American College of Emergency Physicians
Section of Pediatric Emergency Medicine

Pediatric Endocrine Emergencies Quiz Answers 3/04

Paul Ishimine, MD
Department of Emergency Medicine, University of California San Diego Medical Center
Division of Pediatric Emergency Medicine, Children's Hospital and Health Center, San Diego

1. c. This patient has severe diabetic ketoacidosis (DKA). DKA is frequently the initial presentation of a child with new-onset type I diabetes and can be life-threatening. This child presents with signs of shock. Patients with DKA can be profoundly dehydrated, and therefore initial fluid therapy should be directed towards establishing an adequate circulating volume. Therefore initial resuscitation needs to occur with solutions that will stay in the intravascular space. Once the patient is hemodynamically stable, the patient can be switched to half-normal saline (with appropriate potassium and dextrose replacement), but in an unstable patient, the key resuscitation fluid is normal saline. Patients in DKA all require insulin therapy in order to stop ongoing ketone body formation. While subcutaneous insulin can be used in mild DKA, this patient has significant DKA and dehydration; subcutaneous insulin is erratically absorbed in this state and should not be used in this patient. These patients need intravenous insulin infusions. The use of an intravenous bolus in addition to a continuous intravenous infusion is controversial. In patients with DKA, the SERUM potassium level may be high, normal, or low, but all of these patients all have total body potassium depletion. Unless a patient is initially hyperkalemic, potassium should be administered to all patients in DKA, as the serum potassium level inevitably goes down with the administration of insulin and correction of the acidosis. Most patients have adequate respiratory compensation for their metabolic acidosis. Intubation for hyperventilation therapy is not indicated and is associated with worsened outcomes in patients with DKA and cerebral edema. (Marcin JP, Glaser N, Barnett P, et al. Factors associated with adverse outcomes in children with diabetic ketoacidosis-related cerebral edema. *J Pediatr*. 2002; 141: 793-797.)

2. d. The hyperglycemia in patients with DKA often corrects before the acidosis. Insulin is required to stop ongoing ketone body formation (and concomitant acidosis), so the insulin infusion needs to continue until the acidosis is corrected. Therefore, the patient's maintenance fluids should be converted to dextrose-containing solutions (eg, D5/0.45NS) when the blood sugar falls to 250-300 mg/dL.

3. b. The most dreaded complication of DKA is cerebral edema. This is seen more commonly in children than in adults and is the leading cause of death in children with DKA. The mortality rate from cerebral edema is high and many survivors are left with permanent neurologic sequelae. A recent multi-center, case-control study identified a high blood urea nitrogen level, low partial pressure of arterial carbon dioxide, and the use of bicarbonate as risk factors associated with (but not necessarily the causative factors of) the development of cerebral edema (Glaser N, Barnett P, McCaslin I, et *al*. Risk factors for cerebral edema in children with diabetic ketoacidosis. *N Engl J Med*. 2001; 344: 264-9.). Treatment with mannitol needs to be immediate when cerebral edema is suspected clinically and should not be delayed while awaiting the results of imaging studies.

4. c. Children are prone to hypoglycemia because they have increased glucose demands and decreased availability of glycogen and gluconeogenic precursors than adults. Because of the relative lack of glycogen reserves in the liver, glucagon is less effective in children than adults, as the effectiveness of glucagon is dependent on the availability of glycogen for glycogenolysis and subsequent glucose production. The most common cause of hypoglycemia in previously healthy children presenting to the ED is idiopathic ketotic hypoglycemia (Pershad J, Monroe K, Atchison J. Childhood hypoglycemia in an urban emergency department: epidemiology and a diagnostic approach to the

problem. *Pediatr Emerg Care*. 1998; 14(4): 268-271. Daly LP, Osterhoudt KC, Weinzimer SA. Presenting features of idiopathic ketotic hypoglycemia. *J Emerg Med*, 2003; 25(1): 39-43.). This is characterized by symptomatic hypoglycemia associated with ketonuria in previously healthy young children after a moderate fast with resolution of symptoms after glucose administration. Children with hypoglycemia need rapid correction of their hypoglycemia. However, $D_{50}W$ is an extremely hyperosmolar solution and can lead to significant irritation and extravasation injury, especially in children with small veins. Therefore, in younger children, 2-4 cc/kg of $D_{25}W$ or 5-10cc/kg of D10W is preferred over $D_{50}W$.

5. a. This is a patient with congenital adrenal hyperplasia (CAH). In most forms of CAH, an enzymatic defect results in the blockage of cortisol or aldosterone synthesis and the subsequent excessive formation of steroid precursors. The most common form of CAH (90%) results from a deficiency of 21-hydroxylase. Because this deficiency results in accumulation of androgenic compounds, girls are virilized in utero and are born with ambiguous genitalia. Children commonly develop salt-wasting crises between two and five weeks of age. The clinical presentation is usually non-specific and is often manifested by lethargy, irritability, vomiting, poor weight gain, and dehydration. The diagnosis should be considered when a child has hyponatremia and hyperkalemia. Dehydrated patients should be resuscitated with normal saline; hypoglycemic patients will need dextrose containing fluids as well. Glucocorticoid replacement, usually with hydrocortisone (50-100 mg/m^2 or 1-2 mg/kg IV as a bolus followed by 12.5-25 mg/m^2 IV q6h) needs to be given immediately; patients do not need mineralocorticoid replacement on an emergent basis. When possible, blood should be obtained for steroid profile testing before treatment with glucocorticoids, as these agents can suppress production of precursor steroids within hours; however, treatment should not be delayed to help facilitate a diagnosis.

