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QUESTION 1

Loss of the hepatic protein hepcidin can lead to severe iron overload with symptoms resembling those of hemochromatosis. Which of the following functions of hepcidin accounts for the iron overload when the protein is deficient?

A. activates the expression of the ironresponse element-binding protein that regulates transferring receptor and ferritin mRNA translation

B. decreases the level of intestinal membrane iron transporters, resulting in reduced iron uptake

C. facilitation of the interaction of transferring with the transferrin receptor

D. forms a complex with ferritin allowing for higher intracellular storage

E. promotes the formation of hemosiderin, thus detoxifying iron

Correct Answer: B

Section: Biochemistry Hepcidin is a hepatically synthesized iron regulatory protein that functions by inhibiting the presentation of one or more of the iron transporters [e.g., DMT1 and Ireg1 (ferroportin)] in intestinal membranes. With a high iron diet, the level of hepcidin mRNA increases and conversely its levels decrease when dietary iron is low. This is occurring simultaneous to reciprocal changes in the levels of the transporters proteins themselves. Loss of hepcidin activity would then lead to unregulated iron uptake from the intestines leading to iron overload. In fact, it is now considered that defects in hepcidin function contribute to the development of hemochromatosis. Hepcidin does not activate expression of iron-response elementbinding protein (choice A), facilitate transferring interaction with the transferring receptor (choice C), form a complex with ferritin (choice D), nor promote the formation of hemosiderin (choice E).

QUESTION 2

A 6-year-old girl has been having recurrent pyogenic bacterial infections of the sinus and respiratory tract. The latest one has been caused by N. meningitidis. The results of her diagnostic tests are shown in below figure.

TEST	RESULT
White cell count	Normal
T cell count	Normal
Production of IgM to ploysaccharides	Normal
Production of anti-DNA antibodies	Not detected
Levels of complement componets C3 and C5 to C8	Low
Levels of thyroid-stimulating hormone	Normal
Levels of immunoglobulins	Normal
Intracellular killing of microbes by neutrophils	Normal

These results are consistent with a diagnosis of which of the following?

A. chronic granulomatous disease

B. deficiency in the opsonization of microbes by phagocytes

C. Graves disease

D. systemic lupus erythematosus

E. Wiskott-Aldrich syndrome

Correct Answer: B

Section: Microbiology/Immunology The diagnostic tests ordered by the physician are consistent with a deficiency in the C3, C5C8 components of complement. Individuals with these deficiencies have recurrent pyogenic infections, and show enhanced susceptibility to meningococcal infections. All other choices are not consistent with the laboratory findings. Chronic granulomatous disease (choice A) entails a defect in the intracellular killing of microbes by neutrophils. Graves disease (choice C) involves autoantibody production to thyroid-stimulating hormone receptors. Systemic lupus erythematosus (choice D) involves production of antinuclear antibodies (ANA). Wiskott-Aldrich syndrome (choice E) is associated with recurrent pyogenic infections, but is due to an inability of plasma cells to produce IgM against bacterial polysaccharides, and it occurs only in male infants.

QUESTION 3

In patients with type 4 renal tubular acidosis (RTA) aldosterone deficiency is often a prominent finding. Distal tubular transport of which of the following ions will be affected in these patients?

A. hydrogen and potassium in exchange for sodium

B. hydrogen only

C. potassium only

D. sodium and bicarbonate

E. sodium only

Correct Answer: A

Section: Physiology The RTArefers to related conditions that are disorders of urine acidification even though other renal functions are not impaired. In type 4 RTA, distal nephron dysfunction is due to either inadequate aldosterone production or aldosterone resistance resulting from intrinsic renal disease. Thus, patients develop hyperchloremic acidosis with hyperkalemia, due to impaired distal tubular secretion of both potassium and hydrogen ions. Treatment of patients is directed at controlling serum potassium. Choices B, C, D, and E are inconsistent with the known actions of aldosterone.

QUESTION 4

After an abdominal surgery for removal of ovarian cysts, a 56-year-old female has had low-grade fever for the past 2 weeks. She has a history of rheumatic fever as a child. Three of the blood cultures grew gram-positive cocci. Which of the following is the most likely etiologic agent?

