Applied Knowledge in Practice – Sample Paper 1
Answer Key

Question 1 (C 1691 SP)
Subject: Dermatology
Answer: B - Eczema herpeticum

Reasoning:
The child has widespread vesiculo-pustular lesions involving the face and upper trunk which is typical of eczema herpeticum – atopic dermatitis infected with herpes simplex. The history of a mild skin rash since the age of two months suggests underlying chronic condition - eczema.

Answer A is wrong
The rash of chicken pox evolves from pruritic, erythematous macules to papules and then vesicles. The rash occurs in crops, so it is characteristic to find lesions at various stages. There is usually relative sparing of the face in chickenpox. Varicella in an immune-compromised child may cause this appearance, but it is much less likely than eczema herpeticum, particularly with the history that he had been previously been well.

Answer B is the best answer

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

Answer D is wrong
Primary herpes simplex infection presents with gingivo-stomatitis. Vesicles develop in the mouth and on the tongue and lips, but would not involve the rest of the face and the upper trunk.

Answer E is wrong
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 that may include the eyes, oral cavity, upper airway and G-I tract.

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

Question 2 (X 0020 SP)
Subject: Respiratory Medicine with ENT
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. This child is basically well with a cough which will exclude those answers leading to significant lung compromise.

Answer A is wrong
There are no bowel gas patterns within either lung field.

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

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

Question 3a (EMQ 0007 SP a)
Subject: Respiratory Medicine with ENT
Answer: A - Bronchiectasis

Reasoning:
The child has a persistent cough and focal signs after a lower respiratory infection.

Acute infections such as chlamydia and pertussis might have caused the original illness, but not the sequelae.

There is no history given to suggest TOF, aspiration, gastro-oesophageal reflux, hyper IgE or tuberculosis. Obliterative bronchiolitis is a potential consequence of LRTI and can be associated with bronchiectasis but would normally have associated wheeze with fixed lower airway obstruction.

PCD is a plausible cause of chronic chest symptoms and signs, but would normally have presented with earlier onset of symptoms, and associated ear and nose problems, rather than a single LRTI.

Thus, bronchiectasis is the best explanation of these symptoms and signs.

Further Reading
Up to Date: https://www.uptodate.com/contents/epidemiology-clinical-presentation-and-evaluation-of-parapneumonic-effusion-and-empyema-in-children Fakhoury K; Kanu A [Accessed May 2018]

Question 3b (EMQ 0007 SP b)
Subject: Respiratory Medicine with ENT
Answer: I - Recurrent aspiration

Reasoning:
Recurrent cough and wheeze indicates a chronic condition.

It is likely that a child who suffered from severe birth asphyxia has neurodevelopmental consequences, of which the commonest is cerebral palsy. The likeliest cause of respiratory symptoms in a child with cerebral palsy is aspiration, either from gastro-oesophageal reflux or pharyngeal incoordination.

Thus, recurrent aspiration is the most accurate answer to the question.

Further Reading

Up to Date: https://www.uptodate.com/contents/management-and-prognosis-of-cerebral-palsy. Author: Patterson MC; [Accessed May 2018]

Question 3c (EMQ 0007 SP c)
Subject: Respiratory Medicine with ENT
Answer: D - Hyper IgE syndrome

Reasoning:

The key features in this case are the recurrent skin sepsis and recurrent cough and sputum, colonised with Staphylococci aureus. Staphylococci are unusual pathogens in sputum, and two conditions are suggested by this - cystic fibrosis and Hyper-IgE syndrome.

The former is not in the list, and the latter is associated with skin sepsis because of the specific immune deficiency.

Further Reading

Up to Date https://www.uptodate.com/contents/autosomal-dominant-hyperimmunoglobulin-e-syndrome Authors: LaPine A; Kumánovics TR; and Hill HR; [Accessed May 2018]

Question 4 (EH 39 SP)
Subject: Musculoskeletal
Answer: A - Anti-double stranded DNA

Reasoning:
The photograph shows a young person with a facial rash across both malar regions and the bridge of the nose – a 'butterfly distribution'. The history suggests sensitivity to sunlight, chest and joint symptoms along with episodic pyrexia. A likely chronic anaemia and raised CRP are seen. All these observations suggest a diagnosis of Systemic Lupus Erythematosus (SLE) - in this case exacerbated by UV light (excess sunlight) exposure.

