



Welcome to the MRCPCH Applied Knowledge in Practice Specimen Paper

This document provides a specimen Applied Knowledge in Practice (AKP) paper, designed to illustrate the types of questions used in that examination. It includes 24 sample questions with accompanying reasoning and references.

Important Note for Candidates

This specimen paper illustrates the format and style used in the AKP examination. Although the examination is based on current UK practice, the questions are written to ensure international candidates will understand the context. Please note that this paper is for illustrative purposes only and cannot reflect future examination papers in terms of distribution of questions (the Blueprint) or level of difficulty. While it can be a helpful resource for exam preparation, it should not be used as a definitive guide to the actual exam content.

In those situations where terminology of diagnoses, pathology findings or management descriptions has recently changed, the AKP exam will include both the new and the previous used terms in the answer key.

All questions that provide laboratory results will also include the age-adjusted normal ranges. These may differ slightly from local laboratories but are appropriate for interpretation of a given question

Expected exam conduct

It is strictly forbidden to talk, read the work of, or attempt in any way to communicate with other candidates whilst the examination is in progress. Please exercise vigilance to ensure that no other candidate can attempt to copy your work.

The College has tools that can identify the copying of answers or collusion between candidates to share answers. If such a situation were to arise, all identified parties will be investigated.

Unauthorised use of copyrighted exam materials or any form of malpractice may lead to suspension, permanent exclusion, and notification of the GMC or relevant medical board.

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Question 1 - Best answer and reasoning

Stem Question:

Which of the following is the most appropriate management?

Correct answer D: Reassurance

Reasoning

Also known as Benign Migratory Glossitis, the appearances are sometimes described as a 'geographic tongue'. There are sharply demarcated, irregular 'map-like', red, smooth patches with surrounding white plaques of desquamating material. These features are restricted to the dorsum of the tongue. The aetiology remains unclear.

Answer A is not the best answer

The papillae on the tongue are visible but – typically – fewer in number, not distorted and there is no evidence of erythema that would suggest malignancy or infection. Biopsy is not indicated.

Answer B is not the best answer

From the history, the child is well and therefore HIV would not be likely.

Answer C is not the best answer

This question is looking for confidence in a straightforward clinical diagnosis – a follow up review indicates unhelpful uncertainty.

Answer D is the best answer

This is a harmless condition that usually resolves with time. The mother and boy should be reassured. Further tip: Research images for other tongue pathologies such as 'Strawberry tongue', oral candida, lichen planus.

Answer E is not the best answer

Nystatin would be indicated if this was a fungal infection but again the papillae would be covered and usually includes the buccal mucosa (though not visible here).

Further Reading

<https://dermnetnz.org/topics/geographic-tongue>

Question 2 - Best answer and reasoning

Stem Question:

Which of the following can be concluded from the data?

Correct answer: E There was no difference in the level of neurological complications between treatment groups

Reasoning

'Odds ratios' (OR) compare the odds of a particular outcome occurring following one intervention and the outcome occurring following another intervention:

$$\text{OR} = \frac{\text{Outcome with Intervention 1}}{\text{Outcome with Intervention 2}}$$

If the ratio equals 1 (or the confidence interval given spans across 1) then there is no difference in the two interventions.

Answer A is not the best answer

The OR confidence interval crosses 1 and therefore there is no difference between the two interventions.

Answer B is not the best answer

If the paper has already been published in a recognised journal, it is likely that the methodology would have been reviewed and therefore any answer that indicates a poor design of the trial is unlikely to be correct. This is also a general statement not based on the data provided so can be excluded.

Answer C is not the best answer

There is no reference to this point in the data provided so it is not possible to draw this conclusion.

Answer D is not the best answer

There are no details of the clinical outcome, just information of the possible long-term effects. A change of practice cannot be proposed from the data given.

Answer E is the best answer

The OR provided crosses 1 meaning there is no difference in the level of neurological complications between the two treatment groups.

Question 3 - Best answer and reasoning

Stem Question:

Which of the following additional investigations is most likely to help establish the diagnosis?

Correct answer: B Galactose-1-phosphate uridyl transferase

Reasoning

A 6-week-old baby girl presenting with poor feeding, vomiting, hypoglycaemia, seizures, probable sepsis and a conjugated hyperbilirubinaemia raises the likelihood of a metabolic condition.

Answer A is not the best answer

Congenital CMV infection is a cause of conjugated hyperbilirubinaemia. However, there are none of the other usual clinical features suggestive of CMV. All answers should be plausible – one will be more plausible than the rest.

Answer B is the best answer

This history would explain both the early neonatal events and the recent presentation. The symptoms are all consistent with a diagnosis of galactosaemia. The question doesn't specify the cause of the sepsis. Including the fact that it was *E. coli* would make it too easy!

Answer C is not the best answer

Although hepatitis may give some of the findings listed, it would not explain the early neonatal history.