A. Group A streptococci

B. group B streptococci

C. group C streptococci

D. group D streptococci

E. S. viridans

Correct Answer: D

Section: Microbiology/Immunology There are at least 12 species of enterococci, and all are classified as group D streptococci (choice D). E. faecalis is the most common and causes 8590% of enterococcal infections. As long as enterococci remain in the intestinal tract, they are seldom involved in disease. Yet, they are extremely efficient opportunists. The enterococci are among the most frequent causes of nosocomial infections, especially in intensive care units. They are commonly found in complications of intra-abdominal surgery, often as part of a mixed culture of microorganisms. Antibiotic therapy (cephalosporins, etc) is often a selective feature since enterococci are resistant to many of the antimicrobials in common use. Group Astreptococci (choice A) would be represented by S. pyogenes and is the group which causes about 95% of human infections (skin, septicemia, organs, etc), but would seldom be found in intra-abdominal complications. Streptococci B (choice B) are most often found in infections of infants since type B streptococci are found routinely in the vaginal flora. Group C streptococci (choice C) usually occur in the nasopharynx and may cause sinus, blood, or heat infections. S. viridans (choice E) are usually found in the oral cavity and may be opportunists involving blood and heart infections.

QUESTION 5

A 3-year-old girl develops gastroenteritis and a number of other children at the day care nursery that she attends are also affected. This was most likely caused by which of the following agents?

A. Balantidium coli

B. Cryptosporidium parvum

C. Entamoeba histolytica

D. Giardia lamblia

E. rotavirus

Correct Answer: E

Section: Pathology and Path physiology Rotavirus is the most common cause of severe diarrhea worldwide and is most widespread in very young children. It is passed readily to others and can result in outbreaks such as described here. B. coli (choice A) is a ciliated intestinal protozoal parasite that is found mostly in the tropics. It is a rare cause of diarrhea in the United States. C. parvum (choice B) is a spore-forming protozoan that can cause severe diarrhea in immunocompromised individuals. E. histolytica (choice C) is another protozoan parasite. It infests the colon forming flask-shaped ulcers (i.e., having narrow neck and a wide base that undermines the adjacent mucosa). These organisms can also be carried in the portal circulation to the liver where they form characteristic "anchovy paste" abscesses. G. lamblia (choice D) is an intestinal flagellate that infests the upper small intestine. It is very common worldwide and is the most common intestinal protozoan parasite in the Western world. The majority of infected individuals remain asymptomatic but it can cause diarrhea that is usually mild but can be severe.

QUESTION 6

An overworked surgical resident is resecting an acinic cell carcinoma from the inferior aspect of the parotid gland when he makes a careless incision and nicks a branch of the facial (seventh cranial) nerve. Which of the following muscles may be affected by this injury?

A. omohyoid

B. sternohyoid

C. sternothyroid

D. stylohyoid

E. thyrohyoid

Correct Answer: D

Section: Anatomy The stylohyoid is innervated by the facial (seventh cranial) nerve. During the embryonic period, the muscles developing from the second pharyngeal arch include the stylohyoid, stapedius, posterior belly of the digastric, auricular muscles, and muscles of facial expression. The nerve supplying the second pharyngeal arch, and thus all these muscles, is the facial (seventh cranial) nerve. All the other muscles, omohyoid (choice A), sternohyoid (choice B), sternothyroid (choice C), and thyrohyoid (choice E) form the infrahyoid muscle. They are supplied as a group by branches of the ansa cervicalis derived from C1, C2, and C3.

QUESTION 7

A 57-year-old woman had her last menstrual period at the age of 46. However, for the past 4 months she has experienced intermittent vaginal hemorrhage. A right ovarian mass is identified. Which of the following is the most likely diagnosis?

A. arrhenoblastoma

B. Brenner tumor

C. dysgerminoma

D. granulosa cell tumor

E. Sertoli-Leydig cell tumor

F. teratoma

Correct Answer: D

Section: Pathology and Path physiology An ovarian mass raises the possibility of a tumor and vaginal hemorrhage in a postmenopausal woman and suggests an estrogenproducing tumor. One of the first estrogenproducing tumors to consider is granulosa cell tumor. Arrhenoblastoma (Sertoli-Leydig cell tumor) (choice A) secretes androgens, not estrogens. Brenner tumor (choice B) is an uncommon ovarian neoplasm without hormonal activity. Dysgerminoma (choice C) is the female counterpart of the male seminoma and does not produce hormones. Sertoli-Leydig cell tumor (choice E) is a synonym for arrhenoblastoma (choice A). Teratoma (choice F) is also hormonally inactive.

QUESTION 8

Your patient presents in your office complaining of hoarseness. During your examination, you find that one vocal fold has deviated toward the midline and does not abduct during deep inspiration or vocalization. You also observe that touch sensation in the vestibule of the larynx appears to be intact.

