

Pediatrics Assessment Exam

1. During an annual visit, a 5-year-old boy is noted to have a heart murmur. The murmur is soft systolic, grade II in intensity, best heard along the left lower sternal border, and does not radiate. The murmur decreases in intensity in the sitting position. The first and second heart sounds are normal and no gallop is heard. The patient is an active child with no symptoms of tiredness or breathlessness, and he has a good appetite. He was hospitalized as an infant for bronchiolitis. There is no other history of lung infections. His weight and height parameters are at the fiftieth percentile. Lung examination reveals good breath sounds and no rales. There is no hepatomegaly or peripheral edema. The parents are concerned when they are told about the murmur. Which of the following is the most appropriate next step in management?
 - (A) Obtain a chest x-ray
 - (B) Obtain a hematocrit
 - (C) Perform an electrocardiogram
 - (D) Reassure the parents that the child is healthy
 - (E) Send the child for an echocardiogram
2. A 7-year-old boy is brought to the physician because of a new-onset rash on the trunk that appeared after a recent family camping trip in central Pennsylvania. The mother remembers that her son had a short-lived “flu” for several days on returning from camping that resolved with over-the-counter cold medication. Approximately 2 weeks later they noticed a red spot develop on his flank that has since been enlarging. They tried applying cortisone cream to it with no result. The boy is in good overall health and has no past history of serious medical problems. When the physician inquires about possible insect bites during the trip, the mother reports that she had removed several ticks from her son during their trip. No other family member has had similar symptoms and the family history is unremarkable. On physical examination the patient is in no acute distress. His vital signs are within normal limits. Inspection of the skin reveals a large, annular, erythematous plaque with central clearing present on the right flank. The largest diameter of the skin lesion is approximately 12 cm. Axillary and inguinal lymph nodes are not palpable. Potassium hydroxide preparation of skin scrapings from the edge of the lesion is negative. Which of the following is the most appropriate treatment for this patient?
 - (A) Intramuscular triamcinolone
 - (B) Oral amoxicillin
 - (C) Oral doxycycline
 - (D) Topical hydrocortisone
 - (E) Topical ketoconazole

3. An 8-year old boy is brought to the emergency department because of wheezing. According to the parents, the child has “always” had asthma and uses an albuterol inhaler at home when he has wheezing. This episode started 2 days ago but did not respond to several doses of inhaled albuterol. He has been coughing frequently and vomited twice in the previous day. Examination shows that he has diffuse wheezing with decrease in breath sounds on both sides, and prefers to sit up in a tripod-like position. Vital signs are: pulse 142/min, respirations 38/min, blood pressure 108/72 mm Hg, and temperature 37.2 C (99.0 F). Pulse oximetry shows that the oxygen saturation is 89%. Which of the following is the most appropriate first step in management?
- (A) Albuterol by nebulizer
 - (B) Chest x-ray
 - (C) Intravenous fluids
 - (D) Methylprednisolone
 - (E) Oxygen by mask
4. A 16-year-old girl is brought to the physician because of “swollen glands” on the neck that were first noticed 3 days earlier. The girl is otherwise feeling well and denies sore throat, fever, chills, and headaches. Her past medical history is unremarkable, and she does not take any medications on a regular basis. The family history is also non-contributory and no other family members have similar disease. On physical examination the patient does not seem to be in any acute distress. Her vital signs are within normal limits. On the right side of her chin three faintly pink 3-mm papules are noted. When specifically questioned about the papules she reports that these “pimples” arose a couple of weeks earlier after her pet kitten scratched her, but that they are now almost resolved. Inspection of the neck is significant for marked swelling of the right side below the jaw line, and palpation reveals enlarged, tender, mobile lymph nodes in that area. Which of the following studies is indicated at this time?
- (A) Nasopharyngeal swab for culture
 - (B) Needle biopsy of the lymph nodes
 - (C) No further studies are indicated
 - (D) Radiography of the head and neck
 - (E) Serum antibody titers
5. A previously healthy 6-year-old girl has a low-grade fever, malaise, and a new-onset rash on her face and arms of 2 days’ duration. She attends kindergarten but the parents are not aware that any other children have been reported sick. They have a 5-month-old son also, but he has not shown any signs of illness. The family history is unremarkable. On physical examination the patient seems to be in mild distress. Her temperature is 37.3 C (99.1 F), pulse is 120/min, and respirations are 28/min. Physical examination shows hyperemic oropharyngeal mucosa and edematous tonsils with scant clear secretion. The patient’s face has bright red erythema of the cheeks bilaterally. There is a reticulated, erythematous exanthem on the trunk and upper extremities. The palms and soles are unremarkable. Which of the following is the most likely etiologic agent of this patient’s disease?
- (A) Coxsackievirus A16
 - (B) Herpes simplex virus type 1
 - (C) Human herpesvirus type 6
 - (D) Parvovirus B19
 - (E) Rubella virus
6. A 7-month-old infant is brought to the clinic for a follow-up visit. Two days ago the infant was taken to the emergency department because of a large bruise over his thigh without any obvious history of trauma. Coagulation tests had revealed that the factor VIII level was less than 1% (normal 70 to 130%), which suggested a diagnosis of hemophilia A. In the intervening period, there had been no further episodes of bruising or bleeding. On today’s visit the infant appears active and playful, with no pain in the affected extremity. History reveals that there are two other siblings in the family, both female, who have no bleeding tendency. Neither of the parents has history of excessive bleeding, but one of the maternal uncles has hemophilia A. The mother is pregnant with her fourth child now, but is worried that the baby may be born with hemophilia A. In advising the parents, which of the following is the best estimate of the risk of hemophilia in the next child?
- (A) 25% for a female offspring, 50% for a male offspring
 - (B) 25%, regardless of the offspring’s sex
 - (C) None if it is a female offspring, 50% for a male offspring
 - (D) No risk if the father is normal
 - (E) Risk cannot be determined unless mother’s carrier status is determined