Answer A is the best answer
A positive result for anti-double stranded DNA will confirm the diagnosis of SLE

Answer B is wrong
Although temperatures, chest signs and arthropathy can be seen in Mycoplasma infection, the rash is not seen.

Answer C is wrong
Parvovirus gives the 'slapped cheek' facial rash but the illness is usually short lived. It has been associated with arthropathy but this is not a common feature.

Answer D is wrong
Systemic symptoms are seen in rheumatoid disease along with similar abnormalities shown n the blood results but the rash is more characteristic of SLE.

Answer E is wrong
The symmetrical distribution of the rash is not characteristic of a superficial infection.

Answer F is wrong
Porphyria can lead to a photosensitivity of the skin but would not produce the marked arthropathy described here

Further Reading
Nelson Textbook of Pediatrics. 20th Edition. Kliegman RM et el. Elsevier. Philadelphia 2016. Chapter XVI Rheumatic Diseases of Childhood.

Question 5 (SH 2278 SP)
Subject: Neonatology
Answer: B - Blood sugar
Answer: C - Calcium

Reasoning:
All of these answers are plausible and indeed all of them might be requested. But the question asks which two would be most helpful in his *immediate* management.
Hypoglycaemic seizures in infancy are associated with poor neurological outcome so it is crucial to check the glucose as the infant arrives and correct it as necessary.
Similarly, hypocalcaemia is easily and quickly diagnosed and must be corrected. 'Treat the Treatable is a good adage.

Answer A is wrong
Although the history may be consistent with a urea cycle defect, his mental state does not suggest this, and the condition would not be addressed immediately at presentation

Answer B is the best answer
Crucial investigation that allows immediate correction

Answer C is the best answer
Crucial investigation that allows immediate correction

Answer D is wrong
There is no history of a bleeding disorder

Answer E is wrong
May be an appropriate investigation for the presentation but request, implementation and interpretation would not be immediate.

Answer F is wrong
May be an appropriate investigation for the presentation but request, implementation and interpretation would not be immediate.

Answer G is wrong
Anaemia and thrombocytopaenia may be causes for the presentation but there is no history of pallor or bruising.

Answer H is wrong

Abnormalities of sodium, potassium, creatinine and urea would be important to know and abnormal sodium could lead to seizure activity but are less likely than hypocalcaemia in this child.

Further Reading

UpToDate https://www.uptodate.com/contents/etiology-and-prognosis-of-neonatal-seizures
Author: Shellhass R. [Accessed May 2018]

Question 6 (C 2064 SP)
Subject: Gastroenterology and Hepatology
Answer: D - Meckel's diverticulum

Reasoning:
This is an unusual scenario but the history is highly suggestive of pathology.

Blood mixed with the stools suggests lower gastro-intestinal bleeding and merits investigation. This is a radio-isotope scan that is used to make a diagnosis of a Meckel's diverticulum – the injected material is taken up by the mucosal cells found in stomach (or elsewhere). The image shown is of the abdominal area of the child and reveals the expected uptake of the stomach mucosa (dense area at the top right), the contrast in the bladder (at the bottom middle of the picture) and the ectopic mucosa of the Meckel's diverticulum (in the centre of the scan).

Answer A is wrong
The child with an appendix abscess would have a short history and would likely be toxic. The 99mTc pertechnetate scan would not be used in such a situation.

Answer B is wrong
The child with an intussusception would have a short history and would likely be acutely unwell. The 99mTc pertechnetate scan would not be used in such a situation.

Answer C is wrong
The child with a malrotation would have a short history and would likely be acutely unwell. The 99mTc pertechnetate scan would not be used in such a situation.

Answer D is the best answer

Answer E is wrong
The history is not that of vesico-ureteric reflux and a MAG3 scan would be the more appropriate investigation.

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]

Question 7 (C 0005 SP)
Subject: Dermatology
Answer: E - Intraocular pressure measurement

Reasoning:
This is a two-stage question. The first stage is to make a diagnosis. The 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.

Answer A is wrong
Sturge-Weber syndrome is not genetically determined and therefore chromosome analysis is irrelevant.

