THE COLLEGES OF MEDICINE OF SOUTH AFRICA

Incorporated Association not for gain
Reg No 1955/000003/08

Examination for the Subspeciality Certificate in Gastroenterology
of the College of Paediatricians of South Africa

2 March 2017

Paper 1 (3 hours)

All questions to be answered. Each question to be answered in a separate book (or books, if more than one is required for the answer)

Please note the meaning of the following terms:
(i) 'List' means to enumerate i.e. specify a list or an outline form of reply in concise form.
(ii) 'Describe' means to answer in a brief narrative form.
(iii) 'Discuss' means that you need to analyse carefully, and construct an answer that provides an overview of the various factors that may contribute to a particular problem or treatment - this type of question calls for a more complete response.

Question 1

A severely obese 15-year-old boy is referred to you for further management of constipation.

a) On physical examination, what clinical features are suggestive of organic disease instead of functional constipation? (5)
b) What organic causes of constipation need to be excluded in this teenager? (5)
c) List six orally-ingested pharmacologic agents that may be used as maintenance therapy (not disimpaction) for constipation. (3)

On history, the mother remembered that the boy "screamed, cried, and would turn red or purple in the face with each effort to defaecate" when he was a young infant.

d) i) What is infantile dyschezia? (2)
ii) Briefly describe the natural history of infantile dyschezia and the recommended treatment. (2)

The boy's mother also reports that he frequently complains about "heartburn" especially when lying down.

e) List complications of gastro-oesophageal reflux disease (GERD) in adolescents. (5)
f) Briefly describe the management of GERD in adolescents under the following headings
i) Lifestyle changes. (3)
ii) Pharmacologic management. (5)
g) Except for functional constipation and GERD, list four other gastrointestinal diseases associated with obesity. (2)

PTO/Page 2 Question 1h)…

As part of his evaluation, liver function tests (LFT) were performed because the boy complained of right upper quadrant pain and hepatomegaly was noted on examination. LFT results revealed elevated levels of liver transaminases (alanine aminotransferase [ALT], and aspartate aminotransferase [AST]), alkaline phosphatase, and gamma glutamyl transpeptidase (GGT). A liver biopsy revealed steatosis and inflammation.

h) What do you understand by the term "Non-alcoholic fatty liver disease (NAFLD)"? (3)

The mother reports that her son has had major difficulty in losing weight and makes enquires about bariatric surgery for her son.

i) List the medical indications for bariatric surgery in obese children. (5)

[40]

Question 2

A 4-year-old HIV-uninfected girl presents with acute diarrhoea and requires hospitalisation. An infectious gastroenteritis is suspected.

a) List ten common infectious agents responsible for
   i) Acute watery diarrhoea.
   ii) Invasive (bloody) diarrhoea. (5)

The child is given oral rehydration therapy. The diarrhoea, however, continues unabated for more than 14 days.

b) Briefly discuss the pathogenesis of persistent diarrhoea triggered by infection. (5)

The child receives two courses of broad-spectrum antibiotics but the diarrhoea persists. Enteral nutrition is commenced. At this stage, infection due to *Clostridium difficile* is suspected.

c) List the risk factors for *C. difficile* infection in children. (4)

d) Briefly describe the management of treating *C. difficile* infection in children (but do not describe the drug doses or describe the duration of therapy). Briefly describe the treatment options for
   i) The first episode of *C. difficile* infection.
   ii) Recurrent episodes of *C. difficile* infection.
   iii) Therapeutic options for a child with fulminant colitis. (5)

Subsequently, it becomes clear that the patient does not have post-infectious persistent diarrhoea and you consider other causes of chronic diarrhoea. The child does not suffer from any form of primary of acquired immunodeficiency.

e) List other non-infectious causes of diarrhoea that are mediated by an abnormal host immune response. (5)

f) List two dysmotility or bowel obstructive disorders that can present with diarrhoea. (1)

g) List any four causes of congenital secretory diarrhoea and/or causes of steatorrhoea in the neonatal period or early infancy. (4)

h) List two neuroendocrine tumours that can present with diarrhoea. (1)

PTO/Page 3 Question 2i)…

i) When investigating a child with chronic diarrhoea not due to an infectious cause, briefly discuss the tests that should be performed on stool specimens. The interpretation of these stool tests should be discussed in your answer. (10)

[40]

Question 3

An adolescent boy (15-years-old) with cystic fibrosis (CF) suffers from steatorrhoea and exocrine pancreatic insufficiency is suspected.

a) Briefly describe the indirect (non-stimulatory) and direct (stimulatory) tests that can be performed to assess pancreatic function in children. (10)

A month later, the child develops severe abdominal pain and pancreatitis is diagnosed.

b) How do you categorise pancreatitis in children? (3)

c) How would you investigate a child with suspected pancreatitis? (in this instance, assume that the child does not have cystic fibrosis). (10)