You suspect that a nerve has been damaged, but which nerve is most likely involved?

A. external laryngeal

B. glossopharyngeal

C. inferior laryngeal

D. internal laryngeal

E. superior laryngeal

Correct Answer: C

Section: Anatomy The inferior laryngeal nerve is motor to the intrinsic muscles of the larynx except the cricothyroid muscle, and is sensory to the larynx below the level of the vocal fold. The external laryngeal nerve (choice A) is the branch of the superior laryngeal nerve that contains the motor fibers to the cricothyroid muscle and to the cricopharyngeus muscle. The glossopharyngeal nerve (choice B) does not supply either motor or sensory fibers to the larynx. The internal laryngeal nerve (choice D) is the branch of the superior laryngeal nerve that contains the sensory fibers to the laryngeal mucosa above the vocal fold. The superior laryngeal nerve (choice E) is sensory to the laryngeal mucosa above the vocal fold and also includes motor fibers to the cricothyroid muscle.

QUESTION 9

Which of the following is the most treatable dementing disease?

A. Alzheimer\\'s disease

B. Creutzfeldt-Jakob disease

C. multi-infarct dementia

D. Pick disease

E. Wernicke syndrome

Correct Answer: E

Section: Behavioral Science and Biostatics Wernicke syndrome is caused by thiamine deficiency associated with alcohol withdrawal, and can be treated effectively. Alzheimer\\'s disease (choice A) is the most common primary dementia in the elderly. About 50% of brains of demented patients show evidence of Alzheimer\\'s disease. Creutzfeldt-Jakob (choice B) disease and kuru are slow virus infections that show clinical and histologic features similar to Alzheimer\\'s disease. Multiinfarct dementia (choice C) is secondary to repeated cerebrovascular accidents (CVAs) in patients with underlying atherosclerosis or hypertension. It is more common among men than women. Pick disease (choice D) is a rare cause of cortical dementia. The presence of unilateral cortical atrophy in Pick disease contrasts with the bilateral atrophy evident in Alzheimer\\'s disease. Microscopically, Pick bodies composed of densely packed neurofilaments are seen within diseased neurons.

QUESTION 10

Which of the following statements concerning total body energy storage is correct?

A. Most of the body\\'s energy store is held as carbohydrate.

B. Most of the body\\'s energy store is held as lipid.

C. Most of the body\\'s energy store is held as plasma glucose.

D. Most of the body\\'s energy store is held as protein.

E. Total body\\'s energy storage approximately equals resting metabolic rate.

Correct Answer: B

Section: Physiology Lipid is the most concentrated form of energy storage, holding 9.4 kcal/g. For a typical 70 kg human over 130,000 kcal is typically stored as fat. Storage of energy as available protein (choice D) is about 20,000 kcal, while storage as carbohydrate (choice A) is about 3,000 kcal. Though blood glucose (choice C) is critically important, in particular as a source of energy for the brain, blood glucose does not provide a significant fraction of total body energy storage. While total body energy stores typically exceed 150,000 kcal, resting metabolic rate (depending on age and body mass) is about 2100 kcal/day (choice E).

QUESTION 11

Which of the following receptor types is associated with the most rapid response time?

A. G-protein-coupled receptors (e.g., muscarinic receptors)

B. ion channel receptors (e.g., nicotinic receptors)

C. Jak-STAT receptors (e.g., for cytokines)

D. steroid receptors (e.g., for corticosteroids) E. transmembrane enzyme receptors (e.g., for insulin)

Correct Answer: B

Section: Pharmacology Response times for ion channel receptors are measured in milliseconds, for G proteincoupled receptors (choice A), transmembrane enzyme receptors (choice E), and Jak-STAT receptors (choice C) in seconds to minutes, and for steroid receptors (choice D) in hours or days.

QUESTION 12

Which of the following personality traits would most likely result from anal-phase fixation?

A. dependency

B. fierce competitiveness

C. parsimony

D. narcissism

Correct Answer: C

Section: Behavioral Science and Biostatics The anal phase of development occurs around age 2, around the time of toilet training. Developmental difficulty around this time may result in "anal-phase fixation," in which some characteristics of this phase may unduly influence the adult personality. Such characteristics include struggles over the issue of control.

Excessive cleanliness and orderliness, or the opposite, as well as parsimony and hoarding may occur. Dependency (choice A) is characteristic of oral phase, and fierce competitiveness (choice B) may be characteristic of the Oedipal phase. Narcissism (choice D) is a complex phenomenon that develops from very early life and not confined to any

particular stage of development.