Applied Knowledge in Practice – Sample Paper 2
Answer Key

Question 1 (C 0012 SP)
Subject: Infection, Immunology and Allergy
Answer: E - Offer reassurance

Reasoning:
The appearances are sometimes described as a 'geographic tongue' – where there are areas of sharply demarcated, irregular red patches with surrounding white plaques. These features are restricted to the dorsum of the tongue. The aetiology remains unclear.

Answer A is wrong
The papillae on the tongue are visible and not distorted and there is no evidence of erythema that would suggest malignancy or infection. Biopsy is not indicated.

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

Answer C is wrong
This question wants you to be confident and reassure – a follow up review indicates you are unsure!!

Answer D is wrong
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).

Answer E is the best answer
Also known as Benign Migratory Glossitis. The white coating is desquamating material. You can reassure.

Tip: research images for 'Strawberry tongue', oral candida, lichen planus.

Further Reading
Nelson Textbook of Paediatrics. 20th Edition. Kliegman RM et el. Elsevier. Philadelphia 2016. Chapter XXXI. The Skin

Question 2 (D 0009 SP)
Subject: Neonatology
Answer: B - Low maternal platelet count in pregnancy

Reasoning:
This child presents with bruising on day 3 of life.

Answer A is wrong
A positive family history is indicative of an inherited clotting problem and not a thrombocytopaenia.

Answer B is the best answer
In maternal ITP, Neonatal Alloimmune Thrombocytopenia can occur due to trans-placental transfer of maternal antiplatelet autoantibodies.

Answer C is wrong
The given history does not indicate that mother was taking anti-epileptic medication

Answer D is wrong
The child is well so sepsis is unlikely to be the cause of the low platelets.

Answer E is wrong
Vitamin K is given to protect against haemorrhagic disease of the newborn – a problem of clotting, not thrombocytopaenia.

Further Reading
Nelson Textbook of Pediatrics. 20th Edition. Kliegman RM et el. Elsevier. Philadelphia 2016. Chapter XXI. Diseases of the Blood

Question 3 (EBM 0011 SP)
Subject: Science of Practice
Answer: D - Short course treatment resulted in a significantly reduced length of hospital stay.
Answer: F - There was no difference in the level of neurological complications between treatment groups.

Reasoning:
'Odds ratios' compare the odds of a particular outcome occurring following one intervention and the outcome occurring following another intervention [OR=Outcome with Intervention 1/ 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 wrong
The OR confidence interval crosses 1 and therefore there is no observed difference between the two interventions.

Answer B is wrong
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 a general statement and not based on the data provided so should be excluded

Answer C is wrong
There is no Further Reading to this point in the provided data and so not possible to comment.

Answer D is the best answer
The difference in length of stay is -2.17 days and the CI values are all negative indicating a reduced duration of stay – and, intuitively you would suggest this is true!!

Answer E is wrong
You are not provided with details of the clinical outcome just information of the possible long-term effects. You cannot propose a change of practice from the details given.

Answer F is the best answer
The provided OR crosses 1 and that suggests no difference.

Question 4 (7003 SP)
Subject: Neurology
Answers: A - Benign seizures of childhood

Reasoning:

Answer A is the best answer

The seizures are often at night and are infrequent in nature. The prognosis is very good and seizures will resolve in most children often by the early teenage years.

Answer B is wrong

The history does not describe seizures with abnormal, coordinated actions

Answer C is wrong

The history given does not support this diagnosis

Answer D is wrong

The blood glucose was within the normal range when tested during a recent episode

Answer E is wrong

The episodes are described as 'shaking' and not 'jerking'

Answer F is wrong

The patient would usually present with sudden collapse.

Further Reading

Nelson Textbook of Pediatrics. 20th Edition. Kliegman RM et el. Elsevier. Philadelphia 2016. Chapter XXVII. The Nervous System

Question 5 (D 0010 SP)
Subject: Metabolism and Metabolic Medicine
Answer: B - Galactose-1-phosphate uridyl transferase level

Reasoning:
A 6 week old baby girl presenting with poor feeding, vomiting, hypoglycaemia, seizures and a conjugated hyperbilirubinaemia. Raise the likelihood of a metabolic condition.

Answer A is wrong
Intracranial abnormalities would not give a conjugated hyperbilirubinaemia.

