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QUESTION 1

A 34-year-old woman with a history of type 1 diabetes mellitus presents to your office for a routine follow-up visit. She is feeling well and has no complaints. Her fasting blood sugars usually run 140-160 and her HgbA1C was recently measured at 8.2. She tells you that she would like to become pregnant but wants to know if there are any risks for her and a baby due to her diabetes.

Which of the following preconception counseling statements is true?

- A. All diabetics planning to become pregnant should be placed on ACE inhibitors for renal protection.
- B. Diabetic women should not take folic acid because all commercially available supplements contain sugar.
- C. The goal HgbA1C level during her pregnancy is approximately 9%.
- D. Insulin pump treatment is contraindicated during pregnancy.
- E. Women with good preconception diabetic control have infants with a lower incidence of congenital malformations than women with poor preconception diabetic control.

Correct Answer: E Section: (none)

Explanation: Explanations: Pregestational diabetes is associated with numerous risks to both the mother and the fetus. Stillbirths are more common in pregnancies to diabetic women and stillbirths without an identifiable cause, called "unexplained" stillbirths, are a well-described phenomenon. Similarly, preterm births are more common in diabetics than nondiabetics. While congenital malformations are more common in pregnancies to diabetic women, fetal chromosomal abnormalities are not more common. Children of women with diabetes have an approximately 13% incidence of developing type 1 diabetes. While earlier obstetrical teaching suggested that maternal diabetes delayed fetal lung maturation, more recent studies do not support this. Gestational age is likely the most significant factor in the development of respiratory distress. While there are significant maternal risks from the interaction of diabetes and pregnancy, with the possible exception of diabetic retinopathy, the long-term course of diabetes does not appear to be affected by pregnancy.

Pregnancy neither exacerbates nor modifies diabetic nephropathy and the development of diabetic peripheral neuropathy during pregnancy is uncommon. While preeclampsia is a significant risk and the perinatal mortality rate is 20 times higher in preeclamptic diabetic women compared to normotensive women, the occurrence of preeclampsia does not appear to be related to diabetic control. Diabetic ketoacidosis is a serious complication with an approximately 20% rate of fetal loss. However, it is estimated to occur in 1% of pregnancies of diabetic women. Infections occur in approximately 80% of pregnancies in insulin-dependent diabetics, with candida vaginitis, urinary tract infections, and respiratory infections being common. Preconception counseling in diabetic women who desire to become pregnant is a critical issue that often is best served by a team that includes the obstetrician, primary care physician, endocrinologist, and diabetic educators. When possible, attempts should be made to attain optimal diabetic control. Women with good diabetic control have been shown in observational studies to have a lower rate of having infants with congenital anomalies than women with poorer diabetic control. Optimal diabetic control has been defined as glycated hemoglobin levels within or near the upper limit of the normal range. This can be obtained with multiple daily insulin injections or, in selected patients, a continuous infusion via an insulin pump. All women--diabetic or not should be counseled to take folic acid prior to conception in order to lower the rate of neural tube defects. ACE inhibitors are contraindicated during pregnancy and should, whenever possible, be discontinued prior to conception.

QUESTION 2



A 31/2-year-old female presented with a left upper quadrant abdominal mass. The child had no previous history of medical illnesses. An ultrasound examination revealed a markedly deformed left kidney with 12 cm nonhomogenous soft tissue mass arising from the upper pole. Medial displacement of the bowel loops

was also noted.

Characteristically, Wilms tumors are histologically recognizable for which of the following?

- A. classic triphasic combination of blastema, stromal, and epithelial cells
- B. epithelial elements alone
- C. blastemic elements
- D. focal keratinization
- E. glandular formation

Correct Answer: A Section: (none)

Explanation:

Wilms tumor is the most common primary renal tumor in childhood, usually diagnosed between the ages of 2 and 5. The risk of Wilms tumor is increased in association with at least three recognizable groups of congenital malformations exhibiting alteration in at least two distinct chromosomal loci. A few familial cases of Wilms tumor not associated with identifiable lesions or mutations involving either the WT-1 or the WT-2 gene suggest that there may be another locus that plays a role in some tumors, but that still remains unknown. Wilms tumor presents as a large solitary mass and in 10% of cases may be bilateral. Microscopically, the Wilms tumor is characterized by recognizable attempts to recapitulate different stages of nephrogenesis. The classic triphasic combination of blastemic, stromal, and epithelial cell types is observed in the majority of the lesions. Occasional skeletal muscle differentiation can be seen, as well as squamous, mucinous epithelium, cartilage, or bone. The combined therapy of chemo, radiation, and surgery has dramatically improved the results of long-term survival in these patients, up to 90%

QUESTION 3

Which of the following conditions usually causes hypoglycemia at birth?

