RACP Written Examination February 2018

Paediatrics & Child Health

Medical Sciences questions 101–170
101. Which of the following is the most common anatomical abnormality seen in tracheo-oesophageal fistula?

A. Image A.
B. Image B.
C. Image C.
D. Image D.
E. Image E.
102. What is the most appropriate statistical test to compare the proportion of sporting injuries in a group of children who play netball to a group of children who play basketball?

A. Chi-square test.
B. Kappa coefficient.
C. Linear regression.
D. Paired t-test.
E. Pearson correlation.

103. An outbreak of hospital-acquired infection occurs in three wards of a paediatric tertiary hospital. All staff are swabbed to identify potential carriers of the infective agent. The proportion of carriers in each ward is determined and compared.

What type of study is this?

A. Case series.
B. Case-control.
C. Cohort.
D. Cross-sectional.
E. Ecological study.
A term infant presents with respiratory distress soon after birth. A chest x-ray is performed and is shown below.

What is the diagnosis?

A. Congenital lobar emphysema.
B. Congenital pneumonia.
C. Cystic adenomatoid malformation.
D. Hyaline membrane disease.
E. Pneumothorax.
105. Jaw pain is a common side effect of which chemotherapeutic agent?

A. Bleomycin.
B. Cisplatin.
C. Clofarabine.
D. Doxorubicin.
E. Vincristine.

106. Which of the following medicines is a strong inhibitor of cytochrome P450 3A enzymes?

A. Carbamazepine.
B. Clarithromycin.
C. Ethinyloestradiol.
D. Fluoxetine.
E. Rifampicin.

107. Other than molecular size, which property affects filtration of substances by the glomerular basement membrane?

A. Antigenicity.
B. Concentration.
C. Electrical charge.
D. Lipophilicity.
E. Solubility.
108. Which infectious disease gives rise to the highest number of secondary cases in an unvaccinated, non-immune population?

A. Influenza.
B. Measles.
C. Mumps.
D. Polio.
E. Tuberculosis.

109. Mutation in which gene increases the likelihood of developing Crohn disease?

A. ABCB4.
B. EPCAM.
C. JAG1.
D. MYO5B.
E. NOD2.

110. A mitral opening snap occurs in what phase of the cardiac cycle?

A. Atrial systole.
B. Early diastolic ventricular filling.
C. Isovolumic ventricular relaxation.
D. Late diastolic ventricular filling.
E. Late ventricular ejection.
111. In a randomised controlled trial of a novel antidepressant in children and adolescents with obsessive-compulsive disorder, a number of participants are noted to develop suicidal ideation. Results from this trial are shown below.

<table>
<thead>
<tr>
<th></th>
<th>Novel antidepressant</th>
<th>Placebo</th>
</tr>
</thead>
<tbody>
<tr>
<td>Suicidal ideation</td>
<td>20</td>
<td>3</td>
</tr>
<tr>
<td>No suicidal ideation</td>
<td>980</td>
<td>997</td>
</tr>
<tr>
<td></td>
<td>1000</td>
<td>1000</td>
</tr>
</tbody>
</table>

What is the number needed to harm (NNH) for the novel antidepressant compared with placebo?

A. \( \left( 1 - \frac{20}{3} \right) \times 100 \)
B. \( \frac{17}{1000} \)
C. \( \frac{20}{3} \)
D. \( \frac{20}{980} \div \frac{3}{997} \)
E. \( 1 \div \frac{17}{1000} \)

112. An increase in which of the following will cause a shift to the left of the oxygen dissociation curve?

A. Carboxyhaemoglobin.
B. Hydrogen concentration.
C. PCO₂.
D. Red cell 2,3-diphosphoglycerate level.
E. Temperature.
113. What condition leads to an unreliably low glycosylated haemoglobin (HbA$_{1C}$) immunoassay despite sustained hyperglycaemia?

A. Hereditary spherocytosis.
B. Iron deficient anaemia.
C. Sickle cell trait.
D. Thalassemia trait.
E. Vitamin B$_{12}$ deficiency.