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.

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

Answer D is wrong
A biliary atresia needs to be considered but such a diagnosis would not explain the early neonatal history.

Answer E is wrong
Congenital hypothyroidism produces an unconjugated hyperbilirubinaemia

Answer F is wrong
Would not explain the early neonatal problems and will give limited information on anatomy.

Further Reading
Nelson Textbook of Pediatrics. 20th Edition. Kliegman RM et el. Elsevier. Philadelphia 2016. Chapter XI. Metabolic Disorders

Question 6 (EBM 0043 SP)
Subject: Science of Practice
Answers: B - Assessment of whether the child looks ill is a highly sensitive test for serious invasive bacteraemia.

Reasoning:
Sensitivity could also be called the 'True Positive Rate' – the ability of the test to identify those with the condition of interest. In this example the condition of interest is 'bacteraemia'.

Specificity could also be called the 'True Negative Rate' – the ability of the test to exclude those who do not have the condition of interest.

Answer A is wrong
The white count is a valuable assessment tool but that is not the same as – 'All febrile children with petechiae should have a white blood cell count performed'.

Answer B is the best answer
'53 patients appeared ill, including all 6 with serious invasive bacteraemia'. Observation was able to identify all with bacteraemia though the false positive rate was high.

Answer C is wrong
The defined outcome is 'bacteraemia' and the question asks about 'specificity' of 'looking ill'. So, could the 'test' (clinical assessment) exclude those with the condition (bacteraemia). Eight children had bacteraemia but 53 were judged as unwell. The test is not specific.

Answer D is wrong
Although the study suggests that those children with white cell count between 5,000 and 15,000 are not in the likely to have a bacteraemia it does not lead onto saying they should be allowed home. Eight children had bacteraemia but 53 looked unwell.

Answer E is wrong
Low or high white counts were sensitive of bacteraemia but there is no comment about the relationship of intermediate white count. Therefore, one cannot make any decision on management based on this unreported result.

Answer F is wrong
It is always important to ask whether the population studied is the same as the population encountered in one's clinical practice. In this case children in the UK are very similar to those in the United States.

Question 7 a (EMQ 0005 SP a)
Subject: Endocrinology and Growth
Answer: H - Premature thelarche (common, self-limiting)

Reasoning:

Answer A is wrong

Constitutional delay of growth and puberty causes delay of puberty.

Answer B is wrong

Growth hormone deficiency causes delay of puberty.

Answer C is wrong

Hypothalamic hamartoma is associated with abnormal, early puberty with both breast and pubic hair development.

Answer D is wrong

Idiopathic precocious puberty is associated with abnormal, early puberty with both breast and pubic hair development.

Answer E is wrong

Klinefelter syndrome will only affect boys

Answer F is wrong

McCune-Albright Syndrome is associated with abnormal, early puberty with both breast and pubic hair development.

Answer G is wrong

Premature adrenarche is due to mild androgen production from the zona reticulosa of the adrenal gland as it matures and so causes pubic hair that is almost always confined to the vulva.

Answer H is the best answer

Answer I is wrong

Testicular tumour will only affect boys

Answer J is wrong

Turner Syndrome causes delayed puberty in girls.

Further Reading

UpToDate https://www.uptodate.com/contents/definition-etiology-and-evaluation-of-precocious-puberty Authors: Harrington J; Palmert MR. [Accessed May 2018]

Question 7 b (EMQ 0005 SP b)
Subject: Endocrinology and Growth
Answer: J - Turner syndrome

Reasoning:

By definition, menarche is not considered to be delayed until the age of 16 years. It comes towards the end of normal puberty ie 2-3 years following the first signs of secondary sexual characteristics.

Answer A is wrong

Constitutional delay is possible especially if the parents are short but this information is not provided in the question stem.

Answer B is wrong

Growth hormone deficiency is possible if a positive family history was provided.

Answer C is wrong

Hypothalamic hamartoma is associated with abnormal, early puberty with both breast and pubic hair development.

Answer D is wrong

Idiopathic precocious puberty is associated with abnormal, early puberty.

Answer E is wrong

Klinefelter syndrome will only affect boys

Answer F is wrong

McCune-Albright Syndrome is associated with abnormal, early puberty.

