

Block

THREE

Questions

Setting 3: Inpatient Facilities

You have general admitting privileges to the hospital. You may see patients in the critical care unit, the pediatrics unit, the maternity unit, or recovery room. You may also be called to see patients in the psychiatric unit. A short-stay unit serves patients who are undergoing same-day operations or who are being held for observation. There are adjacent nursing home/extended-care facilities and a detoxification unit where you may see patients.

80 • Blueprints Q&A Step 3 Pediatrics

120. A 10-month-old baby is noted to be chronically irritable. His mother, who wears a veil and gown and is a vegetarian for religious reasons, has been breastfeeding the baby exclusively. The baby's fontanelle is still open and is noted to be bulging. On physical exam you find frontal bossing, widening of the wrists, and bowed legs. Because of the irritability and bulging fontanelle, you order an MRI scan. It is reported as normal aside from showing evidence of increased intracranial pressure. An LP yields normal cerebrospinal fluid and an elevated opening pressure. The next step in the management of this patient would be to:

- A. Perform serial LPs
- B. Prescribe acetazolamide (Diamox)
- C. Administer corticosteroids
- D. Prescribe vitamin D and calcium supplements
- E. Refer patient to a neurosurgeon

121. A baby seen in the newborn nursery is found to have a loud heart murmur. The baby has been feeding poorly. The chest radiograph demonstrates decreased pulmonary vascular flow and you are considering tricuspid atresia as a possible diagnosis. In preparing to tell the parents about their baby's heart defect, which of the following is most accurate?

- A. In this condition, blood cannot flow from the right atrium to the right ventricle
- B. In most cases, a patent ductus arteriosus (PDA) allows blood to get to the lungs
- C. Most patients do not have cyanosis in the newborn period
- D. The cardiogram generally shows right ventricular hypertrophy
- E. The goal of medical and surgical treatment is to decrease blood flow to the lungs

122. A 3-day-old preterm infant develops intermittent apnea spells lasting greater than 20 seconds, accompanied by bradycardia. Investigation reveals no apparent infection or metabolic imbalance. These episodes seem to respond to gentle cutaneous stimulation. Which one of the following would be true for the management of this baby?

- A. The baby is too young to consider this idiopathic apnea
- B. No medication has been shown to be beneficial for frequent apnea
- C. Correction of anemia would have no effect on the frequency of apnea
- D. If the episodes are frequent, nasal continuous positive airway pressure (CPAP) might be beneficial
- E. Apnea of prematurity usually resolves at 2 months of age

123. A 4-year-old boy is seen because of increased thirst and increased urination. He was toilet trained at 2 years, but he still wets the bed every night. Your concern is that this boy could have diabetes insipidus (DI) or psychogenic water drinking. A morning urine specimen after an 8-hour fast shows a specific gravity of 1.005. His serum sodium is 140 mEq/L. What would be your next step?

- A. Administer vasopressin to see whether his urine can concentrate
- B. Administer a sodium load to see whether his kidneys can respond by increasing sodium excretion
- C. Increase the overnight fasting test to 12 to 15 hours to see whether his urine can concentrate
- D. Perform a daytime water deprivation test, measuring any changes in urine concentration and serum osmolality
- E. Because his serum sodium is normal, advise the parents that there is no need for further tests

124. An infant is born with port-wine nevus distributed and well-demarcated over the distribution of the right trigeminal area of the face. Which one of the following may apply to this syndrome?

- A. Pulsed dye laser treatments are frequently used and are quite effective for the skin lesions
- B. While seizures are common, most patients can be readily controlled with anti-convulsant therapy
- C. Medical management is the only avenue for the treatment of recalcitrant seizures
- D. Glaucoma on the affected side is always apparent in the first month of life
- E. Subsequent siblings have a 25% risk of being affected because this condition is autosomal recessive

125. You are called by the nurse to evaluate a term 3-hour-old infant who was born to a 25-year-old mother with unremarkable prenatal labs and an uncomplicated pregnancy. The baby was born via scheduled cesarean section without previous rupture of membranes secondary to a previous child born via cesarean section. On physical exam you notice a mild to moderately tachypneic infant with some coarse breath sounds bilaterally and mild intercostal retractions; the remainder of the physical exam is normal. The child is saturating 98% on room air. A chest radiograph is obtained (Figure 125). Your next step in management would be:



Figure 125 • Image courtesy of the Department of Radiology, Phoenix Children's Hospital, Phoenix, Arizona.