Answer D is not the best answer

A HIDA scan shows flow of a radioactive tracer through the hepatobiliary tree and helps diagnose biliary atresia. This needs to be considered as a cause of conjugated hyperbilirubinaemia but would not explain the early neonatal history.

Answer E is not the best answer

Would not explain the early neonatal problems and will give limited information on anatomy. It is also unlikely to be diagnostic as the question requests.

Further Reading

Nelson Textbook of Pediatrics. 22nd Edition.
Kliegman RM et al. Elsevier. Philadelphia 2024.

Applied Knowledge in Paediatrics MRCPC Mastercourse
M Hewitt. R Adappa. Elsevier, 2022

Question 4 - Best answer and reasoning

Stem Question:

Which of the following investigations is most likely to be diagnostic?

Correct answer: E Urinary catecholamines

Reasoning

This boy is unwell with clinical features highly suggestive of widespread malignancy involving the bone marrow (pale), bones (reluctant to be moved), lymphoreticular system (nodes) and abdomen (mass). The question asks for a *diagnostic* investigation and is a good example of the need to read the question carefully.

Answer A is not the best answer

An ultrasound would be requested and helpful to define the location of the mass but it would not be diagnostic as the question requests.

Answer B is not the best answer

A FBC would be requested and is likely to be abnormal but it would not be diagnostic.

Answer C is not the best answer

Liver function tests would be requested and are likely to be abnormal but would not be diagnostic.

Answer D is not the best answer

A raised alpha fetoprotein would be indicative of a hepatoblastoma, but the abdominal lesion described is more that of discrete lesion rather than an enlarged liver. Hepatoblastoma also usually presents before the age of 2 years and as a swollen abdomen in an otherwise well child.

Answer E is the best Answer

The history and examination are highly suggestive of neuroblastoma. The pallor, likely bone pain and the presence of left suprasternal lymph nodes (Virchow nodes) are indicative of metastatic disease. In this clinical context raised urinary catecholamines are diagnostic of neuroblastoma (although they can be negative in 5% of affected children).

Further Reading

Nelson Textbook of Pediatrics. 22nd Edition.

Kliegman RM et al. Elsevier. Philadelphia 2024.

Applied Knowledge in Paediatrics MRCPCH Mastercourse

M Hewitt. R Adappa. Elsevier, 2022

Question 5 - Best answer and reasoning

Stem Question:

What is the most likely diagnosis?

Correct answer: B Crohn colitis

Reasoning

Most diagnoses are made on the history. This girl is chronically unwell – she has pain, rectal bleeding, weight loss and, importantly, no longer feels well enough to participate in sport. The blood tests show a microcytic anaemia, likely to be due to iron deficiency, plus evidence of inflammation somewhere with the high CRP. The low albumin also suggests chronic illness, especially in the bowel. Finally, the faecal calprotectin is high which is a very useful indicator of bowel inflammation when neutrophils migrate to the bowel wall. Therefore, the most likely diagnosis is Crohn colitis.

Answer A is not the best answer

Isolated anal fissures cause pain and often fresh bleeding on defaecation, but they are not associated with chronic illness and elevated calprotectin unless linked to Crohn which can present with perianal disease.

Answer B is the best answer

This girl's symptoms, signs and investigation results make Crohn colitis the most likely diagnosis.

Answer C is not the best answer

Polyps can cause elevated calprotectin but not the chronic symptoms, signs and other test results described in this girl.

Answer D is not the best answer

Meckel diverticulum can cause elevated calprotectin but not the chronic symptoms, signs and other test results described in this girl.

Answer E is not the best answer

Gluten enteropathy is certainly plausible but not as the cause of the fresh blood in the stools. It is a disease of the small intestine.

Further Reading

Nelson Textbook of Pediatrics. 22nd Edition.
Kliegman RM et al. Elsevier. Philadelphia 2024.

Applied Knowledge in Paediatrics MRCPCH Mastercourse
M Hewitt. R Adappa. Elsevier, 2022

Question 6 - Best answer and reasoning

Stem Question:

What is the most likely diagnosis?

Correct answer: B Diabetes insipidus

Reasoning

This question tests knowledge of clinical biochemistry. The baby has chronic intermittent vomiting, longstanding faltering growth and deranged biochemistry specifically hypernatraemia with an inappropriately low urine osmolality. These features are characteristic of diabetes insipidus.

Answer A is not the best answer

This is a chronic history since birth making pyloric stenosis highly unlikely. An affected baby would also have normal physiology and be able to reduce their urine output and concentrate it appropriately when fluid intake is sub-optimal.

Answer B is the best answer

The urine osmolality is inappropriately low in the face of a high serum osmolality (easy to calculate if there is also have a glucose level but it can be estimated without) so diabetes insipidus is the correct answer.

Answer C is not the best answer

Cow's milk protein intolerance can lead to faltering growth and vomiting but this does not explain the abnormal biochemistry.