Answer G is wrong

Premature adrenarche is due to mild androgen production from the zona reticulosa of the adrenal gland as it matures and so causes pubic hair that is almost always confined to the vulva.

Answer H is wrong
Breast development is not mentioned in the history.

Answer I is wrong
Testicular tumour will only affect boys

Answer J is the best answer

Further Reading
UpToDate: https://www.uptodate.com/contents/clinical-manifestations-and-diagnosis-of-turner-syndrome Author: Backeljauw P
UpToDate: https://www.uptodate.com/contents/approach-to-the-patient-with-delayed-puberty Authors: Crowley WF; Pitteloud N. [Accessed May 2018]

Question 7 c (EMQ 0005 SP c)
Subject: Endocrinology and Growth
Answer: A - Constitutional growth and pubertal delay

Reasoning:
Of the causes of delayed puberty in boys on this list the possible answers are Klinefelter syndrome, growth hormone (GH) deficiency and constitutional delay of growth and puberty (CDGP).

Answer A is the best answer
The question asks for the 'most likely diagnosis' which is therefore the common CDGP. This can be a source of misery for affected young men and responds well to a short course of low dose testosterone.

Answer B is wrong
GH deficiency is possible but very rare to be diagnosed at this age.

Answer C is wrong
Hypothalamic hamartoma is associated with abnormal, early puberty.

Answer D is wrong
Associated with early puberty.

Answer E is wrong
Klinefelter characteristically causes disproportionate relative tall stature.

Answer F is wrong
Associated with early puberty.

Answer G is wrong
The history is about delay in puberty.

Answer H is wrong
The history is about delay in puberty.

Answer I is wrong
Testicular problem not mentioned in history

Answer J is wrong
Found only in females

Further Reading
UpToDate: https://www.uptodate.com/contents/approach-to-the-patient-with-delayed-puberty Authors: Crowley WF; Pitteloud N. [Accessed May 2018]

Question 8 (C 0017 SP)
Subject: Cardiology
Answer: E - Subacute bacterial endocarditis

Reasoning:
Dark purple/black (ischaemic) lesions on the big toe and on the plantar surface of the foot. These are appearances of the effect of vascular occlusion.

Answer A is wrong
Deep vein thrombosis would usually lead to swelling of the distal limb and this is not evident here

Answer B is wrong
The resulting lesions from ITP would be distributed throughout the torso and limbs

Answer C is wrong
Meningococcal septicaemia causing such lesions would be indicative of overwhelming and rapidly progressive infection. The child would be septic and significantly unwell.

Answer D is wrong
The individual with Protein S deficiency produces thrombophilia and an increased risk of thromboembolic events. Most events are DVT, pulmonary embolism or cerebral thrombosis.

Answer E is the best answer
This young boy is at risk of developing Subacute bacterial endocarditis following his surgical repair and these lesions would be consistent with embolic thrombi from the endocarditis. The past history makes this the more likely explanation.

Answer F is wrong
There is no history of the characteristic butterfly rash or joint pain to make this a possible diagnosis.

Further Reading
Nelson Textbook of Pediatrics. 20th Edition. Kliegman RM et el. Elsevier. Philadelphia 2016. Chapter XX. The Cardiovascular System

Question 9 (7004 SP)
Subject: Haematology and Oncology
Answer: F - Urinary catecholamines

Reasoning:

Answer A is wrong

Sepsis must always be considered and may explain many of the features here. An intra-abdominal abscess could produce a mass, one would expect the child to be toxic.

Answer B is wrong

An infective cardiac lesion would not produce an abdominal mass and the cardiac murmur fits the description of a flow murmur.

Answer C is wrong

The FBC is likely to be abnormal but would not lead to a definitive diagnosis of the underlying aetiology.

Answer D is wrong

The liver function tests are likely to be abnormal but would not lead to a definitive diagnosis of the underlying aetiology.

Answer E is wrong

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.

Answer F is the best answer

The history and examination are highly suggestive of neuroblastoma. The pallor and the presence of left suprasternal lymph nodes (Virchow nodes) are indicative of metastatic disease. Raised urinary catecholamines are indicative of neuroblastoma although they can be negative in 5% of children.