7. A 4-year-old boy is brought to the emergency department because of fever, malaise, and photophobia of 4 days' duration. The child's illness began with a runny nose and loss of appetite but has worsened progressively despite the use of over-the-counter cold medication. This morning his parents noticed a rash on the head and neck. The child was adopted recently from an orphanage in Eastern Europe and was in good health when he arrived in the United States 2 weeks earlier. The parents had been assured there was no significant past medical or family history. Apart from seeming slightly malnourished, the couple had not noticed any health problems. They are not sure if he had received all the necessary immunizations. His temperature is 40.1 C (104.2 F). He is quite bothered by the examination light and has significant conjunctival injection bilaterally. There are bluish-gray specks located on the buccal mucosa. There is a maculopapular erythematous rash on the forehead and behind the ears, descending on the sides of the neck. Administration of which of the following vaccines most likely would have prevented this disease?
- (A) Hepatitis B vaccine
 - (B) Influenza vaccine
 - (C) Measles-mumps-rubella vaccine
 - (D) Meningococcal vaccine
 - (E) Varicella vaccine
8. A 7-year-old girl is brought to the clinic for a routine school physical examination. The child has been regularly followed in the same clinic, and all previous visits were uneventful. She has demonstrated normal gain in weight and height, and her school performance in the previous year was satisfactory. At today's visit, both the height and weight are at the 75th percentile. Her blood pressure is normal and there is no cardiac murmur. Lungs are clear and the abdomen shows no palpable masses. Genitourinary examination reveals normal female genitalia, with no clitoromegaly. There is sparse growth of pubic hair. There is no facial or axillary hair, and breast development is absent. The mother is concerned about the appearance of pubic hair in her daughter. Which of the following best describes this patient's condition?
- (A) Congenital adrenal hyperplasia
 - (B) Exogenous androgen effect
 - (C) Normal puberty
 - (D) Premature adrenarche
 - (E) Turner syndrome
9. A previously healthy 12-month-old girl is brought to the clinic because of sudden-onset high fever and runny nose. She became very fussy and irritable the prior evening and spent most of the night crying. She has not eaten well since the illness began. The patient goes to daycare where several other children have been reported ill in the prior week. Her temperature is 40.3 C (104.5 F), pulse is 130/min, and respirations are 30/min. Physical examination reveals bilateral conjunctivitis, rhinorrhea with edematous nasal mucosa and clear secretions, and palpable occipital lymph nodes. Symptomatic treatment is recommended. Two days later the fever and runny nose have suddenly ceased but the child has broken out in a generalized skin rash consisting of hundreds of pinkish-red spots over almost the entire skin surface. Which of the following is the most likely diagnosis?
- (A) Measles
 - (B) Roseola infantum
 - (C) Rubella
 - (D) Scarlet fever
 - (E) Varicella
10. A 14-year-old girl with asymptomatic skin lesions is referred to the dermatologist from a psychiatric hospital where she was being evaluated for mental retardation and emotional problems. There is no record of a seizure disorder. The family history is unavailable because the child was adopted. She is on no medication and denies allergies to medication. On physical examination, the patient is in no acute distress. Her blood pressure is 100/50 mm Hg, pulse is 85/min, and respiratory rate is 16/min. Neurologic examination reveals a cooperative girl with mild mental retardation and low affect. Inspection of the skin is significant for multiple homogeneously brown, well-demarcated patches 2 to 5 cm in diameter and several pedunculated, soft, flesh-colored papules on the right inner thigh. Funduscopic examination is normal. Mild scoliosis is detected on skeletal exam. A complete blood count with differential white count was performed on admission to the psychiatric institution and was within normal limits. Which of the following radiologic findings is most likely to be present?
- (A) Jaw cysts
 - (B) Osteopoikilosis
 - (C) Osteopathia striata
 - (D) Thinning of the long-bone cortex
 - (E) "Train track" calcifications of the skull

11. A 5-year-old boy has dysuria, fever, leukocytosis, and right flank pain. On physical examination he is tender to fist percussion over the right costovertebral angle. Urinalysis shows grossly infected urine. He is treated with appropriate antibiotics and the infection clears. This is verified by repeated negative urinary cultures. The family indicates that he has had two previous similar episodes that were also successfully treated with antibiotics. No further workup was done following those two episodes. The reason for this recurrent problem is most likely to be diagnosed with which of the following studies?
- (A) Cystoscopy
 - (B) Intravenous pyelogram
 - (C) Renal ultrasound
 - (D) Retrograde urethrogram
 - (E) Voiding cystourethrogram
12. A previously healthy 12-year-old Middle Eastern boy is brought to the office because of low-grade fever and a skin rash of 2 days' duration. He complained of a sore throat and mild neck pain several days earlier. The entire family immigrated to the United States a year earlier as refugees from a war-torn country. This is the child's first visit to a physician since then. No other family members have been affected with the illness; however, they live in close quarters with several other relatives and are concerned that the infection might spread. The boy's temperature is 39.2 C (102.6 F), pulse is 90/min, and respirations are 22/min. He is in the fiftieth percentile for height and weight. Inspection of the nasopharynx reveals mild hyperemia and a serous exudate on the posterior pharyngeal wall. Faint erythematous macules are present on the soft palate. He has tender, enlarged posterior cervical and posterior occipital lymph nodes. In addition, there are many erythematous macules and papules on the face, chest, and proximal extremities. Who of the following would be most likely to develop complications from contact with this patient while he is infectious?
- (A) His 4-month-old niece
 - (B) His 15-year-old sibling
 - (C) His 23-year-old pregnant aunt
 - (D) His 60-year-old grandmother with systemic lupus erythematosus
 - (E) His 75-year-old grandfather with diabetes mellitus
13. A 13-year-old boy has been complaining of pain just below the knees for the past several months. He plays for the school basketball team and practices regularly. There is no history of fever or change in weight, appetite, or level of energy. There is no family history of rheumatic illness. Examination reveals that he has normal weight and height and is in Tanner stage III of pubertal development. Knee examination shows that the tibial tuberosities are prominent and tender on both sides. The movements of the knee are unrestricted on both sides and there is no redness or swelling of the joint. Radiographs reveal mild elevation of the tibial tuberosity, but are otherwise normal. Which of the following is appropriate management?
- (A) Obtain a complete blood count
 - (B) Obtain an erythrocyte sedimentation rate
 - (C) Obtain a magnetic resonance imaging of the knees
 - (D) Reassure the family that the condition is self-limiting
 - (E) Refer the patient for orthopedic consultation
14. A 2-week-old boy is brought to the clinic for routine checkup. The infant was born at full term after a normal pregnancy by normal vaginal delivery, and the birth weight was 7 lb (3.15 kg). He was breast-fed from birth and the mother reports that he was feeding well for the first 10 days, but lately he has become fussy and often interrupts feeding. The infant's weight today is 7 lb, 8 oz (3.38 kg) and he seems comfortable. His temperature is 37.2 C (98.9 F), and pulse and respiratory rate are normal. Examination of the mouth reveals white plaques on the buccal mucosa, tongue, and palate, which on removal leave behind punctate bleeding spots. The lung and heart examinations are normal. There is no evidence of hepatosplenomegaly. There is no rash on the perineum or genital areas. Which of the following is the most appropriate management?
- (A) Amoxicillin therapy
 - (B) Amphotericin B therapy
 - (C) Change to formula feeding
 - (D) No treatment is required
 - (E) Nystatin therapy