Answer B is wrong
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 wrong
Cranial ultrasound will not show a vascular abnormality and is not indicated

Answer D is wrong
Although around 80% of children with Sturge-Weber Syndrome develop epilepsy an EEG is not indicated until the child presents with a seizure.

Answer E is the best answer
Glaucoma affecting the ipsilateral eye is a common complication in infancy and appropriate assessment is mandatory

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

Question 8 (D 0003 SP)
Subject: Infection, Immunology and Allergy
Answer: C - HIV/AIDs

Reasoning:
This child has recently arrived in the UK and so past medical history is not known. The child may come from an area where some conditions are more prevalent. This child has poor feeding, loose stools and faltering growth. The child is also anaemic and has raised immunoglobulins.

Answer A is wrong
The child with chronic granulomatous disease has neutrophil dysfunction and therefore usually presents with recurrent and multiple abscesses

Answer B is wrong
Common Variable Immune Deficiency is characterised by hypogammaglobinaemia and that is not demonstrated by the immunoglobulin results.

Answer C is the best answer
HIV / AIDS presents with failure to thrive, chronic diarrhoea, chronic parotid swelling, lymphocytic interstitial pneumonitis (LIP) and oral thrush.

Answer D is wrong
In Severe Combined Immune deficiency, the lymphocytes will be low – often less than 0.5x109/l - and immunoglobulin levels are reduced.

Answer E is wrong
In X-linked hypogammaglobinaemia the immunoglobulins will be low

Further Reading
Nelson Textbook of Pediatrics. 20th Edition. Kliegman RM et el. Elsevier. Philadelphia 2016. Chapter XVII Infectious Diseases

Question 9 (C 0007 SP)
Subject: Dermatology
Answer: D - Staphylococcal scalded skin syndrome

Reasoning:
The child has diffuse erythema of the face with peeling of the skin periorally, 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 and pemphigus.

Answer A is wrong
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 wrong
Herpes simplex at this age is usually a primary infection that presents with gingivo-stomatitis. Vesicles develop in the mouth and on the tongue and lips.

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

Answer D is the best answer

Answer E is wrong
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.

Further Reading
Nelson Textbook of Pediatrics. 20th Edition. Kliegman RM et el. Elsevier. Philadelphia 2016. Chapter XVII Infectious Diseases

Question 10 (X 0011 SP)
Subject: Neonatology
Answer: B - Fluid resuscitation with 10 ml/kg 0.9% saline

Reasoning:
The clinical story is that of a preterm infant who is in circulatory shock -tachycardia, hypotension and peripheral oedema. His blood gas shows a profound metabolic acidosis. Abdominal x-ray shows dilated loops of bowel, thickening of the bowel wall and free air at the very top of the film – features of NEC and perforation. Resuscitation is required with urgency. The question asks for the 'most appropriate immediate management'; although more than one action may be necessary in clinical management; the question is asking you to identify the response with the greatest urgency.

Answer A is wrong
Although the baby has a metabolic acidosis, the cause of this is primarily due to circulatory shock. Bicarbonate would not address the cause of the metabolic problem.

Answer B is the best answer

Answer C is wrong
Increasing ventilatory pressures and rates would drop the CO2 but this is currently within the normal range and the action would again not address the causative problem.

Answer D is wrong
No place in the described situation.

Answer E is wrong
These would be needed promptly but they would not be the 'most appropriate immediate management'

Further Reading
Nelson Textbook of Pediatrics. 20th Edition. Kliegman RM et el. Elsevier. Philadelphia 2016. Chapter XII. The Fetus and Neonatal Infant

Question 11 (7000 SP)
Subject: Neurology
Answer: G - Reflex anoxic seizures

Reasoning:

Answer A is wrong

The episodes do not occur at night

Answer B is wrong

The history does not describe seizures with abnormal, coordinated actions

Answer C is wrong

There is nothing in the given history that raises this as a diagnosis and probably there were independent witnesses to the event at the nursery.

Answer D is wrong

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

Answer E is wrong

No record of glucose results so makes this explanation unlikely.