114. What cell in the pancreatic islets of Langerhans is responsible for the secretion of glucagon?

A. Alpha.
B. Beta.
C. Delta.
D. Epsilon.
E. Gamma.

115. A patient with diabetes is confirmed to have maturity-onset diabetes of the young (MODY) due to an HNF4α mutation.

Which class of non-insulin medication is indicated?

A. Biguanide.
B. Glitazone.
C. GLP-1 agonist.
D. SGLT2 inhibitor.
E. Sulfonlurea.
116. Which feature of the immunoglobulin molecule is determined by the Fab region?

A. Antigen binding.
B. Circulating half-life.
C. Isotype specificity.
D. Placental transfer.
E. Proteolysis susceptibility.

117. Which cytokine plays a role in promoting the Th2 immune response?

A. Interferon gamma.
B. Interleukin 2.
C. Interleukin 4.
D. Interleukin 7.
E. Tumour necrosis factor alpha.

118. Dysregulation of which pathway is responsible for the clinical manifestation of tuberous sclerosis?

A. Activation of insulin-like growth factor 1 (IGF-1).
B. Activation of mTOR.
C. Activation of neuregulin 1 (NGR1).
D. Inhibition of interleukin 6 (IL-6).
E. Inhibition of oncogene Ras.
A 2-year-old girl presents with her second episode of meningococcaemia.

What is the most appropriate initial test to screen for possible underlying complement deficiency?

A. Complement activity, alternate pathway (AH50).
B. Complement C2.
C. Complement C3.
D. Factor D.
E. Total haemolytic complement (CH50).
On the cardiac performance curves below, an increase in which physiological parameter alters the ventricular performance from point A to point B?

A. Heart rate.
B. Myocardial contractility.
C. Oxygen consumption.
D. Systemic resistance.
E. Venous filling.
121. The HEADSS adolescent psychosocial risk assessment is used as a framework for conversation to develop rapport with a young person.

What does the "A" stand for?

A. Academic achievement.
B. Accommodation.
C. Activities.
D. Affect.
E. Alcohol use.

122. Which histological feature found on biopsy during colonoscopy would favour a diagnosis of Crohn disease over ulcerative colitis?

A. Crypt abscesses.
B. Granulomata.
C. Lymphocytic infiltrate.
D. Paneth cells.
E. Superficial inflammation.

123. What is the main purpose of a phase IV clinical drug study?

A. Determine the dose range for children from the adult data.
B. Determine the final formulation.
C. Examine the use post-marketing in real world situations.
D. Investigate off-label use of a registered medication.
E. Investigate the effectiveness in children compared to adults.
124. What is the most likely cause of oxygen desaturation in a sleeping infant recovering from bronchiolitis?

A. Hypoventilation.
B. Reduced tidal volume.
C. Secondary bacterial pneumonia.
D. Upper airway secretion.
E. Ventilation/perfusion (V/Q) ratio mismatch.

125. What is the most appropriate assessment tool to diagnose developmental delay in a 4-year-old?

A. Ages and Stages Questionnaires (ASQ-3).
B. Griffiths Mental Development Scales, 3rd edition.
C. Parents' Evaluation of Developmental Status (PEDS).
D. Wechsler Intelligence Scales for Children (WISC-V).
E. Wechsler Preschool and Primary Scale of Intelligence, 4th edition (WPPSI-IV).

126. In which organ is 25-hydroxycholecalciferol converted to the active hormone 1,25 dihydroxycholecalciferol?

A. Kidney.
B. Liver.
C. Lung.
D. Parathyroid gland.
E. Skin.
127. Which pharmacokinetic parameter is the most important in determining the steady-state dose?

A. Absorption rate constant ($K_a$).
B. Bioavailability ($F$).
C. Clearance (Cl).
D. Half-life ($t\frac{1}{2}$).
E. Volume of distribution ($V_D$).

128. Omalizumab is a biological product (monoclonal antibody) used in the treatment of severe asthma.

What is its specific molecular target?

A. Circulating IgE.
B. Eosinophilic basic protein.
C. Eotaxin 1.
D. Interleukin 5.
E. Leukocyte immunoglobulin-like receptor 7.