Further Reading

Nelson Textbook of Pediatrics. 20th Edition. Kliegman RM et el. Elsevier. Philadelphia 2016. Chapter XXII Cancer and Benign Tumours

Question 10 (7009 SP)
Subject: Gastroenterology and Hepatology
Answer: B - Crohn's colitis

Reasoning:
Most diagnoses are made on the history. This girl is chronically unwell – she has pain, 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 likeliest diagnosis is Crohns colitis.

Further Reading
UpToDate: https://www.uptodate.com/contents/clinical-presentation-and-diagnosis-of-inflammatory-bowel-disease-in-children Authors: Highuchi L; Bousvaros A; [Accessed May 2018]

Question 11 a (D 0011 SP a)
Subject: Endocrinology and Growth
Answer: B - Diabetes insipidus

Reasoning:
This baby is vomiting, has faltering growth and deranged biochemistry.

Answer A is wrong
The urea is slightly high but not in the range for chronic renal failure.

Answer B is the best answer
Here the urine osmolality is low in the face of a high serum osmolality (easy to calculate yourself if you also have a glucose level but you can estimate it without) so diabetes insipidus is the correct answer.

Answer C is wrong
The abnormally high sodium excludes GORD.

Answer D is wrong
The biochemistry is normal in Hirschprung and the abdomen is likely to be distended, not scaphoid.

Answer E is wrong
In pseudohypoaldosteronism, whatever the type, there is increased renal tubular sodium loss resulting in hyponatraemia and hyperkalaemia.

Further Reading
https://www.uptodate.com/contents/hypernatremia-in-children

Question 11 b (D 0011 SP b)
Subject: Endocrinology and Growth
Answer: E - Water Deprivation Test

Reasoning:
Answer A is wrong
Diagnosis of diabetes insipidus is confirmed by the water deprivation test.

Answer B is wrong

Diagnosis of diabetes insipidus is confirmed by the water deprivation test.

Answer C is wrong

Diagnosis of diabetes insipidus is confirmed by the water deprivation test.

Answer D is wrong

Diagnosis of diabetes insipidus is confirmed by the water deprivation test.

Answer E is the best answer

To make a diagnosis of diabetes insipidus (DI) requires a water deprivation test with, if there is evidence of ongoing polyuria and weight loss, the assessment of the effect of a dose of desmopressin will help distinguish between central and nephrogenic DI. This test is dangerous in inexperienced hands and must only be undertaken by senior, trained nursing and medical staff.

Answer F is wrong

Diagnosis of diabetes insipidus is confirmed by the water deprivation test.

Further Reading

UpToDate: https://www.uptodate.com/contents/hypernatremia-in-children Authors: Somers MJ; Traum AZ. [Accessed May 2018]

Question 12 (D 1136 SP)
Subject: Haematology and Oncology
Answer: E - Microangiopathic intravascular haemolysis

Reasoning:
This is a normochromic, normocytic anaemia with thrombocytopenia and a slightly elevated white count. The blood film shows anisocytosis (red cells of different sizes) and burr cells (or echinocytes which show a crenelated membrane and are changes seen due to environmental causes).

Many of these answers can be excluded as the cause of the anaemia

Answer A is wrong
Acute lymphoblastic leukaemia (normal film, an appropriately elevated reticulocyte count indicating a responsive marrow and normal MCV –CHECK machine MCV of anisocytosis)

Answer B is wrong
Blood loss (nothing in the given history),

Answer C is wrong
HIV can give anaemia but you would expect the child to have a longer history of illness.

Answer D is wrong
Malaria would be identified on the film

Answer E is the best answer
Anaemia and thrombocytopaenia along with granular casts in urine indicate acute haemolysis - microangiopathic intravascular haemolysis.

Answer F is wrong
Sickle cell disease would be identified on the film.

Answer G is wrong
Thalassaemia would give a microcytic anaemia

Answer H is wrong
Tuberculosis would give a microcytic anaemia

Further Reading

Nelson Textbook of Pediatrics. 20th Edition. Kliegman RM et el. Elsevier. Philadelphia 2016. Chapter XXI. Diseases of the Blood

Question 13 (7005 SP)
Subject: Diabetes Mellitus
Answer: C - Cerebral oedema

Reasoning:
The 13 year old girl admitted in DKA and is hyperglycaemic and acidotic. Progresses into coma after 12hours of treatment as her blood glucose falls toward normal.

