THE COLLEGES OF MEDICINE OF SOUTH AFRICA

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Reg No 1955/000003/08

Examination for the Subspeciality Certificate in Paediatric Neurology of the College of Paediatricians of South Africa

25 July 2019

Paper 1 (3 hours)

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

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1 Discuss the current approach in the diagnosis and management of the Acquired Demyelinating Disorders of Childhood, using the following conditions as examples
   a) Acute Disseminated Encephalomyelitis (ADEM). (10)
   b) Multiple Sclerosis. (20)
   c) NMOSD (Neuromyelitis Optica Spectrum Disorder). (20)
   [50]

2 a) You are referred a 6-year-old boy who, over the past 6-months, has been having increasing difficulty keeping up with his peers at play. His parents noticed a deterioration in his behaviour with increasing hyperactivity and report that he is now struggling to keep up at preschool whereas previously he was thought to be very "clever". Clinical examination reveals moderate spasticity in both lower limbs with pathologically increased deep tendon reflexes in all 4 limbs. You order a MRI and the radiologist reports that there is a "leucodystrophy with frontal predominance". Describe your approach to a differential diagnosis based on MRI appearance and the investigations that you would order in this case. (20)
   b) In any Cerebral palsy (CP) clinic there are so-called cerebral palsy mimics. Name these mimics and discuss which investigations are indicated to identify these conditions. (20)
   c) What is your understanding of the term "pharmaco-resistant Epilepsy" and what are the options for managing such a patient? (10)
   [50]

3 A 7-year-old child presents with a three-week history of a droopy left eye lid and unequal pupils.
   a) Discuss possible clinical diagnoses. (15)
   b) What other neurological clinical signs would assist you in making a diagnosis? (15)
   c) Discuss the relevant neuro-anatomical pathways that are necessary to make a clinical diagnosis. (20)
   [50]

4 a) Discuss Pompe's disease in childhood with respect to the following
      i) Clinical presentation. (10)
      ii) Genetics and Diagnostic confirmation. (5)
      iii) Management and complications. (10)

PTO/Page 2 Question 4b)…

b) SMA (Spinal muscular atrophy) is a poor prognostic condition, especially for type 1 and type 2. Discuss recent advances in therapeutic intervention in the following context:

i) Genetic abnormality in SMA and mechanism of action of therapy. (10)

ii) Therapeutic intervention, administration and complication. (10)

iii) Practical considerations in the South African setting. (5)

[50]