6. d. Female pubertal development typically begins with the development of breast buds at 11.15 years of age and the development of pubic hair at 11.69 years of age. (Boys first start developing pubic hair at age 13.90 years). On average, girls experience menarche at 13.47 years of age. Primary amenorrhea is commonly defined as an absence of menses by age 16 years with normal secondary sex characteristics, by age 14 years in the absence of any pubertal development, or 4 years after the onset of puberty. This 16 year old girl has fully developed breasts and pubic hair, and so her amenorrhea abnormal. While there are many causes of primary amenorrhea, the abdominal mass and the monthly cyclic pain in this patient is suggestive of hematocolpos (accumulation of fluid in the vagina) or hydrometrocolpos (accumulation of fluid in the vagina and uterus). This condition results from obstruction of the vaginal, causing fluid to accumulate. Adolescent girls usually present with primary amenorrhea and /or abdominal pain with otherwise normal pubertal development. This pain is frequently worsened by menses, when additional blood accumulates (although the pain is not necessarily cyclic). Examination of the introitus often reveals a bulging, imperforate hymen. Hydrometrocolpos is usually treated operatively. Malignant ovarian tumors are uncommon and account for only about 1% of malignant tumors in girls less than 17 years old. This girl needs a pregnancy test - if you think all teenagers who deny sexual activity are really not sexually active, you probably also believe that the drunk college student lying in your hallway gurney truly drank only "two beers" and the gang member in your trauma bay who was shot three times was honestly "minding my own business."

7. b. This patient has the syndrome of inappropriate antidiuretic hormone (SIADH) secretion. Normally, antidiuretic hormone (ADH) secretion is stimulated by a rise in serum osmolality and a decrease in plasma volume. Excessive secretion of ADH in the setting of normal or increased plasma volume is inappropriate because ADH causes further water retention and resultant decrease in serum osmolality. While there are many causes of SIADH, this is caused most commonly by central nervous system disorders, pulmonary disease, and medications. Clinical manifestations of SIADH are primarily those

associated with hyponatremia: headaches, nausea and vomiting, changes in mental status, seizures, and coma. These patients are euvolemic or slightly hypervolemic and should not have signs of dehydration. Laboratory studies reveal hyponatremia and low serum osmolality, as well as a high urine osmolality and urine sodium concentration. Treatment for mild cases of SIADH is fluid restriction. In patients who are seizing, hypertonic saline should be administered until the seizures stop. However, overly rapid correction may lead to central pontine myelinosis; therefore once the patient's neurologic condition has stabilized, further correction to the normal range of normal should occur over a longer period of time (e.g., 10% increase of serum sodium over 24 hours).

8. a. This child most likely has rickets, a constellation of symptoms characterized by inadequate mineralization of the bone matrix. Although there are many causes of rickets, the most common cause is inadequate dietary intake of vitamin D. Children at risk include malnourished children, vegetarians, premature infants, children who are not exposed to sunlight (important in vitamin D synthesis), children who are on certain medications (e.g., furosemide, phenytoin or phenobarbital) and patients with malabsorptive or renal disease. This inadequate bone mineralization is most clinically evident in areas of rapid bone growth, leading to widening and irregularities of the epiphyses and metaphyses of growing bones. This irregular growth bone growth frequently leads to the knobby chest wall ("rachitic rosary") and splaying and flaring of the wrists. In the ED, x-rays reveal these characteristic bone irregularities. In vitamin D-deficiency rickets, the calcium and phosphate levels are normal or low, and treatment is with oral vitamin D supplements. In severe cases, calcium therapy may need to be initiated before starting vitamin D. While rickets is the most likely diagnosis in this patient, non-accidental trauma should always be considered in a child with seizures and extremity pain. An otherwise non-toxic appearing child is not likely to have seizures as a result of meningitis. Tuberous sclerosis is associated with nontender hypopigmented macules, intracranial tumors, and ocular lesions.

9. c. Neonatal thyrotoxicosis is almost always seen in children whose mothers have Graves' Disease. Overall, however, the incidence of hyperthyroidism in children is relatively low. The vast majority of pediatric patients with hyperthyroidism have Graves' Disease. The majority of patients with congenital hypothyroidism are detected on universal neonatal screening tests since most have subtle clinical signs and symptoms. More severely affected neonates have are hypoactive, feed poorly, and are frequently constipated. These infants are frequently hypothermic, jaundiced, and have large posterior fontanelles. They later develop coarse facies and enlarged tongues if left untreated. Treatment of thyroid storm includes beta-blockers in order to minimize sympathomimetic symptoms, antithyroid medications (e.g., propylthiouracil) FOLLOWED by iodine so that the iodine is not incorporated into the synthesis of new hormone, glucocorticoids (which decreases the peripheral conversion of T4 to T3), and cooling measures for hyperthermia.

10. d. A white discharge from a neonate's nipples (commonly known as "witch's milk") is normal and is due to the withdrawal from maternal estrogen. This is seen in both boys and girls and usually resolves within the first two weeks of life. Neonates can develop mastitis, which can cause purulent drainage from the nipple; however, this patient's clinical presentation is not consistent with this disease. While CNS tumors (e.g., pituitary adenomas) and breast cancers can cause breast discharge, these entities are exceedingly rare in neonates.