Essential Paediatric Revision

Question 1

Which of the following is part of the routine assessment of a child following a first febrile convulsion? (single best answer)

1. CT brain
2. EEG
3. Developmental assessment
4. Lumbar puncture
5. MRI brain

Question 2

Case history
Kaya was born at 25 weeks gestation and is now 29 weeks corrected gestational age. She is not tolerating feeds and for several hours has had bilious aspirates and bloody stools.

Observations
HR 160 bpm, RR 60 bpm, Frequent desaturations to 80%, CRT 5 secs

Examination
Irritable when handled; Cardiovascular and respiratory system normal
Abdomen distended and shiny with visible bowel loops

Single best answer
What is the most likely diagnosis?

A Pyloric stenosis
B Gastro-oesophageal reflux
C Hirschprung disease
D Necrotizing enterocolitis
E Oesophageal atresia
Question 3: short notes question

A twenty month old boy can crawl but is not yet able to walk.

A Name three diagnostic possibilities.

B What three points would you want to ask in the history?

C What would you expect social milestones to be at this age?

D How would you assess vision in a child of this age?

E Physical examination shows arms are normal but legs are hypertonic and scissor when he is picked up. What is the underlying diagnosis?

F What is the specific name given to this pattern of disability?
Questions 4
A three year old girl whose parents are originally from Japan has had a prolonged fever for more than a week. In the last few days she has developed a rash on her hands which is starting to peel and her lips have become dry and cracked. Mum reports that her eyes have been very red with a watery discharge particularly when her temperature is high.

What is the likely diagnosis?

What serious complication may occur over the next few weeks?

What is the treatment?

Question 5
What is the diagnosis? Write short notes on this condition.
Question 6

Jamal is a 9 year old boy who has sickle cell disease. He presents to A&E with acute severe pain in his left leg and yellow sclera.

List three different types of sickle cell ‘crises’ that occur in children?

How would you manage Jamal?
Question 7
Male infant James Smith was born at 38 weeks gestation by spontaneous vaginal delivery following prolonged rupture of membranes. Mum is a primigravida, with pregnancy induced hypertension, rhesus negative with a normal haemoglobin, and antenatal testing was positive for vaginal group B strep.

James is now 12 hours old with poor feeding and felt by staff to be “not right”

Observations
Temp 35.4
HR 190bpm
RR 60
Sats 95% in air

On examination
Cold, pale, mottled
HS I II no murmur, femorals palpable; CRT 4 secs
Intermittent grunting but no recessions and chest clear
Abdomen normal

What is the most likely diagnosis? Single best answer
a) Cyanotic congenital heart disease
b) Group B streptococcal sepsis
c) Rhesus incompatibility
d) Neonatal stroke
e) Neonatal hypoglycaemia

Question 8
16 month old Petr has been previously well with no significant medical history. He was at his grandparents’ house, holding on to a table. Suddenly his eyes rolled upwards and he fell backwards onto a carpeted floor. He went very stiff and started shaking and this continues after he has been brought in by ambulance 15 minutes after the onset. The ambulance crew have given 5mg of rectal diazepam.

After checking ABC and his blood glucose and giving oxygen, what further treatment would you arrange to terminate his seizure? (Single best answer)
a) Intravenous lorazepam
b) Buccal midazolam
c) Repeat rectal diazepam
d) Intravenous infusion of phenytoin
e) Fast bleep the anaesthetist for rapid sequence induction with thiopentone
Answers

Question 1: Answer: 3: Developmental assessment
This is important because developmental delay is one of the risk factors for developing epilepsy and so this is a routine part of assessing a child with a febrile convulsion. Other risk factors for epilepsy include a family history of epilepsy or presentation with a complex seizure: eg a seizure lasting more than 15 minutes or recurring several times in first 24 hours or with focal features.

An EEG would not be performed as it would be likely to be normal following a febrile convulsion and in any case would not alter management.

Brain imaging would mainly be relevant if there were focal features or significant developmental delay or abnormal neurological examination findings.