15. A 2-year-old boy is brought to the emergency department because of noisy breathing for the last 2 days. He was well until 3 days ago when he developed a fever up to 38.5 C (101.3 F), cough, and runny nose. One day later, the breathing became noisy with a loud sound during inspiration, which is more prominent during night. He is able to eat and drink fluids and has no vomiting. There is no previous history of similar episodes or chronic cough or choking. On examination, the child has mild respiratory distress with a loud inspiratory stridor. He is alert and afebrile, and oxygen saturation is 98% in room air. Examination reveals good breath sounds on both sides and no wheezing. There is no cardiac murmur and abdominal examination is normal. Chest x-ray shows that the subglottic space is narrowed and resembles a pencil point. Which of the following is the most likely causal organism?
- (A) *Haemophilus influenzae*
 (B) Parainfluenza virus
 (C) Respiratory syncytial virus
 (D) *Streptococcus* group A
 (E) *Streptococcus pneumoniae*
16. A 2-year-old boy is brought to the physician for a routine health maintenance examination. He was previously followed at another clinic, but due to change in medical insurance he had to switch physicians. He was born at full term with a birth weight of 7 lb, 12 oz. He was fed an iron-containing formula from birth and since the age of 1 year he is on whole cow's milk. He also eats various other foods that the mother cooks for the family. He is able to join two words together and has a vocabulary of well over 50 words. He was never sick enough to require hospitalization. At the 1-year visit, he was found to have hemoglobin of 10.2 g/dL. He was put on oral iron (ferrous sulfate drops), which the mother administered regularly. However, 2 months later his hemoglobin was still 10.3 g/dL. The dose of iron was increased and the hemoglobin was rechecked 3 months later and found to be unchanged. Thereafter, the mother had continued the ferrous sulfate drops until this visit. Today a complete blood count shows:

Hemoglobin	10.3 g/dL
Hematocrit	31%
Mean corpuscular volume	62 fL
RDW	13.1
Reticulocytes	1.8%
Leukocyte count	$10.5 \times 10^9/L$
Differential:	Neutrophils 35%, lymphocytes 54%, monocytes 8%, eosinophils 1%
Platelet count	$280 \times 10^9/L$

Which of the following studies is most likely to reveal the cause of his anemia?

- (A) Bone-marrow aspiration
 (B) Hemoglobin electrophoresis
 (C) Serum B₁₂ level
 (D) Serum iron and ferritin level
 (E) Upper and lower gastrointestinal endoscopy

17. A 16-year-old boy has a milky white discharge from his urethra, most prominent in the morning when he wakes up. There is also a mild burning sensation when he urinates. It started approximately 2 weeks earlier and is not showing any signs of improvement. He is in good general health and has no other concerns. He is sexually active but does not have a steady sexual partner. He does not use a condom regularly. In the previous 6 months he had sexual relations with four different women. On physical examination, the patient is a well-developed young man in no acute distress. His vital signs are within normal limits. Inspection of the genitals reveals mild erythema of the urethral orifice and a purulent discharge that is easily expressed with manual pressure. The inguinal lymph nodes are not palpable. There do not appear to be any lesions present on the penis or adjacent skin. A urethral swab is most likely to show which of the following findings?
- (A) Colorless pyriform flagellates in a saline smear
 - (B) Chains of Gram-negative bacilli in macrophages
 - (C) Gram-negative intracellular diplococci in polymorphonuclear cells
 - (D) Motile spiral spirochetes on darkfield microscopy
 - (E) Multinucleated giant cells in a Tzanck smear
18. A full-term newborn infant girl is noted to be cyanotic and in severe respiratory distress. She has no breath sounds on the left and cardiac sounds are heard only faintly on the left side. Breath sounds can be heard on the right, but they are obscured by prominent heart sounds on that side. The abdomen is scaphoid. X-ray shows the presence of bowel loops on the left hemithorax and deviation of the mediastinal structures to the right. Which of the following is the most appropriate next step in management?
- (A) Emergency surgical intervention
 - (B) Endotracheal intubation and mechanical ventilation with supplemental oxygen
 - (C) Insertion of a chest tube on the left
 - (D) Pharmacologic agents to increase the pulmonary vascular resistance
 - (E) Positive pressure ventilation through a face mask
19. A 16-year-old boy comes to the physician because of a nonhealing ulcer on the left heel that has been present for 3 months. The family has recently moved to the area from the West Coast and his medical records have not yet been transferred. He has had problems with his legs since he was about 10 years old—it started as vague weakness and increased clumsiness but progressed over the years to the point that he now needs a cane to walk and special orthotics to support his normal weight. The family history is significant for similar disease in multiple members of his mother's family. He is currently taking only nonsteroidal antiinflammatory drugs for muscular pain, and he denies allergies to any medication. On physical examination, the patient is in no acute distress. His vital signs are within normal limits and a basic chemistry panel and complete blood count performed a month earlier show no abnormalities. Inspection of the patient reveals considerable leg muscle wasting with a stork-leg appearance. There is pronounced bilateral pes cavus deformity and thoracic scoliosis. Deep tendon reflexes are absent on the lower extremities bilaterally and proprioception is significantly decreased. You note the presence of essential tremor while examining the patient. On the hands there are palpable enlarged peripheral nerves. A small, shallow, clean ulcer on the right heel corresponds to a rough protruding part on his right prosthesis and you assure him that getting a new prosthesis will expedite the healing process. Which of the following is the most likely diagnosis?
- (A) Charcot-Marie-Tooth disease
 - (B) Duchenne muscular dystrophy
 - (C) Friedrich ataxia
 - (D) Guillain-Barré syndrome
 - (E) Werdnig-Hoffman disease

