome (**choice E**) describes the condition in which the newborn has a karyotype of 45,X. In most cases it results from the loss of the paternal X chromosome. It is not the cause of Down syndrome.

98.

The correct answer is E. The studies show extremely marginal liver function, which would be tipped into overt liver failure by an anesthetic and an operation. He is not a surgical candidate.

Choice A obviously misses the gravity of his situation.

Vitamin K (**choice B**) works only when there is a functioning liver that can use it. In the absence of adequate liver function, it will not correct the prothrombin time.

Albumin (**choice C**) can be given, but it will have a short life span and will not correct the liver dysfunction. The low albumin is not the main problem per se, it is a symptom of how bad his liver is.

The same is true of bilirubin (**choice D**). It is a symptom, not the problem. We can operate on patients with much higher bilirubin if it is not due to intrinsic liver disease.

99.

The correct answer is B. This patient has Guillain-Barré syndrome, characterized by the three As: acute, areflexic, ascending. It is an autoimmune-mediated, demyelinating ascending paralysis that is often idiopathic but is also associated with HIV and *Campylobacter* infection. A lumbar puncture can often help clinch the diagnosis, although nerve conduction studies, if available, are often more consistent, as CSF findings can be ambiguous early in the disease. The classic finding is CSF with few cells and a high total protein (**choice B**).

Glucose level (**choice D**) and opening pressure (**choice A**) are usually fairly normal.

A pleocytosis, whether red blood cells (**choice E**) or white blood cells (**choice C**), should prompt one to think of another diagnosis.

100.

The correct answer is D. This boy has developed mumps. The most common complication in prepubertal children is meningoencephalomyelitis. Mumps is a viral infection with a paramyxovirus that causes painful enlargement of the salivary glands, predominantly the parotid glands. Transmission of the mumps virus occurs via airborne droplets, direct contact, and fomites contaminated with saliva. It is more common in the winter and spring. Outbreaks are related to lack of immunization. The treatment of mumps is supportive.

Arthritis (**choice A**) may develop during the course of mumps but is rather rare and usually mild in nature. It should be treated with nonsteroidal antiinflammatory medication or systemic corticosteroids, depending on the severity of the problem.

Dacryoadenitis (**choice B**) is a rare complication of mumps in which the lacrimal gland and duct are inflamed as a consequence of the viral infection.

Infertility (**choice C**) is not a common complication of mumps, even with bilateral orchitis.

Mumps oophoritis may develop in postpubertal females but it does not affect fertility.

Orchitis (**choice E**) is an uncommon complication that is usually not seen in prepubertal children. It may be unilateral or bilateral and is treated with support and bedrest. In 30% of cases it may be bilateral. Even in those cases, infertility is rare.