Answer F is wrong

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

Answer G 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

http://www.childneurologyfoundation.org/disorders/breath-holding-spells/ [Accessed May 2018]

Question 12 a (C 0011 SP a)
Subject: Safeguarding
Answer: E - Inadequate calorie intake

Reasoning:
The growth chart shows you that the head circumference follows the 50th centile and the most recent measurement of length is also on the 50th. The weight of the child follows the 50th centile until 20 weeks then falls to the 0.4 centile. The child is admitted to hospital and the weight moves up to the 9th centile. The stem to the question does not indicate that any specific intervention or treatment given – therefore assume that no active treatment was provided. It is clear that the hospital admission confirms good weight gain can be achieved when adequate calorific intake is assured.

Answer A is wrong
Although the start of poor weight gain coincided with the introduction of solids, a child with coeliac disease would not usually present with vomiting and any weight gain would have been the result of a move to a gluten-free diet (not mentioned). The weight gain would not have been so dramatic.

Answer B is wrong
A child with CMPI would usually have diarrhoea along with the vomiting and a change in diet would have been mentioned in the text.

Answer C is wrong
If the child had cystic fibrosis then the observed increase in weight would have been the result of an intervention – and this would have been mentioned in the text.

Answer D is wrong
If the child had GORD then the observed increase in weight would have been the result of an intervention – and this would have been mentioned in the text.

Answer E is the best answer
This child has therefore not been given sufficient calories for weight gain and neglect is the most likely cause.

Further Reading
Clinical Cases for MRCPCH Applied Knowledge in Practice RCPCH 2016. Ed R Dinwiddie.

Question 12b (C 0011 SP b)
Subject: Safeguarding
Answer: A - Discharge planning meeting

Reasoning:
The most appropriate answer to this second part of the question depends upon recognising the appropriate diagnosis from part 1.

Answer A is the best answer
The chart shows you that the most likely explanation is neglect with inadequate calories provided. A full assessment by all professionals involved – hospital staff, community team and social workers is needed to assess the potential risk to the child if discharged. The first concern has to be the safety of the child.

Answer B is wrong
Inappropriate investigation where safeguarding is the recognise cause of the problem.

Answer C is wrong
Inappropriate investigation where safeguarding is the recognise cause of the problem.

Answer D is wrong
Inappropriate investigation where safeguarding is the recognise cause of the problem.

Answer E is wrong
Inappropriate investigation where safeguarding is the recognise cause of the problem.

Further Reading
Clinical Cases for MRCPCH Applied Knowledge in Practice RCPCH 2016. Ed R Dinwiddie.

Question 13a (EMQ 0003 SP a)
Subject: Neurology
Answer: B - Lamotrigine

Reasoning:
First line treatment of Juvenile absence epilepsy should be with one of the following - ethosuximide, sodium valproate or lamotrigene. A girl of this age - who may become pregnant - should not be offered sodium valproate due to its teratogenic effect. Ethosuxamide is not on the list of options and so lamotrigine is the most appropriate treatment.

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

Question 13b (EMQ 0003 SP b)
Subject: Neurology
Answer: I - Vigabatrin

Reasoning:
The first line option in infantile spasms would either be ACTH or vigabatrin. ACTH is not an option offered here,

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

Question 13c (EMQ 0003 SP c)
Subject: Neurology
Answer: D - No treatment

Reasoning:

The patient has benign childhood epilepsy with centrotemporal spikes.

The option of not treating the child should be considered particularly if not impacting on lifestyle. The seizures are often at night and are infrequent in nature. Status epilepticus is very uncommon. The prognosis is very good and seizures will resolve in most children often by the early teenage years.

Further Reading

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

Question 14 (EBM 0013 SP)
Subject: Science of Practice
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 wrong
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 wrong
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 wrong
A systematic review identifies those publish 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 wrong
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:F6–F10

Question 15 (SH 0004 SP)
Subject: Cardiology
Answer: D - Transposition of the great arteries with VSD

Reasoning:

Answer A is wrong

Aortic stenosis could produce the symptoms recorded but would not produce the observed central cyanosis. Improvement in the oxygenation with the prostaglandin can be seen and this would not occur in aortic stenosis.

Answer B is wrong

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 wrong

Babies with pulmonary atresia become acutely unwell as the duct closes within the first few hours. Prostaglandin E_1 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 or 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 wrong

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

Question 16 (SH 0016 SP)
Subject: Neonatology
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 wrong
Symptoms from drug withdrawal are not confined to sleep and would not be increasing in frequency at this age.