20. A 20-year-old pre-med student comes to the clinic because of a painful outbreak on his genitals that started 2 days earlier. He also complains of malaise and low-grade fever that is partially responsive to aspirin. Reluctantly, he admits that about a week before this illness he had sexual intercourse with a woman he met at a local fraternity party. He used a condom but noticed after their encounter that it had ripped at the tip. He is in good overall health and works out on a regular basis. On physical examination, he is a well-developed, fit young man with a temperature of 37.9 C (100.2 F) and a pulse of 90/min. His respiratory rate and blood pressure are within normal limits. Inspection of the genitals reveals multiple tense, grouped vesicles with an erythematous base on the glans and shaft of the penis. There are also several painful small ulcerations. The inguinal lymph nodes are symmetrically enlarged bilaterally, and tender to palpation. Which of the following diagnostic studies is indicated at this time?
- (A) Bacterial culture
 - (B) Complete blood count
 - (C) KOH preparation
 - (D) Tzanck preparation
 - (E) Skin biopsy
21. A 1-year-old child is brought to the pediatric emergency department by paramedics. The child was in the care of his baby-sitter, who reports she found the infant lethargic in his playpen. She denies that the child could have swallowed any medication or other substance. The child's parents are currently not available to provide any information. The infant is semi-arousable and irritable when examined. There is soft tissue swelling over the right parietal area, subconjunctival hemorrhages bilaterally, and multiple ecchymoses over the back and upper chest. Which of the following is the most appropriate next step in management?
- (A) Await parental consent before obtaining any necessary diagnostic procedures on the child
 - (B) Confront the baby-sitter
 - (C) Consult a social worker first thing in the morning
 - (D) Protect the child's confidentiality at all costs
 - (E) Report the suspicion to the appropriate state agency immediately
22. A newborn infant who was born full-term is discharged home from the hospital with his mother on day 3 of life. At age 1 week the infant is seen by a pediatrician for his first newborn visit. Nutritional intake has been adequate and the infant has been gaining weight appropriately. Physical examination is normal. As part of the routine behavior assessment, the physician educates the infant's mother about what she can expect from the child developmentally. Which of the following will most likely be found in this infant?
- (A) Absence of grasp reflex
 - (B) Absence of rooting reflex
 - (C) Asymmetric tonic neck reflex
 - (D) Long latency between a painful stimulus and a cry
 - (E) Rolls over from back to stomach
 - (F) Turns head toward human voice
23. A 16-year-old girl is brought to the emergency department by paramedics on a Saturday night. She is not accompanied by a family member, and is lacking available history. Physical examination reveals delirium, dilated pupils, dry and flushed skin and mucous membranes, peculiar myoclonus-like twitching, tachycardia, and fever. Her clinical condition appears to be deteriorating during a period of 15 minutes of observation. The attending physician suspects that the patient is having a reaction to an ingested drug. Which of the following medications would be most appropriate to treat this patient?
- (A) Edrophonium
 - (B) Phenobarbital
 - (C) Phenothiazine
 - (D) Physostigmine
 - (E) Thiamine

24. A 2-year-old boy has a cough and occasional wheezing for the past 3 weeks. Initially, his mother gave him an over-the-counter cough mixture but there was no improvement. He has had no fever, runny nose, or choking episodes during this time. He has been active with good appetite and does not seem to have lost any weight. He has a normal birth history and no previous episodes of coughing. There is no family history of asthma, tuberculosis, or cystic fibrosis. There is one older sibling, a 5-year-old sister who is healthy. Examination reveals an active, playful child in no distress. Throat, nose, and ears look normal. Chest examination reveals diminished breath sounds on the right side at the apex, and an intermittent wheeze is heard on the right side. Cardiac examination is normal and there is no enlargement of the liver or spleen. Extremities are normal with no clubbing of the fingers. Which of the following investigations is most likely to reveal the etiology of his cough?
- (A) Barium swallow
 - (B) Chest x-ray
 - (C) Esophageal pH probe study
 - (D) Skin allergen testing with house dust mite antigen
 - (E) Sweat chloride test
25. A 4-month-old girl is brought to the physician for a follow-up visit. She was delivered by cesarean section at 40 weeks' gestation after a high-risk pregnancy that was marked by reduced movements in utero and reduced spontaneous activity after birth. The Apgar scores were 5 and 6 at 1 and 5 minutes, respectively. After birth, the patient showed signs of bulbar weakness, with sucking and swallowing difficulty and a weak cry. In resting position, she assumed a jug-handle position of the arms and frog-leg position of the legs due to proximal symmetrical muscle weakness. At age 4 months, the physical examination shows a poorly active infant in the 8th percentile for height and weight. The arms are flaccid, with abducted and internally rotated shoulders. Her hips are abducted, with flexed knees and externally rotated feet. She cannot hold her head when in upright position because of spinal muscle atrophy. Inspection of the mouth reveals fasciculations of the tongue. The deep tendon reflexes are absent. Her parents are very concerned about her condition. They should be told which of the following?
- (A) It is associated with severe mental retardation
 - (B) It is inherited in an autosomal recessive trait
 - (C) The gene mutation is located on the X-chromosome
 - (D) Serum creatine kinase levels are the diagnostic test of choice
 - (E) She will never attain normal sphincter function