Note that only 1% of those presenting with convulsions have meningitis as the underlying cause. If there were a clinical suspicion of meningitis being the underlying cause, imaging to exclude raised intracranial pressure might be performed prior to lumbar puncture. For example, if a non-blanching rash had been detected, immediate treatment with IV antibiotics would have been started prior to CT scan and subsequent LP (See NICE guideline on meningitis)

Notes on febrile convulsions
• Commonest seizure disorder of childhood
• Typically from 6 months – 5 years of age
• A seizure associated with a fever
• Seizures are typically generalised and short in duration
• Fever may be due to UTI, URTI or other infection

Management of febrile seizures
• Always remember ABC (+ DEFG- don’t ever forget glucose)
• Symptomatic relief of high fever with anti-pyretics
• Find a focus for the fever, and treat with antibiotics if indicated
• Frightening to witness- it is normal for parents to think their child is dying
• Educate the parents about prognosis and outcomes in febrile seizures

Prognosis
• Febrile convulsions are not epilepsy
• The risk of recurrent febrile seizures during another febrile illness is approximately 30%
• If a child is developmentally normal, has no family history of epilepsy and presents with a simple febrile seizure, the risk of developing epilepsy is the same as the population risk
• The risk of epilepsy is significantly increased in those with febrile convulsions associated with neurological developmental abnormalities, a family history of epilepsy or presenting with complex febrile seizures (eg lasting longer than fifteen minutes, with focal features or recurring within the same illness)
Question 2

Answer: D Necrotising enterocolitis

Kaya is premature and has developed typical symptoms and signs of necrotising enterocolitis, which is almost always found as a complication of prematurity. She is tube fed and the bilious aspirate suggests a serious intra-abdominal cause such as NEC for her irritability. The bloody stool is also a typical finding.

This clinical picture would not be consistent with pyloric stenosis or GORD where white or yellow vomit/aspirate would be found and where bloody stools are not a feature. Similarly, bilious vomit would not be found in oesophageal atresia.

Hirschprung disease is commoner in boys and is not particularly associated with prematurity. Lack of ganglionic cells within part of the colon may present soon after birth with failure to pass meconium. Occasionally this can lead on to a functional obstruction- green vomit, abdominal distension and sometimes associated with bloody diarrhoea. However, constipation usually dominates the clinical picture. For all these reasons, Hirschprung disease is less likely to be the underlying cause in Kaya’s case.

There’s a helpful distinction between green bilious vomit and non-bilious:

<table>
<thead>
<tr>
<th>Bilious vomiting (green) in neonates &amp; infants</th>
<th>Non-bilious vomiting (yellow)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Intestinal obstruction eg volvulus, duodenal or jejunal atresia, intussusception</td>
<td>Pyloric stenosis</td>
</tr>
<tr>
<td>Necrotising enterocolitis</td>
<td>Gastro-oesophageal reflux</td>
</tr>
<tr>
<td>Gastroenteritis (severe)</td>
<td>Gastroenteritis (mild)</td>
</tr>
</tbody>
</table>

Necrotising enterocolitis

- Disease of preterms (rarely seen in term babies)
- Bowel ischaemia, inflammation, necrosis and even perforation
- Poor feed tolerance, abdominal distension and bloody stools
- X-ray: air in the intestinal wall (pneumatosis intestinalis) and free gas (in perforation)
- Bowel rest (NBM), total parenteral nutrition (TPN), antibiotics
- Surgery to remove perforated or necrotic bowel

Necrotising enterocolitis: pathophysiology

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Impaired immunity
Abnormal bacterial colonisation with lack of anaerobes
Enteral feeding
Impaired intestinal blood flow

Inflammation & ischaemia of bowel wall

Bacteria invade bowel wall
Sepsis
Ascites
Bowel infarction
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Question 3

A twenty month old boy can crawl but is not yet able to walk. Children are normally able to walk independently by 18 months of age. So this boy is showing some delay in his gross motor skills.

Name three diagnostic possibilities.

- A neurological condition such as cerebral palsy
- A neuromuscular condition such as Duchenne muscular dystrophy
- An orthopaedic problem, such as developmental dysplasia of the hip.

What three points would you want to ask in the history?

- Pregnancy and birth history. For example were all the scans ok in pregnancy? Was he born prematurely or on time? What sort of delivery was he and did he need to go to the Special Care Baby Unit?
- Family history. Is there any history in the family of children having delayed development or learning difficulties? Is there any history of hip problems? At what age did his siblings walk?
- Other developmental milestones. In order to establish whether this is an isolated problem with gross motor development, or associated with a more global delay in developmental skills.

What would you expect social milestones to be at this age?

At the age of 20 months I would expect him to have several words, including being able to say mum and dad (to the right person!). By the age of 2 years he should be able to put two words together in a sentence. For social skills he should be able to say hello and wave goodbye. He should look at people when talking to them and want to direct their attention to something he likes eg showing a favourite toy.

How would you assess vision in a child of this age?