Answer D is not the best answer

The biochemistry is normal in Hirschsprung and the abdomen is likely to be distended, not scaphoid.

Answer E is not the best answer

In hypoaldosteronism, whatever the cause, there is increased renal tubular sodium loss resulting in hyponatraemia and compensatory hyperkalaemia.

Further Reading

Nelson Textbook of Pediatrics. 22nd Edition.

Kliegman RM et al. Elsevier. Philadelphia 2024.

Applied Knowledge in Paediatrics MRCPCH Mastercourse

M Hewitt. R Adappa. Elsevier, 2022

Question 7 - Best answer and reasoning

Stem Question:

In which of the following conditions is this physical sign a clinical feature?

Correct Answer: A Tricuspid atresia

Reasoning

The image shows digital clubbing i.e. loss of the angle between the nailbed and the nail. When associated with lung and heart conditions (commonest causes) it is a sign of chronic tissue hypoxia. However, the pathophysiology is still not fully understood, especially in the other causes such as gluten enteropathy and bronchiectasis, but may be linked to growth factors produced in chronic inflammation. Having identified the clubbing the next step is to look for a causative condition in the list of potential answers.

Answer A is the best answer

Tricuspid atresia is a cause of cyanotic congenital heart disease which can be associated with chronic tissue hypoxia and the development of clubbing, especially if left untreated.

Answer B is not the best answer

Cystic adenomatoid malformation is usually unilateral. It may present in the neonatal period with respiratory distress. Smaller lesions may present later in childhood with recurrent respiratory infections.

Answer C is not the best answer

Acute chest syndrome in a child with homozygous sickle disease may cause oxygen desaturation, but chronic hypoxia is not a feature, and children do not develop clubbing.

Answer D is not the best answer

Polycystic kidney disease may lead to chronic renal failure, but hypoxia and clubbing are not seen in this condition.

Answer E is not the best answer

Pulmonary stenosis is a form of acyanotic congenital heart disease, and clubbing is not a feature.

Further Reading:

Nelson Textbook of Pediatrics. 22nd Edition.

Kliegman RM et al. Elsevier. Philadelphia 2024.

Applied Knowledge in Paediatrics MRCPCH Mastercourse

M Hewitt. R Adappa. Elsevier, 2022

Question 8 - Best answer and reasoning

Stem Question:

Which of the following is the most important action to take before commencing the EEN regime?

Correct answer: C Correct hypokalaemia

Reasoning

The patient has a low BMI for her age as a result of malabsorption from Crohn disease. The significant investigation abnormalities are anaemia and hypokalaemia although others are borderline. The ferritin might be spuriously high in conjunction with the raised CRP as both are acute phase proteins. She is high risk for 'refeeding syndrome' when, amongst other significant changes, commencing nutrition causes rapid cellular uptake of potassium, phosphate and magnesium. The potassium is already extremely low and this should be corrected prior to starting feeds and she will need frequent and careful biochemical and clinical evaluation.

Answer A is not the best answer

Returning the haemoglobin to normal with oral iron will take months and 92 g/l is not life-threatening. Oral iron is also very unpalatable and adherence unlikely to be optimal.

Answer B is not the best answer

Although B12 deficiency is possible, especially with ileocaecal Crohn, and might need replacement, it does not need normalising prior to EEN.

Answer C is the best answer

Correction plus ongoing monitoring is required to minimise the risk of dangerously low levels as feeds are introduced and increased.

Answer D is not the best answer

She will be started on multivitamins, including Vitamin B1 (thiamine), but no correction is needed before EEN.

Answer E is not the best answer

A haemoglobin of 92 g/l is not life-threatening.

Further Reading:

<https://bspghan.org.uk/wp-content/uploads/2023/01/ecco-consensus-paediatric-crohns-2020.pdf> https://www.uhbw.nhs.uk/assets/1/23-639_refeedingsyndromeguideline-4_redacted.pdf

Question 9 - Best answer and reasoning

Stem Question:

Which of the following is the most appropriate management?

Correct answer: B Oral ciprofloxacin and nebulised colomycin for 3-months

Reasoning

The current UK practice for the management of a first *Pseudomonas* infection in patients with cystic fibrosis (CF) aims for eradication using 'dual therapy'. A course of oral antibiotics (IV if clinically unwell) in combination with an inhaled antibiotic via a nebuliser. An extended course of both should then follow. This is expected knowledge and is an example of a question based on NICE guidelines

Answer A is not the best answer

Single agent antibiotic only.

Answer B is the best answer

Dual therapy with oral and nebulised antibiotics.

Answer C is not the best answer

This option does not include an antibiotic. Dornase alpha is licensed for use in some CF patients to improve lung function by reducing sputum viscosity prior to physiotherapy.

Answer D is not the best answer

This option does not include an antibiotic. Hypertonic saline is used as a mucolytic in CF but has no effect in bacterial infections.