26. An infant is born to a 27-year-old primigravid mother after a full-term pregnancy. The delivery and resuscitation were uneventful and the Apgar scores were 7 and 10 at 1 and 5 minutes, respectively. During the newborn physical examination, the baby is discovered to have a heart murmur. He has been feeding well since birth and there is no respiratory distress or enlargement of the liver. Other features noted at this time are small ears and a prominent tongue. Echocardiogram reveals the presence of atrioventricular canal defect. There is no family history of congenital malformations or mental retardation. In planning further tests to establish a diagnosis, it is most appropriate to mention to the parents that the infant might have
- (A) DiGeorge syndrome
 - (B) Prader-Willi syndrome
 - (C) Trisomy 13
 - (D) Trisomy 18
 - (E) Trisomy 21
27. A 2-month-old male infant is brought to the emergency department with wheezing and rapid breathing for the last 2 days. He was born at full term with a birth weight of 7 lb, 2 oz, and was well until 2 days ago when he developed a fever of 38.8 C (101.8 F) and runny nose. He began to cough and then became "wheezy." He has been irritable and unable to feed from the bottle without pausing several times, and has vomited twice. There is a 4-year-old elder brother who has had a runny nose for the past week. Vital signs are: pulse 146/min, respirations 56/min, blood pressure 84/58 mm Hg, temperature 37.3 C (99.1 F), and oxygen saturation (by pulse oximetry) 96% on room air. Examination shows an active and alert baby in mild respiratory distress. The nose is blocked with clear secretions, while the throat and ear examination is normal. He has slightly reduced breath sounds on both sides and bilateral rhonchi. Cardiac examination is normal and there is no hepatosplenomegaly. Which of the following studies is most likely to reveal the etiology of the illness?
- (A) Bronchoscopy
 - (B) Chest x-ray
 - (C) Complete blood count
 - (D) Nasopharyngeal aspirate
 - (E) Throat swab
28. A 3-year-old boy is brought to the emergency department because of drooling. He developed a sore throat 3 days ago with fever up to 38.8 C (101.8 F). His mother gave him acetaminophen and an over-the-counter mixture for what she assumed was a common cold; however, since last night the fever had increased to 40 C (104.0 F) and the child has become increasingly unwell. He did not want to drink and his respirations were becoming noisy. When he woke up this morning he began drooling and appears to be in pain. There was no history of trauma, cough, or vomiting, and his immunizations are complete to date. Examination reveals a toxic and frightened-looking child who refuses to flex his neck. Vital signs are: temperature 40.3 C (104.5 F), pulse 148/min, respirations 32/min, blood pressure 110/68 mm Hg, and oxygen saturation 98% in room air. There is a bulge in the posterior pharyngeal wall, along with erythema and collection of oral secretions. Which of the following investigations would be most helpful in establishing a diagnosis?
- (A) Complete blood count
 - (B) Lateral neck x-ray
 - (C) Lumbar puncture
 - (D) Nasopharyngeal aspirate
 - (E) Throat swab
29. A 1-day-old newborn has developed an erythematous rash. The infant was born by normal vaginal delivery to a 26-year-old primigravid mother at full term with a birth weight of 8 lb, 3 oz. The delivery was uneventful and the Apgar scores were 8 and 10 at 1 and 5 minutes, respectively. The pregnancy was supervised and uncomplicated, with negative serology for syphilis and toxoplasmosis. Since birth the baby was formula fed and he has passed urine and meconium. Examination shows that the infant is alert looking, with good tone and normal Moro reflex. There is a blotchy erythematous rash on the face, chest, and back. A closer look at the rash reveals small vesicles at the center of the erythematous areas. The chest and lung examination is normal and there is no hepatosplenomegaly. Microscopic examination of the vesicular fluid reveals predominance of eosinophils. Which of the following is most likely to be the cause of this infant's rash?
- (A) Allergy to cow's milk
 - (B) Atopic dermatitis
 - (C) Erythema toxicum neonatorum
 - (D) Intrauterine infection
 - (E) Sepsis and disseminated intravascular coagulation