A liver function test performed after the resolution of the pancreatitis showed elevated levels of aspartate aminotransferase (AST), alanine aminotransferase (ALT), and gamma glutamyl transpeptidase (GGT). A liver biopsy was performed and histopathological analysis revealed proliferation of the bile ducts and portal fibrosis, with accumulation of amorphous pink periodic acid-Schiff (PAS) – positive material within the bile ducts.

d) Briefly describe the major clinical manifestations of progressive cystic fibrosis-related liver disease (CFLD). (5)

e) What types of gall bladder disease are seen in patients with cystic fibrosis? (2)

f) Briefly describe the principles of managing bleeding oesophageal varices given that nonselective beta-adrenergic blockers are generally avoided in patients with cystic fibrosis because of their potential to cause bronchoconstriction. (5)

This adolescent is receiving pancreatic enzyme replacement therapy and is assessed regularly by the dietician. In addition to calculating his caloric requirements (which have been adequately met in his diet), the adolescent also receives fat-soluble vitamin supplementation that the dietician has recommended Two-years later, the child develops severe liver disease.

g) Please describe the rationale for lowering the dose of Vitamin A in children with cystic fibrosis who develop severe liver disease. (2)

h) What form of vitamin A is safest to prescribe? (1)

i) What are the complications of chronic vitamin A toxicity? (2)

[40]

Question 4

A 5-week-old female infant presents with jaundice of three weeks duration. Her mother reports that her daughter's stools are now pale. On clinical examination, hepatomegaly is detected but the child is not dysmorphic. There is no cardiac, pulmonary, or neurologic disease. A liver function test (LFT) is performed.

a) What is conjugated hyperbilirubinaemia defined as? (Please provide a general definition i.e. not limited to the above scenario). (2)

b) What treatable disorders would you initially exclude in a young infant presenting with cholestatic jaundice? (Please provide a general approach i.e. not limited to the above scenario). (5)

The LFT results show a normal serum gamma glutamyltransferase (GGT) level.

c) What conditions would you now consider in your differential diagnosis? (3)

The infant is scheduled for a liver biopsy.

d) Please list the major and minor complications associated with the performance of liver biopsy in children. (4)

e) What risk factors predispose to the development of complications associated with liver biopsy in children? (Please list the risk factors for children in general and not just the risk factors pertaining to this clinical scenario). (4)

As part of the management of cholestatic jaundice, the infant is prescribed supplemental vitamins.

f) What forms of vitamin E preparations are suitable for the prevention and treatment of vitamin E deficiency in cholestatic jaundice? (2)

The infant's condition worsens progressively and the infants suffers from severe pruritus.

g) Briefly describe the therapeutic options and procedures that may be considered to improve the cholestasis in this patient. (5)

Despite the above management, the infant's condition worsens and liver transplantation (LT) is considered.

h) What are the most reliable anthropometric assessments that indicate the child's nutritional status pre-transplantation? (2)

i) Aggressive nutritional support prior to LT improves patient and graft survival as well as neurodevelopmental outcome. Briefly describe the recommended fat and protein requirements needed to optimise the child's nutritional status before transplantation. (3)

After appropriate counselling and work-up, liver transplantation is scheduled to proceed. Living-donor liver transplantation is considered.

PTO/Page 5 Question 4j)…

j) Briefly discuss the considerations that need to be met before living-donor liver transplantation can take place. (5)

k) In children undergoing liver transplantation, briefly comment on the neurocognitive and neurodevelopmental status following liver transplantation. Prior to transplantation, which factors contribute to poorer neurocognitive outcomes? (Please comment on these features in all children undergoing transplantation and not just limited to this clinical scenario). (5)

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3 March 2017

Paper 2 (3 hours)

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Please note the meaning of the following terms:

(i) 'List' means to enumerate i.e. specify a list or an outline form of reply in concise form
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Question 1

A 16-year-old-female adolescent with anorexia nervosa is severely malnourished. She is diagnosed with gastroparesis and has no evidence of organic disease.

a. Briefly describe the factors predisposing to acute gastric dilatation in patients with anorexia nervosa. (3)

The adolescent recently experienced acute weight loss, food intolerance, postprandial abdominal pain and distension, bilious vomiting, and weight loss. Her pain was relieved when she assumed the prone, knee-chest, or left lateral decubitus position.

b. What is the likely diagnosis? (1)
c. Discuss the pathophysiology of this condition. (2)
d. How is this condition usually diagnosed? (1)
e. How is this condition specifically managed? (1)

A while later, the adolescent is re-admitted with raised transaminases. Starvation-induced autophagy is suspected.

f. What is autophagy? (1)

g. In a severely malnourished child, briefly describe the pathophysiological mechanisms that lead to autophagy within hepatocytes. (1)
[10]

PTO/Page 2 Question 2…

Question 2

Regarding gastritis in children:

a) What are the typical endoscopic features of gastritis caused by *H.pylori?* (2)
b) Discuss the principles of the rapid urease test – performed on gastric biopsy samples – which is used to diagnose *H.pylori* infection in children. (3)
c) Write short notes on the Zollinger-Ellison syndrome (pathophysiology, clinical presentation, gastroscopy findings and treatment). (5)
[10]