Answer A is wrong
Addison's disease is rare in children and young people but can lead to recurrent hypoglycaemia in patients with type 1 diabetes mellitus. There is insufficient information given to conclude that this diagnosis explains the change in clinical condition.

Answer B is wrong
Although acidotic there is nothing in history to suggest aspirin was administered. Aspirin is contraindicated in those under 16 years because of the increased risk of developing Reye's syndrome.

Answer C is the best answer
During resuscitation this child given fluids and there are rapid shifts of fluid between various body spaces during this time. Such fluid shifts can be rapid and lead to cerebral oedema, coning and death. Paediatric units would be expected to have clear and robust fluid replacement guidelines in place to ensure a slow and controlled rehydration.

Answer D is wrong
The glucose is returning to normal and one would not expect a clinical deterioration with a blood glucose of 10 mmol/l.

Answer E is wrong
Reye's syndrome - hepatic encephalopathy – would produce an encephalopathy often with confusion and seizures. No laboratory results are provided to suggest this diagnosis.

Further Reading
UpToDate: https://www.uptodate.com/contents/cerebral-edema-in-children-with-diabetic-ketoacidosis Author: Haymond MW; [Accessed March 2018]

Question 14 a (X 0015 SP a)
Subject: Nephro-urology
Answers: C - Dilated left renal pelvis

Reasoning:
It is unlikely that you will be asked to interpret a MAG3 scan in clinical practice but an understanding of the investigation helps in explaining the results.

The image on the left of the picture shows the uptake of the radioactive tracer by renal tissue and represents the two kidneys and bladder when viewed from the back of the patient. The right kidney shows increased concentration in the pelvic area (dense black) whilst the left kidney has extensive area of dense tracer presence – a dilated pelvis.

The image on the right plots out the uptake and decay of the contrast as it passes through the two kidneys. There is rapid uptake by both kidneys following the injection of the contrast with the right kidney absorbing slightly more than the left. The contrast tracer is then cleared by the kidney into the bladder and at 13 minutes furosemide (Lasix) is injected to enhance that clearance. The left kidney fails to clear the tracer and the injection of furosemide (Lasix) leads to a clearance of water and a consequent concentration of the tracer within the kidney – shown as a slight elevation in the line of the graph. The abnormality shown is in the left kidney.

Answer A is wrong
There is an asymmetrical response so any 'bilateral' answer is incorrect

Answer B is wrong
There is an asymmetrical response so any 'bilateral' answer is incorrect

Answer C is the best answer
The left kidney has increased density of contrast material and the renal pelvis is dilated.

Answer D is wrong
The abnormality is in the left kidney – the right is normal.

Answer E is wrong
Scarring would be shown by patches of failed uptake of tracer by the parenchyma.

Answer F is wrong
The abnormality is in the left kidney – the right is normal.

Answer G is wrong
The abnormality is in the left kidney – the right is normal. Scarring would be shown by patches of failed uptake of tracer by the parenchyma.

Answer H is wrong
Posterior urethral valves would give a symmetrical appearance and show a delay in excretion in both kidneys in the graph on the right.

Question 14 b (X 0015 SP b)
Subject: Nephro-urology
Answer: C - Delayed excretion on left despite furosemide

Reasoning:
Again any 'bilateral' answer is incorrect as is any answer including the right kidney. Uptake of tracer is good in both kidneys (indicated by the steep gradient of the graph). Ureteric reflux would show a delayed excretion of the tracer and a slower decay in the right hand graph.

Answer A is wrong
There is an asymmetrical response so any 'bilateral' answer is incorrect

Answer B is wrong
There is an asymmetrical response so any 'bilateral' answer is incorrect

Answer C is the best answer

Answer D is wrong
The abnormality is in the left kidney – the right is normal.

Answer E is wrong
Ureteric reflux would show a delayed excretion of the tracer and a slower decay in the right hand graph – but decay would occur.

Answer F is wrong
The left kidney shows a prompt uptake of the tracer – it is the excretion which is abnormal

Answer G is wrong
The right kidney shows a prompt uptake of the tracer – it is the excretion which is abnormal

Answer H is wrong

The abnormality is in the left kidney – the right is normal.

