

MCQs

Instruction:

1. Please use pencil to shade the box for the correct answer (see loose leaf page).
2. Send back the answer sheet to the Hong Kong College of Paediatricians for the award of 4 CME points for those with >50% correct answers.

The following statements are true / false

(A) Current Status of Acute Lymphoblastic Leukaemia in Children

1. Concerning the hypothesis related to the pathogenesis of childhood ALL, the following statement is/are correct:
 - a. Delayed exposure to common infection such as chickenpox may predispose to leukaemogenesis.
 - b. Dietary and environmental exposures of pregnant women or infant to high doses of chemical such as flavonoids may predispose to leukaemogenesis.
 - c. Deficiency of glucose 6-phosphate dehydrogenase, which detoxifies electrophilic metabolites, has been associated with infant leukaemias.
 - d. Many genetic aberrations associated with childhood leukaemia were found to be fetal in origin.
 - e. Frequent exposure to electromagnetic field such as watching TV may predispose to leukaemogenesis.
2. The following is the most important prognostic indicator for treatment outcome in childhood ALL:
 - a. Initial leukaemic blast count.
 - b. Patient's age at diagnosis.
 - c. Patient's sex.
 - d. Presence of specific cytogenetic abnormalities.
 - e. Persistent morphological disease after 2 weeks of treatment.
3. In order to adequately assess the treatment response, currently the most sensitive way in monitoring the minimal residual disease during and after treatment is:
 - a. Determining aberrant surface antigens expression (immunophenotypes) by flow cytometry.
 - b. Semi-quantitative polymerase chain reaction (PCR) analysis of leukaemic specific clonal antigen-receptor gene rearrangements.
 - c. Reverse transcriptase polymerase chain reaction (RT-PCR) analysis of specific genetic chimeric transcripts induced by translocations.

- d. Morphological screening of residual leukaemic blast cells by high power microscopy.
 - e. Serum level of lactate dehydrogenase (LDH) by high resolution chromatography.
4. Currently, relatively good outcome of 70-80% long-term event free survival can be achieved by different chemotherapy protocols. The total duration of treatment will last for:
 - a. 1 to 2 years.
 - b. 2 to 3 years.
 - c. 3 to 4 years.
 - d. 4 to 5 years.
 - e. 5 to 6 years.
 5. With more and more children survived from their leukaemia, rare but significant long term therapy-related complications started to appear and the following are known complications except for one:
 - a. Avascular necrosis of the femoral heads.
 - b. Bone tumour.
 - c. Brain tumour.
 - d. Acute myeloid leukaemia.
 - e. Cognitive function deficit.

(B) Experience of ¹³C-Urea Breath Test in Eight Children

1. The following statement about *H. pylori* infection is/are correct:
 - a. It is an uncommon infection.
 - b. It was estimated that half of the world population is infected.
 - c. Infection in children is more common in developing countries.
 - d. It was significantly associated with recurrent abdominal pain.

2. *H. pylori* infection could be diagnosed by the following method:
 - a. Rapid urease test.
 - b. Histology.
 - c. ¹³C-urea breath test.
 - d. Skin prick test.
3. Urea breath test requires the following preparations:
 - a. Nil by mouth for 4 hours.
 - b. Citric acid test meal.
 - c. Ingestion of radioisotope labelled urine.
 - d. Collection of exhaled gas for analysis.
4. The main problem(s) of urea breath test include the following(s):
 - a. Cost.
 - b. Uncertainty about universal screening for *H. pylori* infection.
 - c. Absence of normal values.
 - d. Age limitation.
5. False negative result occurs in urea breath test under the following circumstances:
 - a. Younger than 2-year.
 - b. Prior antibiotic treatment.
 - c. Diarrhoea.
 - d. Inhaled corticosteroid treatment.
3. The following investigation result(s) is/are NOT typically found in patients with Ataxia-Telangiectasia:
 - a. Thrombocytopenia.
 - b. Deficient levels of IgA.
 - c. Deficient levels of IgE.
 - d. Raised alpha fetoprotein (AFP).
 - e. Raised carcinoembryonic antigen (CEA).
4. The following is/are TRUE about the AT Mutant (ATM) gene:
 - a. Discovered in 1985.
 - b. Identified on chromosome 11q23-q24.
 - c. Produced by ATM protein.
 - d. One unique mutation has been described.
 - e. Defect in only one copy of the gene may also result in hypersensitivity to ionizing radiation.
5. The following is/are NOT a feature of the ATM protein:
 - a. Has multifunctional.
 - b. Possesses a domain which bears strong similarities to phosphoinositol-3 kinases.
 - c. The protein is also involved in DNA repair.
 - d. Affect the rate of apoptotic cell death.
 - e. Participates in insulin-dependent glucose transport.