Answer E is not the best answer

Single agent antibiotic only.

Further Reading

<https://bnf.nice.org.uk/treatment-summaries/cystic-fibrosis/>

Question 10 - Best answer and reasoning

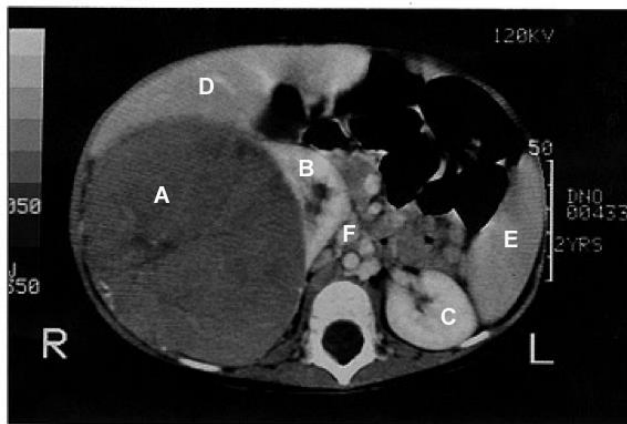
Stem Question:

What abnormality is seen on the CT scan with IV contrast shown below?

Correct answer: C Solid tumour of the right kidney

Reasoning

Questions are asked in this exam that test candidate knowledge of basic anatomy and interpretation of CT, MR and USS images. Here IV contrast has been given which enhances vascular structures making them appear white. The view provided is a transverse section and the view is from the inferior position looking towards the head – the R and L markers indicate the appropriate side



The vertebra is evident posteriorly in the midline at the bottom of the image. Normal sized left kidney (C). Large mass (A) arising from the right kidney (B - enhanced). Liver (D) and spleen (E).

Answer A is not the best answer

The bowel gas pattern is of normal calibre although displaced slightly to the left. The mass is solid with no evidence of abscess formation. An appendix abscess is usually pelvic.

Answer B is not the best answer

There are no cysts evident in the right renal tissue (B). Or in the left kidney (C).

Answer C is the best answer

This is a solid tumour of the right kidney probably a nephroblastoma (Wilms) tumour.

Answer D is not the best answer

An enlargement of the adrenal tissue (neuroblastoma) would displace the right kidney inferiorly but would maintain the normal renal shape.

Answer E is not the best answer

The area of liver visible is normal in appearance and is not displaced.

Further Reading

Nelson Textbook of Pediatrics. 22nd Edition.

Kliegman RM et al. Elsevier. Philadelphia 2024.
Applied Knowledge in Paediatrics MRCPCH Mastercourse
M Hewitt. R Adappa. Elsevier, 2022

Question 11 - Best answer and reasoning

Stem Question:

What is the most likely diagnosis?

Correct answer: A Pulmonary stenosis

Reasoning

The ECG shows that the child is in sinus rhythm with a normal PR interval. There is marked right axis deviation of 150° as shown by a negative QRS complex in Lead I and upward in Lead III. There is also right ventricular hypertrophy (RVH) with tall R waves over the right chest leads.

Answer A is the best answer

Of the congenital heart disease conditions listed, the only type which is acyanotic and associated with right axis deviation and RVH is pulmonary stenosis.

Answer B is not the best answer

The main reason is that a child with Tetralogy of Fallot would be cyanosed. The ECG does show the right axis deviation and RVH typical of uncorrected Fallot but in a 5yr old UK child this situation would be extremely unlikely.

Answer C is not the best answer

In an uncomplicated patent arterial duct, the child is pink and well, and there are no diagnostic ECG features.

Answer D is not the best answer

An ostium primum atrial septal defect (ASD) is also a type of acyanotic congenital heart disease where the ECG typically shows right bundle branch block and right axis deviation but without the RVH. In ostium secundum defects the ECG may be normal or show right axis deviation and/or a tall p wave due to atrial enlargement and/or incomplete right bundle branch block.

Answer E is not the best answer

A child with an uncomplicated ventricular septal defect is pink. The ECG is typically normal.

Further Reading

Nelson Textbook of Pediatrics. 22nd Edition.

Kliegman RM et al. Elsevier. Philadelphia 2024.

Applied Knowledge in Paediatrics MRCPCH Mastercourse

M Hewitt. R Adappa. Elsevier, 2022

Question 12 - Best answer and reasoning

Stem Question:

What is the most likely diagnosis?

Correct answer: D Long QT syndrome

Reasoning

Episodes of sudden collapse with no warning, especially during exertion, are suggestive of a cardiac cause which must be excluded.

Answer A is not the best answer

A plausible answer from the history but the cardiac examination was normal so much less likely.

Answer B is not the best answer

Vasovagal episodes are common especially in rapidly growing teenagers. But there is almost always some warning such as blurred or double vision or dizziness that the patient remembers when they recover.