- A. Place the infant on supplemental oxygen
- B. Immediately administer antibiotics
- C. Draw a CBC/blood culture and then administer antibiotics
- D. Perform an ECHO
- E. Careful observation only

118. **C.** This child has the classic presentation of congenital adrenal hyperplasia (CAH), including ambiguous genitalia, hyperpigmentation, and salt wasting. The child is a genetic female with virilized genitalia. The most common form of CAH is 21-hydroxylase deficiency, which results in salt wasting and hyperkalemia. Low glucose is also common due to cortisol deficiency. These children often present with dehydration and even shock in the first 2 to 3 weeks of life. The treatment for CAH involves the essential replacement of cortisol and mineralocorticoids.
- A.** CAH is inherited as an autosomal recessive trait.
B. 21-Hydroxylase deficiency accounts for 90% of cases of CAH.
D. This child is a genetic female and does not have testes. This diagnosis should be considered in newborn infants with bilaterally nonpalpable testes.
E. Laboratory evaluation includes measurement of elevated 17-hydroxyprogesterone levels.
119. **C.** Fifteen to twenty percent of affected children have a positive family history for natal teeth. Natal teeth are seen in about 1 in 2000 newborn infants.
- A.** Natal teeth usually do not have a deep root system developed.
B. Natal teeth are typically located in the position of the mandibular central incisors.
D. There is a remote risk of aspiration should the tooth become dislodged. Elective extraction may be considered on an individual basis.
E. Natal teeth have been associated with other abnormalities such as cleft palate and Pierre Robin syndrome.
120. **D.** Increased intracranial pressure with a normal MRI scan and normal cerebrospinal fluid (CSF) suggests the diagnosis of pseudotumor cerebri. The treatment of pseudotumor cerebri usually involves treating the underlying condition, in this case vitamin D-deficient rickets. Breastfed babies of women who are gowned (no sun exposure) and vegetarian may develop rickets because of a lack of vitamin D in their mother's breastmilk. Pseudotumor cerebri can be due to many diseases, including those with deranged calcium and phosphorus metabolism, as well as galactosemia, hypoparathyroidism, pseudohypoparathyroidism, hypophosphatasia, prolonged corticosteroid use, excess or deficiency of vitamin A, obesity, and pregnancy. The best treatment for this baby is to provide supplements of vitamin D and calcium.
- A.** Some patients with increased intracranial pressure from pseudotumor cerebri are treated with serial lumbar taps to remove excessive CSF.
B, C. Acetazolamide and corticosteroids can also decrease intracranial pressure, but the underlying etiology should be treated first.
E. A neurosurgical consult would not be indicated in this case.

100 • Blueprints Q&A Step 3 Pediatrics

121. **A.** In patients with tricuspid atresia, blood is obstructed by an atretic valve and prevents flow from the right atrium to the right ventricle.

B. In tricuspid atresia, blood flows from the right atrium through a patent foramen ovale into the left atrium, then through the mitral valve to the left ventricle. In order to get to the lungs, some blood from the left ventricle passes through a VSD to the right ventricle. Shunting from the aorta to the pulmonary artery through a patent ductus is less common.

C. Because oxygenated and nonoxygenated blood mixes in the left atrium, patients with tricuspid atresia will have cyanosis in the newborn period.

D. The left ventricle has an increased load because of the right to left shunt that occurs; as a result left ventricular hypertrophy develops.

E. Treatment is aimed at improving pulmonary blood flow. Prostaglandins should be started immediately and balloon atrial septostomy may be needed as well. The goal of both palliative and definitive surgery is to assure better blood flow to the lungs.

