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

1 March 2018

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)

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1 A 10-month-old boy, with confirmed cystic fibrosis, is seen in your practice

a) The dietitian in the hospital has not treated children with cystic fibrosis before. What guidelines for *macronutrient* support of this child would you give her and how would you monitor his nutritional status? (5)

b) What routine *micronutrient* supplements should this child receive? (5)

c) After 6 months, you notice that he is not growing satisfactorily. How would you manage him? (5)

d) When he is 6-years-old, the results of routine liver biochemistry are

- ALT 145u/l.
- ALP 360u/l.
- GGT 98u/l.
- Total bilirubin 15umol/l.

Discuss

i) The differential diagnosis for these findings. (5)

ii) The life-time risk of developing chronic liver disease. (2)

iii) How you would monitor him for the development of liver disease? (3)

[25]

2 A 6-year-old girl presents to you with the following complaints and special investigation findings: tiredness for 3-months and mild jaundice for 2-weeks; total bilirubin 96umol/l; conjugated bilirubin 41 umol/l; total protein 80g/l; albumin 41g/l; ALT 540u/l; AST 415u/l; GGT 91u/l; ALP 340u/l

a) Discuss the differential diagnosis. (5)

b) If this patient developed dystonia and dysarthria over the next few months, what would the most likely genetic disorder be? (1)

c) Discuss what laboratory and genetic tests you would perform. (5)

d) Discuss the treatment goals and options you would consider for this patient. (5)

e) Discuss the method of action and side effects of each treatment option. (5)

f) Broadly discuss the clinical manifestations of this genetic disorder. (4)

[25]

3 A 3-year-old boy presents with a 2-week history of painless rectal bleeding

a) What conditions would you consider in the differential diagnosis? (5)

b) You decide to perform a colonoscopy.

i) Discuss the preparation of the patient and

ii) The complications of colonoscopy in this child. (10)

c) You find 8 polyps in the colon.

i) Discuss the technical considerations for the removal of the polyps. (3)

ii) What are the most likely causes for this endoscopic finding? (2)

d) The histology of the colonic polyps is reported as: hamartomatous mucosal polyps with a central core of branching smooth muscle. You also notice that the boy has pigmented spots on his lips.

i) What is the inheritance pattern of this polyp (include the genetic defect)? (2)

ii) What intestinal complications can this patient develop? (3)

[25]

4 A 12-year-old child with known Crohn's disease developed an ileal stricture that was removed surgically and closed with an end on end anastomosis. She is stable at present and is referred to you for ongoing management

a) Briefly discuss the risk of recurrence of stricture formation and strategies to reduce the risk of recurrence. (5)

b) Discuss the monitoring of disease activity post-surgery. (5)

c) Discuss the tools available to score disease activity *clinically*. (5)

d) Discuss the extra-intestinal disease manifestations of inflammatory bowel disease and management. (5)

e) The parents have heard of dietary management of Crohn's disease. Discuss dietary treatment, excluding exclusive enteral nutrition, of Crohn's disease. (5)

[25]

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2 March 2018

Paper 2 (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)

1 Write short notes on the following
   a) Human milk oligosaccharides. (5)
   b) Vitamin E deficiency: discuss the causes of vitamin E deficiency in children and the clinical manifestations. (5)
   [10]

2 A pre-primary school located in an affluent area of Johannesburg approached you as paediatric gastroenterologist to address the parents on the use of commercially available formula for children in the age group of 1-3 years
   a) Discuss current guidelines regarding the use of formula in this age group. (5)
   b) One of the parent's requests advice on the intake of sugar in young children. What recommendations would you make based on the most recent recommendations. (5)
   [10]

3 A 20-month-old girl with biliary atresia is referred to your practice. On assessment you notice that she has severe deformity of her legs. Briefly discuss the manifestations, diagnosis and management of bone disease in this patient with chronic liver disease. [10]

4 The 20-month-old girl with biliary atresia (discussed in question 3) is considered for liver transplant
   a) How would you evaluate the nutritional status of this child? (5)
   b) After successful liver transplantation, she develops features suggestive of a lymphoproliferative disorder. How would you confirm the diagnosis and briefly describe the principles of management? (5)
   [10]

5 A 5-year-old child presents to you after falling over the handle of his bicycle the previous day. He complains of severe abdominal pain and occasional vomiting. There are no clinical or radiological signs to suggest intestinal obstruction and there is no free air in the peritoneal cavity. His haemoglobin is 12,3g/dl
   a) What is the single most important cause for his abdominal pain?
   b) What biomarkers would you use to confirm the diagnosis?
   c) What imaging modalities would you use (discuss the most appropriate imaging modalities)?
   d) What analgesia would you give the boy?
   e) How would you support him nutritionally? [10]

PTO/Page 2 Question 6)…

6 A 3-year-old boy presents with the problem of poor growth. His parents give the history that he passes large foul smelling stools. Special investigations reveal a faecal elastase of <50ug/g of stool. Discuss the differential diagnosis of these findings and how you would further investigate this child. [10]

7 You are called to give an opinion on a 10-day-old infant. The infant is jaundiced, has prominent ascites and the following special investigation results: total bilirubin 130umol/l; conjugated bilirubin 84umol/l; INR 8.5. Discuss your differential diagnosis and what relevant special investigations you would perform. [10]

8 You are asked to see a term 8-day-old neonate with the finding of a very prominent liver. On examination the infant is not jaundiced, her pulse rate is 180/min and her respiratory rate is 64/min. According to her mother she tires quickly when feeding and she notices sweat on her forehead. Her liver is palpable 6cm below the costal margin with a normally placed upper border. The spleen is not palpable and there is no ascites. You notice a small haemangioma on her left leg. A full blood count reveals the following: Haemoglobin 13,2g/dl, white cell count 12,5x10^9/l, platelet count 72 x10^9/l

a) What is the single most likely diagnosis and what other causes of severe hepatomegaly would you consider? (5)
b) How would you manage this infant for the most likely cause of the hepatomegaly? (4)
c) For which endocrine abnormality should this infant be assessed? (1)

[10]

9 A 4-year-old boy presents with the complaint of recurrent symmetrical oedema of the lower legs. His parents give the history that he has had this complaint for the past 2-years. In addition, he has frequent episodes of diarrhoea. When he was 1-year-old he was hospitalised for abdominal tuberculosis. Important findings of special investigations are: Total protein 32g/l; albumin 18g/l; cholesterol 2,1mmol/l; white cell count 7,2 X 10^9/l; lymphocyte count 0,6 X 10^9/l; neutrophil count 5,4X10^9/l. His HIV ELISA is negative and he has no history of opportunistic infections or severe infections other than tuberculosis

a) What is the most likely diagnosis? (2)
b) How would you confirm the diagnosis? (4)
c) How would you manage this boy? (4)

[10]

10 A 9-year-old girl presents with a 3-year history of recurrent episodes of vomiting. The episodes all follow the same course. She feels unwell for a few hours before onset of the vomiting. The vomiting is severe and has led to dehydration and hospitalisation on a number of occasions. Extensive special investigations by general paediatricians have been normal. Between the episodes she has no complaints and is thriving

a) What is the most likely diagnosis and what criteria do you use to make the diagnosis (specify the criteria)? (4)
b) How would you manage an episode of vomiting? (3)
c) How would you try to prevent episodes of vomiting from occurring? (3)

[10]