129. What is the mechanism of action of methylphenidate?

A. Blockade of dopamine 2 (D2) receptors.
B. Direct agonism of alpha-1 ($\alpha_1$) and alpha-2 ($\alpha_2$) receptors.
C. Inhibition of GABA.
D. Inhibition of noradrenaline and dopamine reuptake.
E. Inhibition of serotonin reuptake.
**130.** A 4-year-old boy ingests a large dose of aspirin (acetylsalicylic acid). What is the most appropriate therapy to maximise his urinary excretion of aspirin?

A. Allopurinol.
B. Forced diuresis with frusemide.
C. Hyperhydration.
D. Probenecid.
E. Urinary alkalinisation.

**131.** In a child with toxic shock syndrome due to group A streptococcal infection, in addition to penicillin which antibiotic is recommended to decrease toxin production?

A. Cefotaxime.
B. Ciprofloxacin.
C. Clindamycin.
D. Meropenem.
E. Vancomycin.
132. You are asked to see a 2-year-old boy (IV.3 below) who presents with significant motor and cognitive delay. The boy's grandmother (II.2) is present at the consultation and tells you that her 20-year-old son (III.1) presented in a similar fashion and now has a severe intellectual disability.

What is the most likely inheritance pattern in this family?

A. Autosomal dominant with reduced penetrance.
B. Autosomal recessive.
C. Mitochondrial inheritance.
D. X-linked dominant.
E. X-linked recessive.
133. A term neonate is resuscitated using bag and mask ventilation with air, and preductal oxygen saturation monitoring.

At 3 minutes of age what is the lowest oxygen saturation acceptable to continue ventilation in air?

A. 50
B. 60
C. 70
D. 80
E. 90

134. Lissencephaly is a disorder of what stage of neural development?

A. Myelination.
B. Neuronal and glial proliferation.
C. Neuronal migration.
D. Neuronal organisation.
E. Ventral induction.

135. During routine follow-up of a boy with spinal muscular atrophy (SMA) type 2, his mother informs you she is in a new relationship and 20 weeks pregnant with a male fetus.

What is the risk the fetus will be affected?

A. 1 in 2.
B. 1 in 4.
C. 1 in 50.
D. 1 in 200.
E. 1 in 10,000.
136. What is the most important indicator of effective positive pressure ventilation in neonatal resuscitation?

A. Audible bilateral breath sounds.
B. Improving colour.
C. Increasing heart rate.
D. Increasing peripheral oxygen saturation.
E. Visible chest rise.

137. Defects in the FLG gene (coding the Filaggrin protein) are associated with which skin condition?

A. Acne vulgaris.
B. Atopic dermatitis.
C. Epidermolysis bullosa.
D. Infantile haemangiomas.
E. Psoriasis.

138. Hereditary methaemoglobinaemia is most commonly due to deficiency of which red cell enzyme?

A. Cytochrome b5 reductase.
B. Glutathione peroxidase.
C. Heme oxygenase.
D. Methylenetetrahydrofolate reductase.
E. Phosphoglycerate kinase.
139. The thymic and parathyroid abnormalities seen in 22q11 microdeletion syndrome are due to defective development of which embryological structure(s)?

   A. Neural crest.
   B. Pharyngeal arches.
   C. Pharyngeal groove.
   D. Pharyngeal pouches.
   E. Rathke pouch.

140. Which chemotherapy drug inhibits microtubule formation?

   A. Cyclophosphamide.
   B. Dactinomycin.
   C. Doxorubicin.
   D. Etoposide.
   E. Vincristine.

141. Which lipid can be absorbed directly into the portal circulation?

   A. Cholesterol.
   B. Lysolecithin.
   C. Medium chain fatty acid.
   D. Monoglyceride.
   E. Very long chain fatty acid.
142. What is the mechanism of weakness in infantile botulism?

A. Axonal degeneration.
B. Competitive blockade of motor endplate.
C. Inflammatory myopathy.
D. Inhibition of neurotransmitter release.
E. Peripheral nerve demyelination.

143. A 1-year-old child (weight 10 kg) presents with urticaria and wheeze following the ingestion of peanut butter for the first time.

What is the correct management?