122. **D.** Nasal CPAP may help splint the upper airway and prevent any obstructive component of apnea that may be occurring.

A. Generally, apnea of prematurity begins between the second and seventh day of life. Apnea on the first day, or beginning after the second week (or any time in a term infant), warrants immediate investigation.

B. Apnea of prematurity may respond to theophylline by mouth (PO), aminophylline (IV), or caffeine (PO).

C. Apnea associated with significant anemia may respond to packed RBC transfusion.

E. Apnea of prematurity generally resolves by 36 weeks postconceptional age (gestational age at birth plus postnatal age).

123. **D.** The dilute urine after an 8-hour overnight water deprivation is suspicious for DI. It would be dangerous to repeat a long overnight fast if this patient has DI. Therefore, a daytime water deprivation can be done lasting 7 to 8 hours with close observation for significant rise in urine concentration and serum osmolality. After 7 to 8 hours, and no significant urine concentration or rise in serum osmolality occurs, insipidus is confirmed, and the administration of vasopressin will distinguish central from nephrogenic DI. While DI is usually accompanied by hypernatremia, patients with free access to water may retain normal serum sodium concentration.

A. Vasopressin will result in urine concentration if the diagnosis is either psychogenic water drinking or central DI. Vasopressin can be used after an abnormal water deprivation test to distinguish between central and nephrogenic DI.

B. Sodium loading would be dangerous and offer no pertinent information.

C. Dilute urine after an 8-hour overnight water deprivation is suspicious for DI. It would be dangerous to repeat a long overnight fast if this patient has DI.

E. While DI is usually accompanied by hypernatremia, patients with free access to water may retain normal serum sodium concentration.

124. **A.** This is Sturge-Weber syndrome. Pulsed dye laser is frequently effective for the skin lesions and generally avoids scarring. Such therapy can begin in infancy.
- B.** Seizures are frequently unilateral on the contralateral side, and difficult to control.
- C.** For recalcitrant seizures, not well controlled with anticonvulsant medication, hemispherectomy or lobectomy has been effective, and may even prevent the development of mental retardation.
- D.** Glaucoma may develop with time, and therefore regular measurement of intraocular pressure is indicated.
- E.** Sturge-Weber syndrome is not hereditary, and is thought to result from anomalous development of the primordial vascular bed during the early stages of cerebral vascularization. The frequency of occurrence is 1 in 50,000.

125. **E.** The infant in this scenario is experiencing transient tachypnea of the newborn (TTN). TTN represents a transient pulmonary edema resulting in delayed absorption of fetal alveolar fluid secondary to a variety of risk factors that include precipitous delivery, macrosomia, and operative delivery without labor, like the infant in this scenario. Most infants recover well with only supplemental oxygen, if necessary, and observation. Obtaining a chest radiograph is a logical choice in any patient showing signs of respiratory distress. The x-ray in this scenario reveals a prominent right lobe of the thymus or sail sign that may often be confused as an infiltrate of pneumonia.
- A.** Although the use of oxygen will most likely not harm this patient, it is unnecessary with saturations of 98% on room air.
- B.** Simply administering antibiotics without obtaining cultures is generally not sound medical practice unless the patient is extremely toxic appearing.
- C.** Further infectious work-up should be delayed, as there are no risk factors such as prematurity, fever, or prolonged rupture of membranes. If the patient continues to not improve or worsens, then a more thorough investigation would be needed.
- D.** With no murmur on exam, saturations of 98% on room air, and a normal-shaped heart on x-ray, performing an ECHO at this time is not necessary.

126. **D.** The prolonged history of fever, limpness, and radiographic findings of periosteal elevation and subperiosteal fluid collection is consistent with the diagnosis of osteomyelitis. While those affected with sickle cell anemia are more susceptible to osteomyelitis with *Salmonella*, *Staphylococcus aureus* is still the predominant pathogen of osteomyelitis.
- A.** *Pseudomonas aeruginosa* infects avascular cartilaginous structures of the foot following puncture wounds.
- B, C.** *Salmonella* and *Brucella* tend to cause osteomyelitis of the vertebrae.