1. **Regarding pathogenesis of genetically determined cholestatic disorders of the liver the following are TRUE:**
 1. Inheritance of progressive familial intrahepatic cholestasis (PFIC) is autosomal recessive
 2. PFIC 1 is due to mutations in ABCB 11 gene encoding for bile salt excretory protein (BSEP)
 3. BSEP defect results in accumulation of bile salts in the hepatocyte with consequent hepatocellular damage
 4. Benign recurrent intrahepatic cholestasis (BRIC) can occur with mutations on the same genes that cause PFIC
 5. Intrahepatic cholestasis of pregnancy and drug induced cholestasis are not related to these genetic mutations
2. **Regarding the clinical features and diagnosis of PFIC the following are TRUE EXCEPT:**
 1. PFIC 2 is the most severe form with highest propensity to develop liver failure, portal hypertension and hepatocellular carcinoma
 2. Majority of patients with PFIC 3 will present within first 3 months of life
 3. Extrahepatic manifestations are most common in PFIC 3
 4. PFIC 1 and 2 have normal gamma glutamyl transpeptidase levels
 5. Serum alpha-fetoprotein levels are raised in PFIC 2
3. **Regarding treatment of PFIC the following are TRUE:**
 1. Ursodeoxycholic acid (UDCA) is useful in PFIC
 2. Cholestyramine and rifampicin are the drugs of choice in PFIC
 3. Biliary diversion procedures are useful to delay histological progression in patients who do not respond to medical therapy
 4. Ileal bypass is the biliary diversion surgical procedure of choice in PFIC
 5. PFIC can recur after liver transplantation
4. **Regarding the pathogenesis of Gaucher disease the following are TRUE:**
 1. It is an autosomal dominant disease
 2. It results from deficiency of the lysosomal enzyme glucocerebrosidase
 3. Accumulation of glucosylsphingosine correlates with severity of neuronopathic disease
 4. N370S substitution is associated with neurological disease
 5. There is an absolute genotype-phenotype correlation in Gaucher disease
5. **Regarding the clinical features and diagnosis of Gaucher disease the following are TRUE EXCEPT:**
 1. Type 1 Gaucher disease does not have neurological involvement
 2. Type 3 is the acute neuronopathic form of Gaucher disease
 3. Spleen is enlarged disproportionately more than the liver
 4. Cirrhosis is a common presenting feature
 5. Histopathological examination of bone marrow or spleen is the gold standard for diagnosis
6. **Regarding management of Gaucher disease the following are TRUE:**
 1. Bone marrow transplantation is the treatment of choice
 2. Enzyme replacement therapy (ERT) is most useful in type 2 Gaucher disease
 3. ERT does not treat lung parenchymal disease and pulmonary hypertension
 4. Cirrhosis or advanced fibrosis is not reversible with ERT
 5. Drugs targeted at inhibition of glucosylceramide synthesis are useful for induction therapy
7. **Regarding pathogenesis and presentation of portal cavernoma cholangiopathy (PCC) the following are TRUE EXCEPT:**
 1. Biliary system is supplied solely by the hepatic arterial system
 2. PCC may reverse only partially after shunt surgery
 3. Only minority of patients with PCC are symptomatic
 4. Majority of patients with PCC become symptomatic at a young age, soon after the diagnosis of portal cavernoma
 5. PCC term is applicable to biliary changes associated with portal hypertension of any etiology
8. **Regarding management of PCC the following are TRUE:**
 1. Magnetic resonance cholangiography (MRC) is the diagnostic modality of choice
 2. Therapeutic endoscopic retrograde cholangiography (ERC) is required in all cases
 3. Majority of patients with symptomatic PCC cannot undergo shunt surgery due to absence of suitable veins
 4. Majority of patients of symptomatic PCC become asymptomatic after successful portosystemic shunt procedure
 5. Liver transplantation is not feasible in PCC
9. **Regarding hepatic encephalopathy (HE) the following are TRUE EXCEPT:**
 1. Normal neurological examination and normal psychometric testing is characteristic of minimal HE

<http://dx.doi.org/10.1016/j.jceh.2014.05.010>

2. The term covert HE includes minimal HE and grade I HE
3. Covert HE does not need to be treated
4. Magnetic resonance spectroscopy findings of HE are characterized by increased intracellular glutamate/glutamine signal and a decrease in myoinositol, taurine, and choline signals
5. Lactulose acts solely by its laxative effect in HE

10. Regarding non-alcoholic fatty liver disease (NAFLD) the following are TRUE:

1. Asians origin individuals are at higher risk for developing NAFLD irrespective of their current place of residence
2. The association of patatin-like phospholipase domain-containing protein 3 (PNPLA3) gene polymorphisms with NAFLD has been demonstrated for Caucasians only
3. Only a minority of patients will be able to achieve desired weight loss despite intensive lifestyle modification programs.

4. Weight loss medications are not superior to lifestyle modification programs in achieving desired weight loss
5. Transient elastography is unreliable in predicting fibrosis in NAFLD

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(Answers on page 184)