30. A 3-week-old infant is brought to the clinic because of a fever. He was born at full term after a normal pregnancy and weighed 7 lb, 12 oz at birth. The delivery was uneventful and he was discharged from the hospital after 48 hours. He has been formula fed and was doing well until the day before, when the fever was first noticed. Several family members have had a viral upper respiratory infection in the past week, including the mother. Upon examination, the temperature is 38.9 C (102.0 F). The baby appears awake and has good tone in the extremities. Respiratory, cardiovascular, and abdominal examinations are within normal limits. The eardrums are normal and there is no skin rash. In the office he is offered a bottle and is able to feed 3 oz of formula. Which of the following is the most appropriate management?
- (A) Acetaminophen infant drops for fever control and monitoring at home
 - (B) Arrange for another office visit in 2 days' time
 - (C) Refer to emergency room for urgent admission
 - (D) Send for a complete blood count and, if normal, reassure mother that baby has a viral infection
 - (E) Start oral amoxicillin
31. A 14-year-old girl is brought to the clinic for a routine school physical examination. The mother is concerned that her daughter has shown no signs of breast development and has not started menstruating. Past history does not reveal any significant illnesses. Review of the patient's growth reveals she was always just below the 5th percentile until she was 10 years old. However, her current height is well below the 5th percentile with an absence of pubertal growth spurt. Physical examination reveals a short girl with normal facial appearance. There is a prominent cubitus valgus deformity. The neck is short and the posterior hairline is low. There is no breast development, although some pubic and axillary hair is present. Examination of the heart, lungs, and abdomen reveals no abnormality. Which of the following studies is most likely to reveal the etiology of this patient's short stature?
- (A) Abdominal ultrasound
 - (B) Gonadotropin level
 - (C) Growth hormone level
 - (D) Karyotype
 - (E) Thyroid hormone level
32. A newborn infant is diagnosed with dehydration and is transferred to the neonatal intensive care unit. The infant was born at 38 weeks' gestation to a healthy 26-year-old mother. Prenatal care was good, there were no complications during pregnancy, and the delivery was by way of normal, spontaneous, vaginal delivery. Initially the infant seemed well; however, soon after there was poor oral feeding, vomiting, and lethargy. On physical examination in the NICU, the infant is lethargic and has a weak cry. Vital signs are systolic blood pressure 40 mm Hg, pulse 190/min, and respirations 25/min. The abdomen is soft and there are no masses palpable. Examination of the genitalia reveals an enlarged phallus with significant curvature of the phallus. The urethral meatus is in a hypospadiac location. The scrotum is rugated, and the testicles are nonpalpable bilaterally. At this time, which of the following is the most important information to obtain to treat the patient?
- (A) Arterial blood gas
 - (B) Karyotype
 - (C) Pelvic ultrasound
 - (D) Serum electrolytes
 - (E) Serum 17-alpha-hydroxyprogesterone level
33. A 21-year-old primigravid woman comes to the physician at 12 weeks' gestation. Her brother and sister are both affected with Disease X. Her mother also had the disease but her father was not affected. Her affected brother has three children, two of which are girls and both of which are affected with the disease. Her nephew does not have Disease X. Which of the following diseases is consistent with the described inheritance pattern?
- (A) Albinism
 - (B) Congenital adrenal hyperplasia
 - (C) Bruton agammaglobulinemia
 - (D) Hypophosphatemic rickets
 - (E) Duchenne muscular dystrophy

34. A 23-year-old primigravid woman with a history of alcohol and substance abuse is admitted to the hospital in true labor, showing signs of imminent delivery. She is transferred to the delivery room, and 30 minutes later a full-term baby girl is born. The new mother has not had any prenatal care because she “never had any problems during the pregnancy.” Examination of the mother shows signs of severe malnutrition. The newborn is 42 cm long and weighs 2,500 g. She scored 6 and 7 on the Apgar scale at 1 and 5 minutes, respectively. Physical examination of the neonate does not reveal any signs of acute distress. Her vital signs are within normal limits. However, physical examination reveals flaccid paralysis of the lower extremities, subluxed hips, and bilateral clubfoot deformity, and examination of the back reveals a large cystic sac protruding from the midline lumbosacral region. Deep tendon reflexes are absent on the lower extremities. Which of the following nutritional supplements could have prevented this defect?
- (A) Folic acid
 - (B) Vitamin A
 - (C) Vitamin B₂
 - (D) Vitamin B₆
 - (E) Vitamin K
35. A 9-year-old boy is brought to the physician for evaluation of what the parents complain of as “increasing clumsiness.” He used to be very athletic, participating in multiple sports, but has recently dropped out because he cannot keep up with the other children. The parents had also noticed a change in his speech, with difficulty expressing himself to the point that on several occasions he almost choked while talking. His prior medical history is unremarkable and he is on no medications. There is no family history of similar disorders. On physical examination, the child is in no distress and has normal vital signs. He is in the 50th percentile for height and weight. Neurologic evaluation reveals a wide-based gait with constant shifting of position to maintain balance. Sitting and standing are associated with titubation. When he walks there is uneven and irregular striking of the floor by his soles, with wild movements produced in trying to correct the imbalance. His speech is a little slurred and slow. Extensive blood work did not reveal any significant abnormality. An echocardiogram showed symmetric, concentric, ventricular hypertrophy. Which of the following is the most likely diagnosis?
- (A) Friedrich ataxia
 - (B) Refsum disease
 - (C) Spastic hemiplegia
 - (D) Werdnig-Hoffman disease
 - (E) Wilson disease

36. A 9-month-old boy is brought to the physician because the mother noticed “orange sand” in his diapers over the previous several days. The child has been in good health, although a little slow to develop his milestones. He has no significant past medical history and takes no medication. The family history is unremarkable. On physical examination, the patient is in no acute distress. His temperature is 36.7 C (98 F), pulse is 95/min, and respirations are 20/min. The weight and length are in the 20th and 10th percentiles, respectively. He sits with assistance and lifts his upper body when in a prone position, but does not attempt to crawl or stand. Involuntary movements indicative of extrapyramidal dysfunction are noted. There are prominent calluses on both hands and you note that the child repeatedly gnaws on those areas. A complete blood count and chemistry panel reveals macrocytic anemia and hyperuricemia. In addition, which of the following studies is most likely to be abnormal in patients with this condition?
- (A) Cerebrospinal fluid analysis
 - (B) Computerized tomography of the brain
 - (C) Magnetic resonance imaging of the brain
 - (D) Plain abdominal radiograph
 - (E) Ultrasound of the abdomen
37. A 16-year-old girl is brought to the emergency department by her parents because of nausea, vomiting, and malaise that have been progressively worsening over the course of several days. They also noted some speech changes and a fine tremor most prominent when she tried to complete fine tasks, such as threading a needle. They first thought she was on one of her crash diets to lose weight because she would not eat anything, but when she finally complained of feeling sick they decided to have her examined. She has no significant past medical history and her menstrual periods are regular. She denies allergies to medication and intake of any prescription or over-the-counter drugs in the previous month. The family history is significant for a paternal uncle who died at a young age of liver cirrhosis. On physical examination, the patient is in moderate distress. Her pulse is 110/min, respirations are 24/min, and blood pressure is 90/45 mm Hg. Inspection of the skin reveals multiple telangiectatic arterioles in the skin with radiating capillary branches, redness of the palms, and pronounced mild jaundice. There is a hint of digital clubbing. A tan-green color is seen in the limbic area of the eye. Laboratory studies reveal a ceruloplasmin level of 17 mg/dL and low total serum copper. Which of the following studies is necessary to confirm the diagnosis?
- (A) Analysis of 24-hour urine collection
 - (B) Computerized tomography of the brain
 - (C) Liver biopsy
 - (D) Skeletal radiologic survey
 - (E) Slit-lamp examination of the eye