Answer C is not the best answer

The history does not describe any automatisms or unusual behaviours prior to the collapse (typical of Focal Impaired Awareness seizures) nor any abnormal movements during the episodes or a post-ictal phase. This seizure type was previously termed Complex Partial Seizures. In such instances with a recent change in nomenclature candidates will be given both names in the answer key.

Answer D is the best answer

Episodes of loss of consciousness during exertion suggest a cardiac disorder. Any child presenting in this way should have an ECG that in this patient will show a prolonged QTc of greater than 0.46 msec.

Answer E is not the best answer

Hypoglycaemic episodes rarely cause seizures and loss of consciousness because the patient would almost certainly have symptoms such as sweating, hunger, anxiety, weakness and headaches in advance that they or others will recognise and intervene. They will also get warning from any indwelling Continuous Glucose Monitor that most patients now wear.

Further Reading

Nelson Textbook of Pediatrics. 22nd Edition.

Kliegman RM et al. Elsevier. Philadelphia 2024.

Applied Knowledge in Paediatrics MRCPCH Mastercourse

M Hewitt. R Adappa. Elsevier, 2022

Question 13 - Best answer and reasoning

Stem Question:

Which of the following diagnoses best explains this presentation?

Correct answer: C - Congenital lung cyst

Reasoning

The obvious abnormality on this chest x-ray is the round, air-filled lesion in the right lower lobe [*Make sure you have a routine for reviewing x-rays*]. This child is basically well with a cough which will exclude those answers leading to significant lung compromise.

Answer A is not the best answer

Would not lead to a large air-filled space and would also have solid elements visible on x-ray.

Answer B is not the best answer

Congenital lobar emphysema (or congenital lobar hyperinflation) generally affects the upper lobe, or right middle lobe. The hyperlucency would be in a lobar pattern. This is in the lower lobe area and not lobar. The child would be unwell with a tachypnoea.

Answer C is the best answer

A localised air-filled lesion in a well-child would explain the history and findings.

Answer D is not the best answer

The child would be unwell with a tachypnoea. A loculated pneumothorax would be unlikely, as it is normally loculated due to pleural adhesions and would be unlikely to persist for a month.

Answer E is not the best answer

A pneumatocoele is seen in Staphylococcal lung infection and the patient would be extremely unwell.

Further Reading

Nelson Textbook of Pediatrics. 22nd Edition.

Kliegman RM et al. Elsevier. Philadelphia 2024.

Applied Knowledge in Paediatrics MRCPCH Mastercourse
M Hewitt. R Adappa. Elsevier, 2022

Question 14 - Best answer and reasoning

Stem Question:

Which of the following diagnoses best explains this presentation?

Correct answer: E - Reflex anoxic seizures

Answer A is not the best answer

This condition is not usually seen in this age group and is often characterised by seizures involving facial twitching, numbness, or tingling, and speech difficulties. Various previous names including Benign Rolandic epilepsy.

Answer B is not the best answer

The history does not describe seizures with abnormal, coordinated actions.

Answer C is not the best answer

Child has a 'mild elevation of temperature' prior to one these episodes but that is unlikely to cause seizure activity.

Answer D is not the best answer

This condition is not usually seen in this age group and the episodes in the presented scenario here, are described as 'shaking' and not 'jerking'.

Answer E is the best answer

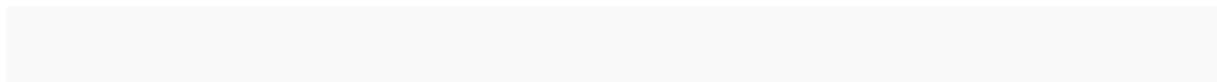
Classical history where pain or anger initiates a 'crying response' and the child collapses often prior to the onset of the cry. Benign, self-limiting and resolve in time.

Further Reading

<https://www.medlink.com/articles/reflex-anoxic-seizures>
[Accessed June 2025]

NICE Guideline: Epilepsies in children, young people and adults (NG217) Last updated January 2025

Applied Knowledge in Paediatrics MRCPCH Mastercourse
M Hewitt. R Adappa. Elsevier, 2022



Question 15 - Best answer and reasoning

Stem Question:

Based on the information provided above, which of the following statements is the most appropriate conclusion to draw?

Correct answer: D - There was a reduction in the need for exchange transfusion in those treated with intravenous immunoglobulin.

Reasoning

The “95% confidence interval” means that we are 95% confident that the population value of interest lies within this given range.

The “relative risk” indicates the risk of an event occurring in an exposed group when compared with the risk of the event occurring in an unexposed group. A “relative risk” (expressed as a ratio) around 1 indicates little or no difference, greater than 1 indicates a greater risk of the event happening in the exposed group and a value less than 1 indicates a reduced risk of the event happening in the exposed group.