A. 0.1 mL of 1:1000 IM adrenaline.
B. 0.1 mL of 1:10,000 IM adrenaline.
C. 1 mL of 1:10,000 IM adrenaline.
D. 0.1 mL of 1:10,000 SC adrenaline.
E. 1 mL of 1:1000 SC adrenaline.

144. Which term describes the situation in which maintenance of a normal phenotype requires the proteins produced by both copies of the gene, and a 50% decrease in gene function results in an abnormal phenotype?

A. Dominant-negative effect.
B. Haploinsufficiency.
C. Heterozygosity.
D. Hypomorphic allele.
E. Uniparental disomy.
145. What is the average life span of normal human platelets in circulation?

A. 24 hours.
B. 72 hours.
C. 10 days.
D. 30 days.
E. 120 days.

146. Neonatal surfactant replacement therapy increases the risk of which serious complication?

A. Intraventricular haemorrhage.
B. Necrotising enterocolitis.
C. Pneumothorax.
D. Pulmonary haemorrhage.
E. Pulmonary interstitial emphysema.

147. Predisposition to bone marrow failure associated with dyskeratosis congenita results primarily from a defect in what critical cellular process?

A. DNA methylation.
B. Histone acetylation.
C. Interstrand crosslink DNA repair.
D. Ribosomal trafficking.
E. Telomere maintenance.
148. Which antibiotic is most likely to cause oesophageal ulceration if not taken with fluids or food and at least 1 hour prior to going to sleep?

A. Amoxicillin.  
B. Azithromycin.  
C. Cephalexin.  
D. Doxycycline.  
E. Rifampicin.

149. Which one of the following contributes most to serum osmolality?

A. Albumin.  
B. Creatinine.  
C. Glucose.  
D. Sodium.  
E. Urea.

150. Which of the following physical processes is primarily responsible for fluid removal during peritoneal dialysis?

A. Active transport.  
B. Facilitated diffusion.  
C. Hydrostatic pressure.  
D. Osmotic gradient.  
E. Solute drag.
151. What formula can be used to estimate the correct size of an uncuffed endotracheal tube for use in children?

A. \((\text{Age} \div 2) + 4\)
B. \((\text{Age} \div 4) + 4\)
C. \((2 \times \text{Age}) - 4\)
D. \(\text{Age} + 4\)
E. \((\text{Age} \div 4) + 2\)

152. A 2-year-old boy is referred with elevated creatine kinase (CK). Calf hypertrophy is noted. There is subtle proximal weakness.

Which feature best differentiates between Duchenne and Becker muscular dystrophy?

A. Creatine kinase level.
B. Exon site of the deletion.
C. Out of frame deletion.
D. Presence of an intragenic deletion.
E. Presence of cardiomyopathy.

153. A 12-year-old boy is undergoing epilepsy surgery for hippocampal sclerosis.

Which post-operative visual field deficit is most likely to occur as a complication of this surgery?

A. Bitemporal hemianopia.
B. Contralateral homonymous hemianopia with macular sparing.
C. Contralateral homonymous hemianopia.
D. Contralateral homonymous inferior quadrantanopia.
E. Contralateral homonymous superior quadrantanopia.
154. What is the earliest gestational age at which pupils first start responding to light?

A. 24 weeks.
B. 28 weeks.
C. 32 weeks.
D. 36 weeks.
E. Term.

155. Which condition is treatable by enzyme replacement therapy with recombinant human acid d-glucosidase?

A. Galactosaemia.
B. Glutaric aciduria.
C. Glycerokinase deficiency.
D. Methylmalonic aciduria.
E. Pompe disease.

156. To which category of base do the DNA bases cytosine and thymine belong?

A. Amine.
B. Lysine.
C. Protamine.
D. Purine.
E. Pyrimidine.
157. In early infancy, what event precipitates congestive heart failure due to large ventricular septal defects?

A. Arterial duct closure.
B. Increase in size of ventricular septal defect.
C. Reduced pulmonary vascular resistance.
D. Reduced systemic pressure.
E. Reversal of the shunt.

158. In addition to pertussis vaccination, which vaccine when given to pregnant women has been shown to reduce the likelihood of disease in their infants up to 3 months of age?