38. A 12-year-old white girl has right distal thigh pain for the past several months that has worsened over the past few days. She recalls that she was struck in gym class by a ball in the affected thigh and that caused a period of excruciating pain. She also feels that her right thigh is slightly larger than her left and that her right calf and foot sometimes swell. She is a well-developed and well-nourished girl. The patient's vital signs are all within normal limits. On examination of the distal right thigh, there is a noticeable difference in circumference of the right thigh versus the left. Range of motion is decreased with respect to the knee secondary to pain. There is 1+ edema to the mid-calf level and distal pulses are normal and symmetric. Radiographs of the distal femur show a bone-forming mass with rays of bone formation radiating from the center of the mass. The periosteum is raised off the bone by the mass, causing fusiform swelling with reactive periosteal bone at the periosteal margin. Which of the following is the most likely diagnosis?
- (A) Benign bone cyst
 - (B) Fracture of the distal femur with callous formation
 - (C) Osteomyelitis of the distal femur
 - (D) Osteosarcoma
 - (E) Paget disease
39. A 20-month-old girl is brought for evaluation. The parents complain that she has been behaving in a peculiar way over the past couple months, as though she was forgetting the skills she had already mastered earlier. Her vocabulary seems to have shrunk, and she has become very placid compared to other children she spends time with. The child has lost interest in playing with her friends and is often found sitting alone, wringing or clapping her hands repeatedly. She no longer makes eye contact with her parents. The most recent development is the occurrence of breath-holding or rapid breathing while she is awake. She sleeps peacefully, though. Her prior medical history, as well as the family history, is unremarkable. On physical examination, the patient is in no acute distress and her vital signs are within normal limits. Her height, weight, and head circumference are in the 20th, 30th, and 5th percentiles, respectively. During the examination, she sits quietly on the examining table and does not make eye contact. Repeated hand wringing and general hypotonia are noted. A Wood lamp examination of the skin is negative. Basic laboratory studies and neuroimaging studies reveal no abnormality. Genetic testing is offered to the family. Which of the following genes is most likely to be sequenced in this patient?
- (A) MECP2
 - (B) NF1
 - (C) PTEN
 - (D) SPINK5
 - (E) TSC1

40. The parents of a 2-week-old child bring the newborn to the clinic for a routine visit. At birth the infant was noticed to have a cleft lip. Both parents are 24 years old and this is their first child. The pregnancy and delivery were uneventful and, apart from the lip, no other congenital malformations were noted. The birth weight was 7 lb, 4 oz, and at this visit the infant has gained 4 oz on formula feeds. Examination reveals an active newborn with unremarkable examination findings. The mother mentions that she has heard about recurrence of congenital abnormalities in subsequent offspring. The parents want to know the risk of having a second child with cleft lip. At this time, which of the following is the best advice?
- (A) Chromosomal testing from the infant and both parents is required to estimate the risk
 - (B) The recurrence risk of cleft palate is 5%
 - (C) The risk of recurrence is 25%
 - (D) The risk to the second child is not increased
 - (E) The risk will increase with increasing maternal age

41. A 12-year old boy is brought to the emergency department due to worsening of asthma. He has a history of several previous admissions due to status asthmaticus. At home he uses inhaled steroids on a regular basis. He started to wheeze 3 days ago and used an albuterol inhaler, which provided partial relief. In the last several hours before coming to the hospital he developed increasing difficulty in breathing and could not talk in full sentences. His vital signs are: pulse 148/min, respirations 32/min, blood pressure 126/74 mm Hg, and temperature 37.8 C (100.0 F). He appears confused and disoriented, lung examination reveals markedly diminished breath sounds, and pulse oximetry shows an oxygen saturation of 87%. He is started on oxygen and nebulized albuterol. Arterial blood gas analysis shows:

pH	7.29
pO ₂	84 mm Hg
pCO ₂	44 mm Hg
HCO ₃	24 mEq/L

Which of the following is the most appropriate next step in management?

- (A) Administer intravenous fluids
- (B) Administer methylprednisolone
- (C) Administer sodium bicarbonate
- (D) Admit for observation
- (E) Intubate and provide mechanical ventilation

42. A 5-year-old boy is brought to the physician for evaluation. The parents complain that he has trouble maintaining his balance and has started walking in a peculiar, waddling way as of lately. He pushes his knees when he wants to stand up and has developed large calf muscles that seem to stand out from the rest of his body. His medical history is significant for several episodes of pneumonia. He is on no medications at this time and has no allergies to medication. The family history is unremarkable. On physical examination, the child is in no acute distress. His vital signs are within normal limits. He has difficulty generating a forceful cough for chest auscultation. Inspection of the trunk and extremities reveals very pronounced calf muscles. Deep tendon reflexes are significantly diminished. There is mild kyphoscoliosis secondary to paraspinal muscle weakness. Which of the following studies is most likely to aid in confirming the diagnosis?
- (A) Electrocardiogram
 - (B) Electromyography
 - (C) Muscle biopsy
 - (D) Radiographs of the spine
 - (E) Serum creatinine phosphokinase

43. A 9-month-old male infant is brought to the clinic because of wheezing and a cough. He has been taken to the emergency room three times in the past 5 months for "breathing problems." On each visit he responded to inhaled bronchodilator therapy. Recently, though, the wheezing and coughing episodes are becoming more frequent. Also, the infant has developed loose stools, which are greasy and foul smelling. His weight today is 16 lb, and the infant's growth chart shows loss of growth percentiles in the last 3 months. Examination reveals a smiling infant in mild respiratory distress. The pulse is 128/min, respirations are 42/min, and oxygen saturation is 98% on room air. Chest examination reveals an increased anteroposterior diameter and bilateral wheezing. There is no cardiac murmur and the abdominal examination shows no hepatosplenomegaly. Which of the following studies is most likely to reveal the etiology of the infant's illness?