Answer I is wrong

Ureteric reflux would show a delayed excretion of the tracer and a slower decay in the right hand graph.

Further Reading

Nelson Textbook of Pediatrics. 20th Edition. Kliegman RM et el. Elsevier. Philadelphia 2016. Chapter XXIII. Nephrology

Question 15 (C 2129 SP)
Subject: Cardiology
Answer: B - Eisenmenger syndrome

Reasoning:
The image shows digital clubbing ie loss of the angle between the nail-bed and the nail. It is a much more reliable sign of chronic hypoxia than cyanosis. Indeed, in this image it is difficult to ascertain whether the child is cyanosed! Furthermore, peripheral cyanosis is most commonly associated with reduced blood flow through the extremities, rather than central cyanosis.
Having decided that the child has finger clubbing, the next step is to look for a condition in the list of possible answers that causes central cyanosis. The main causes of clubbing in children are: Suppurative lung disease (cystic fibrosis, bronchiectasis), cyanotic heart disease, infective endocarditis, inflammatory bowel disease.

Answer A is wrong
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 B is the best answer
Eisenmenger syndrome is caused by a ventricular septal defect leading to pulmonary hypertension and a right-to-left shunt causing central cyanosis and clubbing.

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

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

Further Reading
Nelson Textbook of Pediatrics. 20th Edition. Kliegman RM et el. Elsevier. Philadelphia 2016. Chapter XX. The Cardiovascular System

Question 16 (D 2330 SP)
Subject: Metabolism and Metabolic Medicine
Answers: B - Correct hypokalaemia

Reasoning:
The patient has a low BMI for her age as a result of malabsorption from her Crohn's disease. The significant abnormalities in the investigations are anaemia and hypokalaemia although ferritin, sodium and chloride are all borderline low. The question asks which 'is the most important action to take before commencing the enteral feeding regime?' Hypokalaemia needs prompt correction – the others can wait. Furthermore, were she to start an enteral feeding regimen whilst malnourished child she will be at risk of developing 'Re-feeding syndrome'. One of the consequences would be a further fall in potassium levels – it is therefore crucial that hypokalemia is corrected before enteral feeding is started.

Answer A is wrong
Returning the haemoglobin with oral iron in a well individual will take months and 93 g/l is not life-threatening

Answer B is the best answer
Prompt correction is required.

Answer C is wrong
Although B12 deficiency is possible, it has not been demonstrated on the results provided.

Answer D is wrong
Not an urgent requirement

Answer E is wrong.
A haemoglobin of 93 g/l is not life-threatening

Further Reading
Nelson Textbook of Pediatrics. 20th Edition. Kliegman RM et el. Elsevier. Philadelphia 2016. Chapter VIII. The Acutely Ill Child.

Question 17 (C 0022 SP)
Subject: Nephro-urology
Answer: E - Hereditary angio-oedema
Answer: G - Nephrotic syndrome

Reasoning:
The picture shows periorbital oedema without obvious inflammation or bruising. The most common causes of periorbital oedema in the UK are nephrotic syndrome and conditions presenting with allergy or anaphylaxis.

Answer A is wrong
Nephritis can lead to a nephrotic picture but proteinuria is uncommon (<1%).

Answer B is wrong
Cellulitis is unlikely as there is no obvious erythema in the periorbital area

Answer C is wrong
Cardiac failure can give systemic oedema but it is usually postural and one would need more clinical information about cardiac status.

Answer D is wrong
There is no history or clinical evidence of the characteristic rash.

Answer E the best answer
An acute anaphylactic reaction gives periorbital swelling.

Answer F is wrong
A spurious answer as myotonia may give a ptosis but this is oedema and not ptosis!

Answer G is the best answer
A nephrotic syndrome will lead to hypoabuminaemia

Answer H is wrong
No bruising – another spurious answer

Answer I is wrong
SVC obstruction gives full facial swelling, cyanotic facial appearance and engorged veins.

Further Reading

Up to Date: https://www.uptodate.com/contents/evaluation-and-management-of-edema-in-children Valentini RP; [Accessed May 2018]

Question 18 (SH 0030 SP)
Subject: Palliative Care and Pain Management
Answer: E - Oral ciprofloxacin and nebulised colomycin for 3 months

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Reasoning:
The current practice in the management of Pseudomonas colonisation in patients with cystic fibrosis involves the use of 'duel therapy' – antibiotics given systemically and via a nebulised route.