A. Hepatitis B.
B. Influenza.
C. Measles.
D. Meningococcal B.
E. Varicella.

159. What is the mechanism of the prokinetic effect of erythromycin?

A. Cholecystokinin antagonist.
B. Dopamine agonist.
C. Guanylate cyclase agonist.
D. Motilin receptor agonist.
E. Serotonin agonist.
160. Which of the following arises from the lung bud mesenchyme?

A. Epithelial lining of the alveolar sac.
B. Pseudostratified columnar epithelium.
C. Smooth muscles.
D. Type I cells.
E. Type II cells.

161. When does vertical transmission of human immunodeficiency virus most commonly occur?

A. First trimester.
B. Intrapartum.
C. Postpartum via breast-feeding.
D. Second trimester.
E. Third trimester.

162. Transcription is initiated by attachment of which enzyme to the promoter site upstream of the coding sequence of a gene?

A. DNA phosphodiesterase.
B. DNA polymerase.
C. DNA primase.
D. RNA helicase.
E. RNA polymerase.
163. The primary aim of angiotensin converting enzyme inhibitor (ACE-I) therapy in the treatment of congestive heart failure is to improve which physiological parameter?

A. Afterload.
B. Contractility.
C. Heart rate.
D. Myocardial relaxation.
E. Venous filling.

164. In children with very early onset inflammatory bowel disease (IBD), mutations affecting the immune system are common. In a 9-month-old diagnosed with IBD, mutations in the gene for which receptor are commonly found?

A. Interkelulin-1.
B. Interleukin-6.
C. Interleukin-7.
D. Interleukin-10.
E. Interleukin-28.

165. What is the inheritance pattern of complete androgen insensitivity syndrome?

A. Autosomal dominant.
B. Autosomal recessive.
C. Mitochondrial.
D. X-linked dominant.
E. X-linked recessive.
166. Which diuretic works by inhibiting the sodium-potassium-chloride cotransporter?

A. Amiloride.
B. Frusemide.
C. Hydrochlorothiazide.
D. Mannitol.
E. Spironolactone.
QUESTIONS 167 AND 168 REFER TO THE FOLLOWING INFORMATION

Results of a new screening test for detection of a rare form of childhood cancer among genetically at risk children are shown in the table below.

<table>
<thead>
<tr>
<th></th>
<th>Cancer +</th>
<th>Cancer −</th>
</tr>
</thead>
<tbody>
<tr>
<td>Test +</td>
<td>90</td>
<td>10</td>
</tr>
<tr>
<td>Test −</td>
<td>5</td>
<td>1195</td>
</tr>
</tbody>
</table>

What is the correct calculation for the following?

167. Sensitivity

A. \( \frac{90}{95} \)

B. \( \frac{90}{95} \div \left(1 - \frac{1195}{1205}\right) \)

C. \( \frac{90}{100} \)

D. \( \frac{95}{1205} \)

E. \( \frac{95}{1300} \)

F. \( \frac{1195}{1200} \)

G. \( \frac{1195}{1205} \)

H. \( \frac{1205}{1300} \)
168. Pre-test probability

A. \( \frac{90}{95} \)

B. \( \frac{90}{95} \div \left( 1 - \frac{1195}{1205} \right) \)

C. \( \frac{90}{100} \)

D. \( \frac{95}{1205} \)

E. \( \frac{95}{1300} \)

F. \( \frac{1195}{1200} \)

G. \( \frac{1195}{1205} \)

H. \( \frac{1205}{1300} \)
QUESTIONS 169 AND 170 REFERENCE THE FOLLOWING INFORMATION

Which respiratory parameter is defined by the following statement?

169. The maximal volume of air that can be inhaled above the tidal volume.

A. Anatomic dead space.
B. Expiratory reserve capacity.
C. Forced expiratory volume in 1 second.
D. Forced vital capacity.
E. Functional residual capacity.
F. Inspiratory reserve capacity.
G. Physiologic dead space.
H. Residual volume.

170. The volume of the lung that does not eliminate CO₂.

A. Anatomic dead space.
B. Expiratory reserve capacity.
C. Forced expiratory volume in 1 second.
D. Forced vital capacity.
E. Functional residual capacity.
F. Inspiratory reserve capacity.
G. Physiologic dead space.
H. Residual volume.