I would assess vision by first asking the parents if they have any concerns about their child’s eyesight. I would spend a few minutes just simply watching the child as they sit on their mother’s knee – in particular looking for eye contact and a squint. At this age it would not be possible to perform formal vision testing using for example a Snellen chart. Instead you could present them with a brightly coloured toy and watch them fix on and follow it with their eyes. It is important to remember that fine motor skill development is related closely to visual development – so any visual assessment should include an assessment of fine motor skills as well.

Physical examination shows arms are normal but legs are hypertonic and scissor when he is picked up. What is the diagnosis?

Cerebral palsy

What is the specific name given to this pattern of disability?

This pattern of hypertonia in the lower limbs with scissoring of the legs is suggestive of diplegic cerebral palsy.
**Question 4: answers**

What is the likely diagnosis?  
*Kawasaki’s disease: febrile mucocutaneous syndrome*

What serious complication may occur over the next few weeks?  
*Risk of coronary aneurysms (20%) and small risk of myocardial infarction and sudden death.*

What is the treatment?  
*Treatment with intravenous immunoglobulin and aspirin*

**Notes: diagnosis based on fever for 5 or more days**

Plus 4 out of 5 of the following
- Conjunctivitis
- Lymphadenopathy
- Rash
- Lips- redness, cracking or strawberry tongue
- Extremity changes

**Mnemonic: clear**

<table>
<thead>
<tr>
<th>C</th>
<th>Conjunctivitis</th>
</tr>
</thead>
<tbody>
<tr>
<td>L</td>
<td>Lymphadenopathy and lips</td>
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<tr>
<td>E</td>
<td>Extremity changes</td>
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<tr>
<td>A</td>
<td>Aneurysms</td>
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<tr>
<td>R</td>
<td>Rash</td>
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**Kawasaki’s disease**

- ? Bacterial toxin
- Acute phase response  
  - T cell stimulation & cytokine release
- Early mucocutaneous syndrome
- Late vasculitis

**Prolonged fever >5 days**
- Raised WCC, ESR, CRP
- Thrombocytosis

**Conjunctivitis without pus**
- Red cracked lips / strawberry tongue
- Polymorphous rash on trunk
- Redness around BCG scar
- Hands and feet- red and then peeling

**20% develop coronary aneurysms**
- Best detected on echo
- Small risk of thrombosis and death
- Early immunoglobulin prevents
- Aspirin continued for at least 6 weeks
Question 5

**Cavernous haemangioma**
- “Strawberry naevus” due to proliferation of blood vessels
- Often raised or “lumpy” (unlike capillary haemangiomas or “stork bites”)
- Not usually present at birth but develops in first month
- Typically regresses spontaneously between 2 and 4 years of age
- Commoner in pre-term infants

Question 6

**Sickle cell anaemia**
- Commonest monogenetic condition in the world
- Single nucleotide substitution GTG for GAG
- 2 homozygous genes for defective red blood cells with a tendency to sickle
- Sickled cells cause vaso-occlusion and result in a variety of sickle crises
- Increased infection risk due to auto-infarcted spleen

**Sickle crises**

<table>
<thead>
<tr>
<th>Type</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>PAINFUL</td>
<td>Vaso-occlusion results in ischaemia and pain. Bones, abdomen.</td>
</tr>
<tr>
<td>CHEST</td>
<td>Vaso-occlusion and collapse in the lungs. Commonest cause of mortality – 2%.</td>
</tr>
<tr>
<td>APLASTIC</td>
<td>Temporary bone marrow failure. Severe anaemia. Trigger: Parvovirus B19</td>
</tr>
<tr>
<td>SPLENIC SEQUESTRAION</td>
<td>Life threatening sudden enlargement of the spleen leading to hypovolaemia.</td>
</tr>
<tr>
<td>CEREBROVASCULAR</td>
<td>Vaso-occlusion in cerebral circulation. Cause of significant mortality and morbidity.</td>
</tr>
<tr>
<td>PRIAPISM</td>
<td>Usually nocturnal, with risk of long-term impotence.</td>
</tr>
</tbody>
</table>

**Sickle cell crisis: general principles of management**
- Hydration
- Oxygenation
- Pain relief
- Antibiotics if indicated
- Consider exchange transfusion
Question 7

What is the most likely diagnosis? Single best answer

a) Cyanotic congenital heart disease
b) Group B streptococcal sepsis
c) Rhesus incompatibility
d) Neonatal stroke
e) Neonatal hypoglycaemia

James is clearly unwell, with hypothermia, tachycardia, raised respiratory rate, grunting, prolonged capillary refill and reduced saturations. This combination of signs is strongly suggestive of sepsis and as mum is known to have had prolonged rupture of membranes and was a GBS carrier, this is the most likely aetiology.