Answer A is not the best answer

The concept of ‘Numbers Needed to Treat’ is a measure of how many individuals need to be treated for one individual to benefit. In this study for every 2.7 individuals treated, 1 patient will have a beneficial result with immunoglobulins when compared with the control group. It does not refer to the number of transfusions.

Answer B is not the best answer

This statement is the opposite of what was found. The number of exchange transfusions per infant was less in the group that received immunoglobulin.

Answer C is not the best answer

A systematic review identifies those published research studies which meet a predefined set of criteria and thereby allow specific questions to be addressed. If a study meets the criteria, then it is included. The design of the study cannot then be challenged if it meets these criteria.

Answer D is the best answer

There was a reduction in the need for exchange transfusion in those treated with intravenous immunoglobulin as the weighted mean was a negative value [-0.52] and the confidence interval range was negative – i.e. fewer exchanges needed.

Answer E is not the best answer

While the review is robust, evidence of the effect of intravenous immunoglobulins on the need for exchange transfusion, the introduction of this therapy as routine would require a careful risk-benefit assessment, which cannot be made on the data available.

Further Reading

R Gottstein, R W I Cooke Systematic review of intravenous immunoglobulin in haemolytic disease of the newborn. Arch Dis Child Fetal Neonatal Ed 2003;88:

Applied Knowledge in Paediatrics MRCPCH Mastercourse

M Hewitt. R Adappa. Elsevier, 2022

Question 16 - Best answer and reasoning

Stem Question:

What is the most important initial investigation?

Correct answer: E - Intraocular pressure measurement

Reasoning

For the candidate, this is a two-stage question. The first stage is to make a diagnosis, and the appearance of a unilateral capillary haemangioma (port wine stain) is typical of the Sturge-Weber syndrome.

The second stage is to consider complications of this syndrome that may occur in infancy and consider an appropriate investigation for that condition.

Note that the questions ask:

What is the most important **INITIAL** investigation?

Some of these investigations would be required for this child but would not be required in the first instance.

This illustrates the importance of understanding exactly what is being asked in a particular question.

Answer A is not the best answer

The main cause of SWS is somatic mosaic GNAQ pathogenic variants. The contribution of such a test, if available, to management would need further consideration and a discussion of implications for the family. It is not an initial test.

Answer B is not the best answer

An MRI scan with gadolinium is important to look for an associated intracranial vascular abnormality, but there is no immediate urgency for this.

Answer C incorrect

Cranial ultrasound is unlikely to show a vascular abnormality at this location and is not indicated.

Answer D is not the best answer

Although around 80% of children with Sturge-Weber Syndrome develop epilepsy, an EEG is not indicated until the child presents with seizures.

Answer E is the best answer

Glaucoma affecting the ipsilateral eye is a common complication in infancy (affecting 50% of patients) and appropriate initial assessment is mandatory. It presents in newborns with enlargement of the globe (buphthalmos).

Further Reading

Nelson Textbook of Pediatrics. 22nd Edition.

Kliegman RM et al. Elsevier. Philadelphia 2024.

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M Hewitt. R Adappa. Elsevier, 2022

Question 17 - Best answer and reasoning

Stem Question:

Which of the following diagnoses best explains this presentation?

Correct answer: D - Transposition of the great arteries with VSD

Reasoning

The important features to recognise are 3 days old, cardiac signs, (including cyanosis) metabolic acidosis and response to prostaglandins.

Answer A is not the best answer

Aortic stenosis could produce the symptoms recorded but would not produce the observed central cyanosis. The improvement in the oxygenation with the prostaglandin as described in this question would not occur in aortic stenosis.

Answer B is not the best answer

AVSD would produce increased pulmonary blood flow and possibly heart failure but do not produce cyanosis and would not improve with prostaglandin.

Answer C is not the best answer

Babies with pulmonary atresia become acutely unwell as the duct closes within the first few hours. Prostaglandin E₁ infusion is necessary and early surgical intervention required. The child would not present at 3 days of age.

Answer D is the best answer

A baby with TGA and VSD has some mixing of oxygenated and deoxygenated blood in the ventricles depending on the size of the VSD. If the VSD is small, then the lesion is duct dependent but may present later.

Answer E is not the best answer

A combined outflow tract from both left and right ventricles allows mixing of bloods and is unlikely to present with acidosis. The lesion is not duct dependent, so would not improve with prostaglandin.

Further Reading

Nelson Textbook of Pediatrics. 22nd Edition.

Kliegman RM et al. Elsevier. Philadelphia 2024.

Applied Knowledge in Paediatrics MRCPCH Mastercourse,
Hewitt. R Adappa. Elsevier, 2022

Question 18 - Best answer and reasoning

Stem Question:

Which of the following diagnoses best explains this presentation?

Correct answer: A - Benign sleep myoclonus

Reasoning

The diagnosis of the cause of 'funny turns' or abnormal movements in any child relies predominantly on the history. Here we have jerking movements seen only when the child sleeps. They are of short duration.