Answer C is wrong
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 wrong
Symptoms from infantile spasms are not confined to sleep.

Answer E is wrong
Symptoms from myoclonic epilepsy are not confined to sleep.

Further Reading
UpToDate https://www.uptodate.com/contents/nonepileptic-paroxysmal-disorders-in-infancy Authors: Nguyen TT; Kaplan PW; Wilfong A; [Accessed May 2018]

Question 17 (D 0006 SP)
Subject: Neonatology
Answer: A - Add inhaled nitric oxide at 20 ppm

Reasoning:
This baby has persistent pulmonary hypertension of the newborn (PPHN) as indicated by the normal CO2 and low oxygenation. This is often seen in term babies with meconium aspiration. Pulmonary vasodilatation or ECMO may be necessary.

Answer A is the best answer
The addition of inhaled nitrous oxide would lead to pulmonary vasodilation and thereby improve arterial oxygenation.

Answer B is wrong
Blood gas bicarbonate level is at the lower limit of normal and does not need correction at this stage.

Answer C is wrong
This would drop the pCO2 further and would not be appropriate

Answer D is wrong
The mean arterial blood pressure is currently at an acceptable level and so does not need immediate support.

Answer E is wrong
High frequency oscillation is a reasonable option and is used in the management of PPHN but is usually considered when PIP reach 28-30. The evidence of efficacy of HFOV in PPHN remains limited.

Further Reading
UpToDate https://www.uptodate.com/contents/persistent-pulmonary-hypertension-of-the-newborn Authors: Stark A; Eichenwald E. [Accessed May 2018]

Question 18 (D 1121 SP)
Subject: Neonatology
Answer: E - Passive cooling

Reasoning:

Answer A is wrong
This intervention is not evidence-based

Answer B is wrong
This intervention is not evidence-based

Answer C is wrong
This intervention is not evidence-based

Answer D is wrong
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

Annzopardi 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]

Question 19 (SH 0008 SP)
Subject: Respiratory Medicine with ENT
Answer: E - Short synacthen test
Answer: F - Spirometry with reversibility

Reasoning:
The most common cause of poorly controlled asthma is poor adherence to therapy, especially in a teenager. Other diagnoses also need to be excluded.

Answer A is wrong
This test would be inappropriate until more basic assessments had been carried out.

Answer B is wrong
Nothing in the history suggests a diagnosis of tuberculosis

Answer C is the wrong
Ciliary dyskinesia may be considered at a later stage.

Answer D is the wrong
These may be considered at a later stage in the management.

Answer E is the best answer
If he is taking treatment as prescribed (high doses), then he is likely to have a degree of adrenal suppression, which could be dangerous.

Answer F is the best answer
A more detailed spirometric assessment would give more information than the crude measure of PEF. If obstruction is confirmed on the flow-volume loop, and reversibility is demonstrated then the diagnosis of asthma is strengthened, and the likelihood of non-adherence increases.

Answer G is wrong
CT chest is not generally felt to be worth the radiation exposure.

Answer H is wrong
Cystic fibrosis should be considered but sweat test would not be the next investigation

Answer I is wrong
Nothing in the history suggests pulmonary emboli.

Further Reading

International ERS/ATS guidelines on definition, evaluation and treatment of severe asthma.. Chung et al. European Respiratory Journal 2013; DOI: 10.1183/09031936.00202013. [Accessed May 2018]

Question 20 (SH 0013 SP)
Subject: Neurology
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 wrong
Although Idiopathic (benign) intracranial hypertension does lead to chronic headaches of a persistent nature, they are not associated with abnormal neurological findings.

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

Answer D is wrong 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 wrong
A spinal cord tumour would not give a history of headaches.

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

Question 21 (X 3071)
Subject: Haemotology and Oncology
Answer: D - Hilar lymphadenopathy
Answer: F - Left pleural effusion

Reasoning:
This chest x-ray is grossly abnormal and shows dense shadowing in the mediastinum and the left side of the chest.

The large left pleural effusion and gross bilateral hilar lymphadenopathy are likely the result of some form of non-Hodgkin lymphoma or leukaemia and those which arise in the chest are usually T-cell in origin (from thymic (T) modulated cells).

Answer A is wrong
Cardiac shadow is visible behind the shadowing and is normal

Answer B is wrong
Cardiac shadow is visible and is normal

Answer C is wrong
No evidence of fluid in the right horizontal fissure.