- A. Sturge-Weber syndrome
- B. neurofibromatosis, type 1 (von Recklinghausen disease)
- C. tuberous sclerosis
- D. CHARGE association
- E. Beckwith-Wiedemann syndrome

Correct Answer: E Section: (none)

Explanation:



Of the listed syndromes, only Beckwith Wiedemann syndrome has neonatal hypoglycemia as part of its clinical spectrum. The constellation of macroglossia, hypoglycemia, and visceral organomegaly (hepatosplenomegaly) is a common finding in children with Beckwith-Wiedemann syndrome. The presence of an omphalocele in a newborn would also be concerning for Beckwith-Wiedemann syndrome.

QUESTION 4

A 13-year-old boy is brought into the emergency room with a laceration of his right arm. According to his parents, he received the injury when he fell on the ground while playing at the family farm about 1 hour ago. He has no known history of any medical problems. His parents say that they haven't brought him to the doctor in years. On questioning, they report that he only received one of his "baby shots" and they are not sure which one that was. On examination, he is healthy appearing. He is appropriately apprehensive but calm and consolable. His right arm has a 5 cm linear laceration with visible soil particles in and about the wound. The remainder of his examination is unremarkable. You carefully clean and irrigate the wound and then primarily repair the laceration with sutures.

What immediate tetanus prophylaxis would be optimal in this case?

- A. IM injection of adult Td vaccine only
- B. IM injection of both adult Td vaccine and tetanus immune globulin (TIG)
- C. IM injection of Tdap only
- D. IM injection of TIG only
- E. IM injection of both Tdap and TIG

Correct Answer: E Section: (none)

Explanation: Explanations: The disease tetanus is caused by an exotoxin produced by the anaerobic, gram-positive bacterium *C. tetani*. The spores of *C. tetani* are endemic in soil, particularly in agricultural areas. They can also be found in the intestines and feces of many animals. Human infection usually is the result of the introduction of the spores through a wound, such as a puncture or laceration. The spores can then germinate and toxins are released. Tetanus is characterized by unopposed muscle contractions and spasms. Autonomic nervous system manifestations, seizures, and difficulty swallowing may occur. Recovery may take months, but the disease is often fatal. In the developed world, most cases of tetanus occur in those who either were never vaccinated or who completed a primary vaccine series but have not had a booster in the preceding 10 years. The currently available vaccine is a toxoid which consists of a formaldehyde-treated toxin. It is available as a single antigen vaccine, combined with diphtheria (pediatric DT or adult Td) or combined with both diphtheria and acellular pertussis vaccine (DTaP). Whenever possible, tetanus toxoid should be given in combination with diphtheria toxoid to provide periodic boosting for both antigens. There is little reason to use single antigen tetanus toxoid alone. Management of a potentially contaminated wound initially involves local wound care. Necrotic tissue should be debrided, foreign material removed, and the wound irrigated. The need for active and/or passive immunization against tetanus depends on the wound and the patient's history of immunization. A person who has completed a primary series of three or more doses of tetanus toxoid vaccine will not require passive immunization with TIG, but may require a booster of dT or Tdap. For a clean, minor wound, a Td or Tdap booster would be indicated if it has been more than 10 years since the patient's most recent booster. For all other wounds, a booster would be indicated if it has been 5 years since the most recent booster. In a person who has not completed a primary series, who is completely unimmunized, or in whom the vaccine status is unknown, initiating passive immunization with Td or Tdap is indicated for all wounds. If the wound is clean and minor then TIG would not be recommended. For all other wounds, both Td and TIG would be indicated, as the initial doses of Td/Tdap may not produce immunity and TIG can provide immediate, temporary immunity. Antibiotic prophylaxis against tetanus is not useful. As noted in explanation 9 (above), Tdap is recommended as a substitute for a single Td dose in order to address the increasing rates of pertussis being encountered in the population. As the patient in question 32 has no history of having completed a primary vaccine series



and has a contaminated wound, the optimal management would be to provide both Tdap and TIG. If Tdap were not available, then utilizing Td and TIG would be an acceptable substitute, with a dose of Tdap to be given as part of his "catchup" series in the future.

QUESTION 5

A 42-year-old male with extensive Crohn's disease undergoes a near complete resection of the ileum. A deficiency of which of the following vitamin is likely to result?

- A. niacin
- B. thiamine
- C. vitamin B12
- D. vitamin C
- E. vitamin B6

Correct Answer: C Section: (none)

Explanation:

The distal small bowel (ileum) is the site of absorption of fat-soluble vitamins (vitamins A, D, E, and K) as well as vitamin B12. Vitamin B12 binds with intrinsic factor, a glycoprotein secreted from parietal cells of the gastric fundus and body. Specific receptors in the terminal ileum take up the B12 intrinsic factor complex. Vitamin B12 deficiency leads to megaloblastic anemia. The patient will require monthly vitamin B12 injections

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