Answer A is the best answer

This is not the history of pathological seizures of any type – the episodes are limited to the time the baby is asleep. Benign sleep myoclonus becomes the most likely diagnosis by the exclusion of the other offered diagnoses. Benign sleep myoclonus is a common and self-limiting condition.

Answer B is not the best answer

Symptoms from drug withdrawal are not confined to sleep and would not be increasing in frequency at this age.

Answer C is not the best answer

Hypocalcaemia characteristically causes 'jitteriness' but, if severe, can lead to generalised seizures. Symptoms from hypocalcaemia are not confined to times of sleep.

Answer D is not the best answer

Symptoms from infantile spasms are not confined to sleep.

Answer E is not the best answer

Symptoms from myoclonic epilepsy are not confined to sleep.

Further Reading

UpToDate:

<https://www.uptodate.com/contents/nonpileptic-paroxysmal-disorders-ininfancy> Authors: Nguyen TT; Kaplan PW; Wilfong A; [Accessed May 2018]

Nelson Textbook of Pediatrics. 22nd Edition.

Kliegman RM et al. Elsevier. Philadelphia 2024.

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M Hewitt. R Adappa. Elsevier, 2022

Question 19 - Best answer and reasoning

Stem Question:

Pending transfer to the tertiary NICU, which of the following is the most appropriate next action?

Correct answer: E - Commence passive cooling

Reasoning

This baby has had a traumatic birth, leading to hypoxic-ischaemic insult. Features include the need for resuscitation, neurological depression, seizures and electroencephalographic abnormalities. There is an increasing risk of death or neurodevelopmental abnormalities. Early reduction of body temperature has been shown to improve outcome.

Answer A is not the best answer

This intervention is not evidence-based.

Answer B is not the best answer

This intervention is not evidence-based.

Answer C is not the best answer

This intervention is not evidence-based.

Answer D is not the best answer

This intervention is not evidence-based.

Answer E is the best answer

This intervention has been shown to affect outcome in severe asphyxia. This baby fulfils the cooling criteria from the UK TOBY study register.

Further Reading

Anzopardi DV et al Moderate hypothermia to treat perinatal asphyxial encephalopathy. NEJM 2009;361(14) 1349-58

Strohm, B et al; Temperature control during therapeutic moderate whole-body hypothermia for neonatal encephalopathy. Arch Dis Child Fetal Neonatal Ed 2010; 9(1):39-45

TOBY trial protocols: <https://www.npeu.ox.ac.uk/toby/protocol> [Accessed May 2018]

Therapeutic Hypothermia for Neonatal Encephalopathy. A BAPM Framework for Practice. 2020

<https://www.bapm.org/resources/237-therapeutic-hypothermia-for-neonatal-encephalopathy>

Question 20 - Best answer and reasoning

Stem Question:

Which of the following diagnoses best explains this presentation?

Correct answer: A - Arnold-Chiari malformation type 1

Reasoning

Headaches are a common problem presenting in out-patients and one that depends heavily on a clear history. Certain 'red flag' features can indicate significant pathology. This young person has a relatively long history with some rather strange aspects – frequent, improve when lying down and added description of balance problems with exercise. The neurological examination identifies abnormalities.

Answer A is the best answer

The Arnold-Chiari malformation type 1 is a herniation of the cerebellar tonsils through the foramen magnum [Arnold-Chiari malformation type 2 are often associated with spinal dysraphism and Arnold-Chiari malformation type 3 is extremely rare].

Answer B is not the best answer

Although Idiopathic (benign) intracranial hypertension does lead to chronic headaches of a persistent nature, it is not associated with abnormal neurological findings apart from blurred optic discs.

Answer C is not the best answer

A Becker muscular dystrophy leads to features of myopathy that would mainly include weakness of lower limbs.

Answer D is not the best answer

Although there is a strong family history of migraine, this is not the pattern of headaches seen in classical migraine – these usually with episodic headaches with longer, symptom-free, periods. One would not expect abnormal neurology on examination.

Answer E is not the best answer

A spinal cord tumour would not give a history of headaches.

Further Reading

Nelson Textbook of Pediatrics. 22nd Edition.

Kliegman RM et al. Elsevier. Philadelphia 2024.

Applied Knowledge in Paediatrics MRCPCH Mastercourse

M Hewitt. R Adappa. Elsevier, 2022

Question 21 - Best answer and reasoning

Stem Question:

Which of the following diagnoses best explains this presentation?

Correct answer: D - Intussusception

Reasoning

This girl has an acute abdomen with evidence of shock. In this context the rectal bleeding is suggestive of ischaemic bowel. In this list the only 2 possibilities causing this are volvulus and intussusception. Although intussusception is most common in infants and toddlers, about 10% of cases occur in children over five years and 3-4 % in those over 10 years; and 1% in infants <3 months. Outside the typical age range, it is likely to be associated with a pathological 'lead' point, which may include reactive lymphoid hyperplasia, small bowel lymphoma or a Meckel's diverticulum.