Question 3

In neonates with gastrointestinal bleeding:

a) Apart from vitamin K deficiency bleeding (VKDB), list other common and/or life-threatening causes of upper and lower GI bleeding. (6)
b) How is VKDB diagnosed in neonates? (2)
c) Briefly describe the strategies used to prevent VKDB in neonates. (2)
[10]

Question 4

Regarding IBD in childhood:

a) List the cutaneous extra-intestinal (i.e. skin and mouth) manifestations of IBD. (3)
b) In a child with isolated colitis, what findings on clinical examination of the gastrointestinal system (not imaging, endoscopic or histopathological features) favour a diagnosis of CD over UC? (2)
c) Despite the limited evidence to guide the use of infliximab in children, briefly describe the indications when you would consider its use in a child with Crohn's disease. (2)
d) Briefly describe the serious adverse events associated with the use of infliximab in children. (3)
[10]

Question 5

The placement of percutaneous endoscopic gastrostomy (PEG) tubes in children and adolescents is to ensure safe feeding access and/or the prevention of aspiration.

a) List six other clinical indications for PEG tube placement in children and adolescents. (3)
b) Name two absolute contra-indications to PEG tube placement in children and adolescents. (2)
c) List six major complications and four minor complications of PEG tube placement in children and adolescents. (5)
[10]

PTO/Page 3 Question 6…

Question 6

Regarding coeliac disease in children:

a) Briefly define the following terms
   i) Classic coeliac disease.
   ii) 'Atypical' or non-gastrointestinal coeliac disease.
   iii) 'Silent' or sub-clinical coeliac disease.
   iv) Latent coeliac disease. (4)

b) List four non-gastrointestinal complications of coeliac disease. (2)

c) Briefly describe the measures needed to monitor the response to a gluten-free diet. (2)

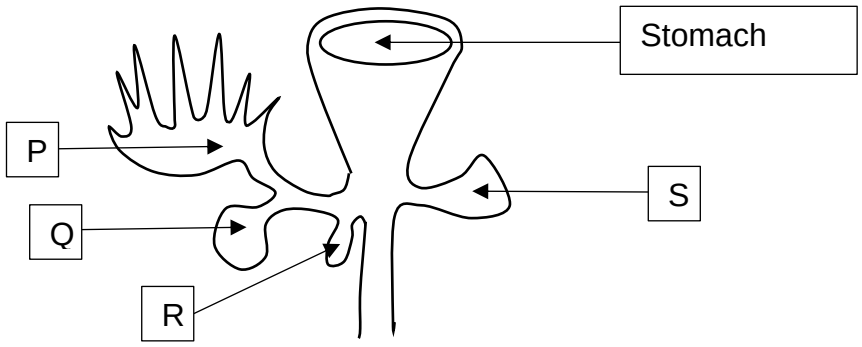
d) Except for first- and second-degree relatives of patients with celiac disease, list at least four other groups of children that should be routinely screened for coeliac disease even if they display no symptoms or signs of coeliac disease. (2)

[10]

Question 7

Regarding the embryological development (at day 35) of the biliary tree and pancreas:

a) Label the following structures P, Q, R, S shown in the diagram below. (4)



b) Briefly describe three postulated mechanisms that may lead to the development of congenital biliary cysts (previously known as choledochal cysts). (2)

c) Except for stone and sludge formation in the cyst and/or gallbladder and/or the liver, what other complications can arise from biliary cysts? (4)

[10]

Question 8

In chronic liver disease:

a) Briefly describe the pathogenesis of portal hypertension. (4)

b) Define the term "hepatopulmonary syndrome (HPS)". (1)

c) What screening tests are used to identify HPS? Indicate which screening test is the most sensitive at identifying HPS. (3)

d) Briefly describe the role of liver transplantation in children with HPS. (2)

[10]

PTO/Page 4 Question 9…

Question 9

A child with short bowel syndrome (but with an intact colon) is been transitioned from enteral nutrition to oral feeds. During this transition to oral feeds, the infant experiences watery diarrhoea. Nonetheless, a decision is made to continue with oral feeds. In this case, briefly describe the recommended dietary composition of the oral feeds with regard to

a) Fats. (2)
b) Carbohydrates. (2)
c) Soluble fibre. (1)

This child was inadvertently given a high carbohydrate meal and became confused, and developed cerebellar ataxia and slurred speech.

d) What complication is this child suffering from? (1)
e) Briefly describe the pathogenesis of this complication. (2)
f) List the pharmacologic agents that may be used to treat a child with this complication. (2)
[10]

Question 10

Regarding digestion in children:

a) List the principle enzyme that is responsible for the luminal digestion of carbohydrates in the oral cavity and duodenum. (1)
b) Briefly describe the action of the brush border membrane hydrolases that are used to digest disaccharides to monosaccharides. (3)
c) Briefly describe the clinical presentation, pathogenesis, and diagnosis of glucose-galactose malabsorption. (3)
d) How is glucose-galactose malabsorption treated? (1)
e) Briefly describe the clinical presentation and pathogenesis of congenital sucrase-isomaltase deficiency. (2)
[10]