- (A) Bronchoscopy
- (B) Chest x-ray
- (C) Lung biopsy
- (D) Sweat chloride testing
- (E) Upper and lower gastrointestinal endoscopy

44. A physician volunteers to participate in a screening program at the local school organized to detect head lice in their students. The program is conducted at the school, where the physician systematically examines the head and scalp hair of all third-graders present. During the examinations, there is a 9-year-old girl with thick, long, curly blonde hair that has abundant white, gritty scale on the retroauricular and posterior occipital scalp with several enlarged, nontender suboccipital lymph nodes. There is also patchy red lichenification and excoriations on the posterior neck. Despite the best effort to characterize the nature of the white particles, which appear to be attached to hair shafts, the physician is unable to make a conclusive decision. Which of the following may prove helpful in making the correct diagnosis in this patient?
- (A) Culture on Sabouraud's agar
 - (B) Hair penetration test
 - (C) Microscopic examination of the snipped hair
 - (D) Potassium hydroxide hair preparation
 - (E) Pull test
45. A concerned mother brings her 2-month-old infant to the clinic for a routine appointment. The child is healthy and the mother has no medical complaints at this time. He has begun to play with a rattle, can hold his head erect temporarily, and often coos and smiles when played with. One of the mother's friends lost a similarly healthy 2-month-old infant to sudden infant death syndrome (SIDS). Understandably the mother wishes to do everything possible to protect her child while he is sleeping. She has heard conflicting information regarding sleeping habits and positions that can possibly reduce the risk for SIDS. She should be advised that her infant should sleep in which of the following positions?
- (A) Any comfortable position
 - (B) Left lateral decubitus
 - (C) Prone position
 - (D) Right lateral decubitus
 - (E) Supine position
46. A mother brings her 4-year-old child to the emergency department for evaluation of persistent fevers. For the last week the young girl has suffered from spiking fevers and malaise. The mother is concerned that she may have caught something while visiting family in Japan 1 month prior, though the child has no known sick contacts. Past medical history and family medical history are unremarkable. Aside from the usual childhood illnesses, the patient has been well. She is up to date on all appropriate immunizations. Physical examination shows bilateral conjunctival injection and dry, cracked mucous membranes. Her lips are fissured and her tongue is erythematous and swollen. She has marked cervical lymphadenopathy, with at least one tender node of approximately 2 cm. Additionally, across her chest is a pleomorphic, erythematous rash. Erythema is also present on the soles of her feet and palms of her hands, as is some mild desquamation. Laboratory studies show a normocytic anemia, a thrombocytosis, and a markedly elevated erythrocyte sedimentation rate. Blood and urine cultures are unrevealing. Given this patient's presentation, which of the following is the most appropriate treatment?
- (A) Acetaminophen
 - (B) Aspirin
 - (C) Aspirin and intravenous immunoglobulin
 - (D) Plasmapheresis
 - (E) Prednisone
47. A 16-year-old girl comes to the clinic for evaluation of delayed puberty. She is concerned about her development, as she feels her body has not changed a bit since she was 9 years old. Although she reports that she has always been shorter than her classmates, she has generally been healthy. She has had no major illness and knows of none that run in her family. Physical examination reveals a short, stocky appearing young woman. She has Tanner stage 1 breasts and sparse pubic and axillary hair. Her ears appear lower than usual and she has a low hairline and a small mouth and mandible. Additionally, her neck is short and she has a wide, broad chest with nipples that are farther apart than normal. She is markedly hypertensive. Which of the following is the most likely cause of this patient's hypertension?
- (A) Aortic coarctation
 - (B) Essential hypertension
 - (C) Ovarian failure
 - (D) Renal artery stenosis
 - (E) Renal failure

48. A 32-year-old multiparous woman delivers a baby boy at full term after an uneventful pregnancy. She has three healthy children at home and has no significant past medical history. She denies taking any medication during her pregnancy. The neonate scores 7 and 8 on the Apgar scale at 1 and 5 minutes, respectively. He is 48 cm long and weighs 3,200 g. His vital signs are within normal limits. On physical examination, asymmetry of the lower extremities is noted. The left leg is less well developed and the foot is slightly deformed. An x-ray of the lumbosacral spine is performed and shows laminal defects in the L5-S1 region with widening of the interpedicular spaces and butterfly vertebrae. Which of the following skin lesions is most likely to be associated with this defect?
- (A) Ash-leaf macule
 - (B) Becker nevus
 - (C) Mongolian spot
 - (D) Patch of hair
 - (E) Salmon patch
49. A 5-year-old white boy is complaining of groin pain. He has been limping for approximately 1 month and it has gotten progressively worse over the past week. He has not had any recent infectious contacts or any known trauma. The boy is in the fifteenth percentile for height and the tenth percentile for weight. His temperature is 36.7 C (98 F). He has decreased right hip internal rotation and abduction. The patient's knee examination is normal. When observing his gait, the physician notes that the boy leans his torso to the right side during the stance phase on the right leg. Which of the following is the most likely diagnosis?
- (A) Developmental dysplasia of the hip
 - (B) Juvenile rheumatoid arthritis
 - (C) Legg-Calve-Perthes disease
 - (D) Septic arthritis
 - (E) Slipped capital femoral epiphysis
50. A 25-year-old woman comes to the emergency department in labor. She delivers a 7-pound baby boy without complications. She tells the obstetric staff that she has been visiting a friend for the last 2 weeks and that her medical records from her obstetrician will not be available for another 2 days. She is carrying a medical note from her doctor stating that she is hepatitis B surface antigen-positive and hepatitis B e antigen-positive. She is concerned that the baby will be affected by her illness. Which of the following is the most appropriate intervention for the newborn?
- (A) Administer first dose of hepatitis B vaccine only
 - (B) Administer first dose of hepatitis B vaccine with hepatitis B immune globulin
 - (C) Administer hepatitis B immune globulin only
 - (D) Explain to the mother that no intervention is indicated
 - (E) Send hepatitis serologies on the mother and treat the newborn on the basis of the results