Answer A is not the best answer

Would expect more of a history of diarrhoea and fever over a shorter period. It can give rectal bleeding but not usually a major feature.

Answer B is not the best answer

Although such patients have abdominal pain, the important description is 'spasmodic pain' and some features may have been seen on previous ultrasound.

Answer C is not the best answer

No description of a characteristic rash or joint pain. Also, the previous history makes less likely.

Answer D is the correct answer

See explanation given at introduction to the reasoning in this question.

Answer E is not the best answer

See explanation given at introduction.

Further Reading

Nelson Textbook of Pediatrics. 22nd Edition.

Kliegman RM et al. Elsevier. Philadelphia 2024.

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Question 22 - Best answer and reasoning

Stem Question:

Which of the following diagnoses best explains this presentation and image?

Correct answer: C - Staphylococcal scalded skin syndrome

Reasoning

The child has a diffuse erythema of the face with peeling of the skin peri-orally, around the eyes and in the neck. The epidermis has separated from the dermis to form blisters that have then ruptured. The folds of lifted epidermis can be seen (above both eyes, left cheek, mouth, chest). This feature is typical of staphylococcal scalded skin syndrome.

Answer A is not the best answer

Atopic dermatitis in infancy characteristically involves the cheeks and scalp. The skin is erythematous with dry crusting lesions. Perioral involvement and desquamation are not features of this condition and the unaffected scalp is also against this diagnosis.

Answer B is not the best answer

Impetigo usually starts with a vesicle or pustule which develops into a crusted, oozing lesion which then gradually spread.

Answer C is the best answer

Answer D is not the best answer

Stevens-Johnson syndrome starts with erythematous macules with central necrosis. The lesions then form vesicles, bullae and desquamation. The condition involves two or more mucosal surfaces which may include the eyes, oral cavity upper airway and G-I tract. Erythromycin is known to cause SJS but the features are not consistent with that diagnosis.

Answer E is not the best answer

Toxic epidermal necrolysis is usually the result of a drug reaction and erythromycin is a recognised cause but, like Stevens -Johnson syndrome, usually involves the mucosal surfaces.

Further Reading

Nelson Textbook of Pediatrics. 22nd Edition.
Kliegman RM et al. Elsevier. Philadelphia 2024.

Applied Knowledge in Paediatrics MRCPCH Mastercourse

M Hewitt. R Adappa. Elsevier, 2022

Question 23 - Best answer and reasoning

Stem Question:

Which of the following conditions should be screened for at this age?

Correct answer: D - Nephropathy

Answer A is not the best answer

Would not form part of an annual review, although offered at diagnosis and if symptomatic.

Answer B is not the best answer

Would not form part of an annual review.

Answer C is not the best answer

Would not form part of an annual review for type 1 diabetes. It is recommended for those with Type 2 diabetes.

Answer D is the best answer

Vasculopathy is a significant complication in all forms of diabetes and will affect renal tissue. Annual check and early intervention for microalbuminuria is vital.

Answer E is not the best answer

Peripheral neuropathy is a recognised complication of type 1 diabetes but is very rare at this age and is not screened for until adulthood.

Further Reading

<https://www.nice.org.uk/guidance/ng18/chapter/Recommendations#type-1-diabetes>

Nelson Textbook of Pediatrics. 22nd Edition.

Kliegman RM et al. Elsevier. Philadelphia 2024.

Applied Knowledge in Paediatrics MRCPCH Mastercourse

M Hewitt. R Adappa. Elsevier, 2022

Question 24 - Best answer and reasoning

Stem Question:

Which of the following diagnoses best explains this presentation?

Correct answer: D - Pneumocystis jirovecii pneumonia

Answer A is not the best answer

Influenza A pneumonia would not be expected to give such marked hypoxia in the early phases.

Answer B is not the best answer

Pneumococcal pneumonia causing such marked hypoxia would produce a toxic presentation in the patient with much higher temperatures.

Answer C is the best answer

Pneumocystis jirovecii pneumonia is seen in patients who are immunosuppressed and they will often have marked hypoxia despite a paucity of chest findings on auscultation. Often occurs after prolonged insult to immune system.

Answer D is not the best answer

Relapse of mediastinal Hodgkin lymphoma may cause some features such as pyrexia but individuals would usually present with symptoms long before they developed such hypoxia.

Answer E is not the best answer

Respiratory Syncytial Virus pneumonia would not be expected to give such marked hypoxia in the early phases.

Further Reading

Nelson Textbook of Pediatrics. 22nd Edition.

Kliegman RM et al. Elsevier. Philadelphia 2024.

Applied Knowledge in Paediatrics MRCPCH Mastercourse
M Hewitt. R Adappa. Elsevier, 2022