a) is unlikely because there are no signs to suggest a cardiac cause
b) is unlikely as this was mum’s first pregnancy and there are no specific signs of cardiac failure, which is the usual result of haemolysis due to rhesus disease
c) is unlikely as this was mum’s first pregnancy and there are no specific signs of cardiac failure, which is the usual result of haemolysis due to rhesus disease
d) there are no signs to suggest a stroke
e) it is always important to check the blood glucose in an unwell child, but hypoglycaemia is unusual in a term baby of a non-diabetic mother. Note though that hypoglycaemia can cause hypothermia, tachycardia and mottled skin with prolonged capillary refill. However it would be more likely to cause apnoeic episodes than a raised respiratory rate

Notes on the unwell term baby: think about

- Sepsis – Group B Strep, E.coli, Listeria
- Hypoglycaemia
- Congenital cardiac anomaly
- Metabolic problem
- Other causes: transient tachypnoea of the newborn, anaemia/haemolysis, birth asphyxia and seizures

Group B streptococcal infection

- ¼ of pregnant women in the UK are group B strep carriers
- Most babies are not affected by GBS sepsis
- 1 in 1,600 will develop early GBS sepsis (within 1st week) with septicaemia +/- pneumonia / meningitis
- 1 in 10 babies with early onset GBS will die
- Maternal antibiotics in labour are associated with 90% risk reduction for early GBS sepsis

Managing GBS sepsis
Always remember the ABC approach!

- Airway
- Breathing – oxygen, CPAP, ventilation
- Circulation – fluids, inotropes
- DEFG – check glucose

Antibiotics
Benzylpenicillin and gentamicin until microbiology results
May require up to 2 weeks benzylpenicillin.
Question 8
16 month old Petr has been previously well with no significant medical history. He was at his grandparents’ house, holding on to a table. Suddenly his eyes rolled upwards and he fell backwards onto a carpeted floor. He went very stiff and started shaking and this continues after he has been brought in by ambulance 15 minutes after the onset. The ambulance crew have given 5mg of rectal diazepam.

After checking ABC and his blood glucose and giving oxygen, what further treatment would you arrange to terminate his seizure? (Single best answer)

a) Intravenous lorazepam  
b) Buccal midazolam  
c) Repeat rectal diazepam  
d) Intravenous infusion of phenytoin  
e) Fast bleep the anaesthetist for rapid sequence induction with thiopentone

Notes
The definition of status epilepticus has changed. It used to refer to a seizure lasting longer than 30 minutes. But now it is regarded as a seizure continuing for longer than 5 minutes, or when shorter convulsive seizures occur one after the other with no recovery in between. Petr’s seizure has already lasted 15 minutes, so he is in status epilepticus.

All of the options given as possible answers are included as options in the NICE (2011) guideline, but only one is appropriate at this stage.

Five step approach
- Onset: check ABC, give oxygen, check glucose, confirm clinical diagnosis
- 5 minutes: buccal midazolam or rectal diazepam or IV lorazepam
- 15 minutes: IV lorazepam, senior help
- 25 minutes: phenytoin infusion over 20 minutes
- 45 minutes: rapid sequence induction of anaesthesia (thiopental)

Initial treatment by the ambulance staff was with rectal diazepam, which is one of the 5 minute options, but this has not worked. That means that the next stage (ideally at 15 minutes, but here at 20 minutes) would be intravenous therapy, which should be faster acting than buccal or rectal administration. A flow chat summarising the steps is shown below.

Causes of status epilepticus
- Febrile convulsion
- Known epilepsy + acute illness
- Meningoencephalitis
- Metabolic/ electrolyte abnormality
- Drugs/ intoxication/ poisoning
- Intracranial haemorrhage
- Trauma
Status epilepticus management flow chart

**ABCDEFG**
5 mins after convulsions start

Yes → **IV Access?**

No → Midazolam Buccal (0.5mg/kg) OR Diazepam Rectal (0.5mg/kg)

**Lorazepam 0.1mg/kg IV/IO**

**After 10 minutes**: If still fitting give 2\textsuperscript{nd} dose of
Lorazepam (0.1mg/kg) + call for senior help

**Definitely a seizure??**

Wait 10 further minutes (but prepare Phenytoin)

**Senior help now needed**
Seek ICU/anaesthetic advice
Give **20mg/kg Phenytoin**
IV/IO infusion over 20 mins

1. Anaesthetist must be present
2. Rapid Sequence Induction (RSI) with Thiopentone unless hypotensive

Source: APLS protocols. © 2010