(C) Ataxia Telangiectasia: Case Report of Three Affected Brothers and Review of the Literature

1. The following is/are TRUE about Ataxia-Telangiectasia:
 - a. The incidence is ~1 in 40,000-300,000 live births.
 - b. Males are affected more than females.
 - c. The disease is more prevalent in Asians.
 - d. It is an autosomal dominant disease.
 - e. Female carriers of the defective gene are prone to breast cancer.
2. The following is/are NOT a clinical feature of Ataxia-Telangiectasia:
 - a. Choreoathetosis.
 - b. Progeria.
 - c. Dilated vessels on cheeks.
 - d. Epileptic tendency.
 - e. Proneness to insulin-resistant diabetes.
1. The following is/are not feature(s) of chronic malabsorption:
 - a. Anaemia.
 - b. Muscle wasting.
 - c. Steatorrhoea.
 - d. Satisfactory weight gain.
 - e. Delayed bone age.
2. The following helps to diagnose allergic gastro-enteropathy:
 - a. Presence of antigen-specific IgE.
 - b. Resolution of symptom with food avoidance.
 - c. Presence of autoimmune markers.
 - d. Positive response to specific food challenge.
 - e. Responsiveness to steroid treatment.
3. The following is/are clinical features of food allergy:
 - a. Skin eczema.
 - b. Diarrhoea.
 - c. Protein-losing enteropathy.
 - d. Rhinitis.
 - e. Malabsorption.

(D) Treatment of Allergic Enteropathy in a 7-year-old Girl with Controlled-release Budesonide

4. The following is/are useful in monitoring the response of allergy enteropathy:
 - a. Gastrointestinal symptoms.
 - b. Blood α -1 antitrypsin level.
 - c. Xylose absorption test.
 - d. Small bowel biopsy.
 - e. Renal function test.
5. The following statement(s) concerning treatment of allergic gastroenteropathy is/are true:
 - a. Elimination of offending allergies is the mainstay of treatment.
 - b. Total parenteral nutrition or elemental diet following by careful food reintroduction is a treatment of choice.
 - c. Systemic steroid is usually not helpful.
 - d. Ketotifen is an effective treatment.
 - e. Oral sodium cromoglycate is an effective treatment.

(E) Acute Rheumatic Fever Presenting with Sydenham's Chorea

1. The following is/are major criteria in diagnosing acute rheumatic fever according to Jones Criteria:
 - a. Carditis.
 - b. Migratory polyarthritis.
 - c. Arthralgia.
 - d. Sydenham's chorea.
 - e. Prolonged PR interval on ECG.
2. Sydenham's chorea is associated with the following neurological impairment:
 - a. Motor restlessness.
 - b. Gait disturbance.
 - c. Visual failure.
 - d. Dysarthria.
 - e. Hearing impairment.
3. The following psychiatric manifestation is/are associated with Sydenham's chorea:
 - a. Obsessive-compulsive disorders.
 - b. Childhood schizophrenia.
 - c. Tics.
 - d. Attention deficit disorder.
 - e. Anorexia nervosa.

4. The following medications have been shown to be effective in controlling Sydenham's chorea:
 - a. Botulinium toxin.
 - b. Haloperidol.
 - c. Sodium Valproate.
 - d. Carbamazepine.
 - e. Phenobarbital.
5. The following treatment is/are appropriate in the management of acute rheumatic fever:
 - a. Bed rest in case of acute carditis.
 - b. 3 days course of penicillin to eradicate group A streptococcus.
 - c. Anti-inflammatory dose of salicylates in case of carditis.
 - d. Once weekly benzathine benzylpenicillin injection for secondary prophylaxis.
 - e. Twice-daily oral penicillin V for secondary prophylaxis.

(F) Immunisation: Opportunities and Challenges

The following is/are correct:

1. Of the vaccine preventable diseases, pertussis (whooping cough) causes the most deaths annually.
2. The immunisation schedule for premature infants should commence according to birth age, not gestational age.
3. Acellular pertussis vaccines have a decreased rate of local and systemic reactions in comparison to whole cell pertussis vaccines.
4. Parental attitudes to immunisation are the most significant cause of poor immunisation uptake in developed countries.
5. The efficacy of the 7-valent conjugate pneumococcal vaccine against vaccine serotype invasive disease is greater than 95%.