Answer A is wrong
Single agent antibiotic only.

Answer B is wrong
Single agent antibiotic only.

Answer C is wrong
This option does not include an antibiotic.

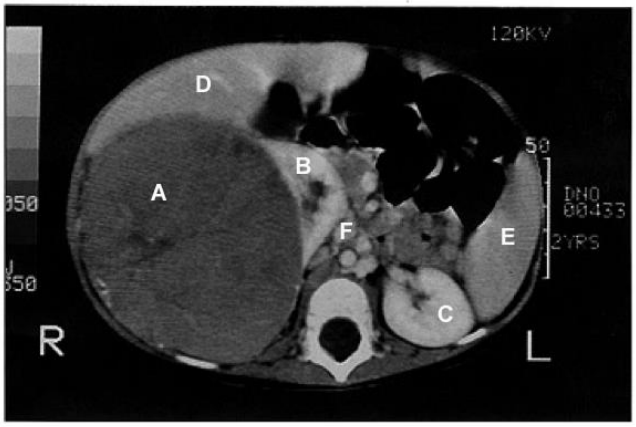
Answer D is wrong
This option does not include an antibiotic.

Answer E is the best answer
Duel therapy with oral and nebulised antibiotics.

Further Reading
Nelson Textbook of Pediatrics. 20th Edition. Kliegman RM et el. Elsevier. Philadelphia 2016. Chapter XIX The Respiratory System

Question 19 (X 0014 SP)
Subject: Haematology and Oncology
Answer: A - Displaced right kidney
Answer: F - Solid tumour of the right kidney

Reasoning:
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 sides.



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 the best answer

Answer B is wrong
The bowel gas pattern is of normal calibre although displaced slightly to the left.

Answer C is wrong
There are no cysts evident in the renal tissue (B or C).

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

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

Answer F is the best answer

Further Reading

Nelson Textbook of Pediatrics. 20th Edition. Kliegman RM et el. Elsevier. Philadelphia 2016. Chapter XXII Cancer and Benign Tumours

Question 20a (EMQ 0019 SP a)
Subject: Endocrinology and Growth
Answer: E - GP review at 6 months

Reasoning:
This is not an uncommon scenario. Most testes that are undescended at birth and complete their descent within the first three to four months of life. Spontaneous descent is rare after six months of age. Clinically this is an otherwise normal baby boy. The likelihood is that the palpable mass is a testis and the other one is either retractile or intra-abdominal. Provided the rest of the genitalia are normal there is no urgency to do anything but wait and see what happens - and reassure the parents.

Further Reading
UpToDate: https://www.uptodate.com/contents/undescended-testes-cryptorchidism-in-children-management Authors: Cooper CS; Docimo SG; [Accessed May 2018]

Question 20b (EMQ 0019 SP b)
Subject: Nephro-urology
Answer: J - Urea and electrolytes

Reasoning:
There are a number of reasons for this appearance, but the most dangerous is a virilised female baby with 21-hydroxylase deficiency. The urgent complications of this condition are hypoglycaemia and adrenal crisis, typically in the first two weeks of life. Thus, the NEXT step would be to check the electrolytes (and a blood sugar), before going on to perform more diagnostic tests.

Further Reading
UpToDate: https://www.uptodate.com/contents/diagnosis-of-classic-congenital-adrenal-hyperplasia-due-to-21-hydroxylase-deficiency-in-infants-and-children Author: Merke DP; [Accessed May 2018]

Question 20c (EMQ 0019 SP c)
Subject: Nephro-urology
Answer: H - Referral to paediatric surgeon

Reasoning:
If the testes are in the scrotum, this is likely to be an inguinal hernia, and most of the other options are irrelevant. Since it is tender, it may be incarcerated or strangulated, and thus surgical referral is indicated.

Further Reading
UptoDate https://www.uptodate.com/contents/inguinal-hernia-in-children Author: Ramsook C; [Accessed May 2018]

Question 21 (7006 SP)
Subject: Neurology
Answer: C - Febrile convulsion

Reasoning:

Answer A is wrong

Although adrenal insufficiency (Addison disease) can cause electrolyte disturbances which may lead to seizure activity, the results are not consistent with such a diagnosis (hypoglycaemia, hyponatraemia, hyperkalaemia).