Answer D is the best answer
Appearances are characteristic of bilateral hilar lymphadenopathy. This is less clear on the left because of the dense shadowing (fluid) occupying the left hemithorax.

Answer E is wrong
No evidence of a left Gohn focus.

Answer F is the best answer
There is a fluid level extending up the lateral chest wall consistent with a large left pleural effusion and this causes some midline shift to the right.

Answer G is wrong
There is some tracheal shift to the right but no air in the pleural cavity on the left.

Answer H is wrong
The upper part of the central shadow is consistent with thymic enlargement but the mass is much larger than could be attributed to just thymus (persistent thymic shadow)

Answer I is wrong

The shape of the cardiac shadow is not consistent with this diagnosis.

Further Reading

UpToDate https://www.uptodate.com/contents/epidemiology-clinical-presentation-and-evaluation-of-parapneumonic-effusion-and-empyema-in-children Authors: Janahi I ; Fakhoury K [Accessed May 2018]

UpToDate https://www.uptodate.com/contents/clinical-assessment-of-the-child-with-suspected-cancer Authors: Neville K; Steuber CP [Accessed May 2018]

Question 22 (7001 SP)
Subject: Gastroenterology and Hepatology
Answer: E - 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.

The palpable mass and the rectal bleeding makes intussusception most likely.

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.

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]

https://www.uptodate.com/contents/intussusception-in-children Authors: Vo N; Sato TT [Accessed May 2018]

Question 23 (SH 0014 SP)
Subject: Diabetes Mellitus
Answer: D - Nephropathy
Answer: F - Retinopathy

Reasoning:

Answer A is wrong
Would not form part of an annual assessment, although offered at diagnosis..

Answer B is wrong
Would not form part of an annual assessment.

Answer C is wrong
Would not form part of an annual assessment.

Answer D is the best answer
Vasculopathy is a significant issue and will affect renal tissue. Annual check for albuminuria is vital.

Answer E is wrong
Peripheral neuropathy is a recognised complication of type 1 diabetes but would not be common at this age.

Answer F is the best answer
Vasculopathy leads to a diabetic retinopathy and is more likely with duration of diagnosis and degree of glycaemic control.

Further Reading
UpToDate https://www.uptodate.com/contents/complications-and-screening-in-children-and-adolescents-with-type-1-diabetes-mellitus Authors: Levitsky, L; Misra, M; [Accessed May 2018]

Question 24 (X 0002 SP)
Subject: Metabolism and Metabolic Medicine
Answer: E - Rickets

Reasoning:
The X-ray of the wrist shows poor mineralisation associated with cupping and fraying of the metaphyseal region.

Answer A is wrong
Lead poisoning gives dense lines along the metaphyseal margin (look for a picture)

Answer B is wrong
Mucopolysaccharidosis is a collection of conditions caused by enzyme deficiencies leading to abnormal storage of glycosamines in connective tissue. The radiological changes are extensive and include short, thick phalanges with proximal widening.

Answer C is wrong
The radiological appearances of osteogenesis imperfect are osteopenia and fractures.

Answer D is wrong
The bones are dense white in osteopetrosis.

Answer E is the best answer
This is classical of Vitamin D deficiency rickets (poor mineralisation associated with cupping and fraying), which occurs a baby who is largely breast fed, fussy eater and who is failing to thrive.

Further Reading
https://www.ncbi.nlm.nih.gov/pmc/articles/PMC3731470/ [Accessed May 2018]

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

Question 25 (7002 SP)
Subject: Haematology and Oncology
Answer: D - Pneumocystis jirovecii pneumonitis

Reasoning:

Answer A is wrong

Bleomycin does cause a fibrosis and could present with some of the features listed here but not so acutely or so profoundly and the patient would not be pyrexial.

Answer B is wrong

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

Answer C is wrong

Pneumococcal pneumonia that was the cause of such marked hypoxia would produce a toxic presentation in the patient with much higher temperatures

Answer D is the best answer

Pneumocystis jirovecii pneumonitis is seen in patients who are immunosuppressed and they will often have marked hypoxia despite a paucity of chest findings on auscultation.

Answer E is wrong

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 F is wrong

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

Further Reading

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