Answer B is wrong

He has only had one episode so this is not any form of epilepsy. This episode does not fit the pattern of BFEC – recurrent, infrequent, often at night.

Answer C is the best answer

The child has an intercurrent illness and is mildly pyrexial when assessed though may have had a higher temperature around the time of the seizure. He is of the expected age for febrile seizure and has no abnormal neurological findings.

Answer D is wrong

There are no features in the history or blood results to support this diagnosis

Answer E is wrong

The clinical history is of an acute onset. The blood results are not consistent with this diagnosis.

Answer F is wrong

Children with MCAD do present during a period of fasting and poor oral intake and can have seizures. However, they have hypoglycaemia.

Further Reading

UpToDate: https://www.uptodate.com/contents/seizures-and-epilepsy-in-children-classification-etiology-and-clinical-features Author: Wilfong A. [Accessed March 2018]

Question 22 (DE 252 SP)
Subject: Cardiology
Answer: D - 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 180^{o} as shown by an equiphasic QRS compex in aVF with an upward QRS complex in aVR. There is also right ventricular hypertrophy with tall R waves over the right chest leads (note that V1, V2 and V3 have been recorded at half the normal standardisation).

Answer A is wrong
Ostium primum atrial septum defect is also a type of acyanotic congenital heart disease, but the ECG is typically associated with right bundle branch block and right axis deviation (without right ventricular hypertrophy) NB PRIMUM V SECUNDUM

Answer B is wrong.
In Fallot tetralogy the child would be cyanosed. The ECG shows right axis deviation and right ventricular hypertrophy. It would be extremely unlikely to see a 5 year old child with uncorrected tetralogy of Fallot in this country.

Answer C is wrong
In uncomplicated patent arterial duct, the child is usually pink and well, and there are no diagnostic ECG features.

Answer D is the best answer
Of the types of congenital heart disease listed, the only type of acyanotic heart disease associated with right axis deviation and right ventricular hypertrophy is pulmonary stenosis.

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

Further Reading
Nelson Textbook of Pediatrics. 20th Edition. Kliegman RM et el. Elsevier. Philadelphia 2016. Chapter XX. The Cardiovascular System

Question 23 (7008 SP)
Subject: Cardiology
Answer: F - Long QT syndrome

Reasoning:

Answer A is wrong

The cardiac examination was normal!

Answer B is wrong

The episodes do not occur at night

Answer C is wrong

The history does not describe seizures with abnormal, coordinated movements

Answer D is wrong

Hypoglycaemia may cause seizures and loss of consciousness, but the individual would experience symptoms such as sweating, anxiety, weakness and headaches before the loss of consciousness and is usually preceded by increasing drowsiness.

Answer E is wrong

The episodes are described as 'collapse' and not 'jerking'

Answer F 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 will show a prolonged QTc of greater than 0.46 msec.

Further Reading

Nelson Textbook of Pediatrics. 20th Edition. Kliegman RM et el. Elsevier. Philadelphia 2016. Chapter XX. The Cardiovascular System

Question 24 (7007 SP)
Subject: Gastroenterology and Hepatology
Answer: D - Juvenile colonic polyp

Reasoning:
This boy is well and the bleeding is painless. He has a microcytic anaemia suggestive of iron deficiency so the bleeding is significant and over a prolonged period of time.

Answer A is wrong
Painless bleeding would make anal fissure very unlikely.

Answer B is wrong.
This is rather a prolonged duration of illness and the fresh blood indicates a bleeding point rather than widespread mucosal damage and inflammation.

Answer C is wrong
The normal albumin and CRP help exclude inflammatory bowel disease.

Answer D is the best answer
The presence of mucous and significant fresh bleeding makes the likeliest diagnosis a juvenile colonic polyp.

Answer E is wrong
This diagnosis is possible but most patients with complications from a Meckel's
are younger and the blood rarely fresh

Further Reading
UpToDate: https://www.uptodate.com/contents/lower-gastrointestinal-bleeding-in-children-causes-and-diagnostic-approach Authors: Patel N; Kay M; [Accessed May 2018]