

The Management of Acute Abdominal Pain in Children with Sickle Cell Disease

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The Management of Acute Abdominal Pain in Children with Sickle Cell Disease

This guideline is aimed at all children in King's College Hospital with sickle cell disease (SCD) and acute abdominal pain. The guideline applies to all patients with SCD who are currently under the care of the Paediatric Haematology team. It is mainly aimed at being a tool for the medical team managing these patients, but any member of the multidisciplinary team may find it useful.

Causes of Acute Abdominal Pain

Acute abdominal pain is a common symptom in children with sickle cell disease. Possible causes include:

Common

- Constipation
- Acute vaso-occlusion (called abdominal painful crisis, girdle syndrome)
- Cholecystitis
- Gastroenteritis
- Urinary tract infection

Less Common

- Pancreatitis
- Parvovirus B19 infection
- Splenic sequestration
- Hepatic sequestration
- Acute pancreatitis
- Acute appendicitis
- Acute chest syndrome (referred pain)
- Gastritis/peptic ulcer disease
- Septicaemia

Rare

- Hepatic infarction
- Viral hepatitis
- Ischaemic cholangiopathy
- Ischaemic colitis
- Papillary necrosis
- Bladder infarction

Initial Management and Assessment

If the child is in pain, analgesia should be given according to the Pain Guidelines. If the child is shocked or collapsed they should be urgently resuscitated according to standard protocols.

History should initially focus on identifying the more common diagnoses including:

- Does pain feel like previous episodes of pain due to SCD?
- Has the child been constipated or using codeine recently?
- Has the child been taking non-steroidal anti-inflammatory drugs frequently?
- Is there diarrhoea or vomiting?
- Is there a history of gall stones or cholecystitis?
- Has jaundice increased?
- Is there a history of splenomegaly or splenic sequestration?

Examination

Full examination should be performed including for:

- Presence of bowel sounds: in severe abdominal sickling bowel sounds are usually absent, but a silent abdomen also raises the possibility of acute surgical complications, including perforation.
- Signs of appendicitis
- Signs of cholecystitis
- Splenomegaly
- Hepatic enlargement or tenderness

Initial Investigations

- FBC, U&E's, reticulocyte count
- Group and save
- CRP
- Liver function tests including ALT
- Serum amylase
- Blood cultures if temperature $>38^{\circ}\text{C}$
- Urine for microscopy and culture
- Pulse oximetry on air
- Stool culture if diarrhoea

Initial Management

As mentioned the child's condition should be stabilised and appropriate analgesia given.

- Intravenous fluids should be started according to guidelines if the pain is severe, there are no bowel sounds or if there is diarrhoea or vomiting.
 - 1-3 years 150 ml/kg/24h
 - 4-6 years 145 ml/kg/24h
 - 7-14 years 105 ml/kg/24h
 - 15-18 years 90ml/kg/24h
- If there are no bowel sounds or a surgical cause such as acute appendicitis is suspected, the child should be made nil-by-mouth and the paediatric surgery team asked to see the patient urgently.
- Penicillin V should be continued at the prophylactic dose unless the temperature is $>38^{\circ}\text{C}$ or there are no bowel sounds; in which case intravenous antibiotics should be started:
 - Cefuroxime (if not allergic) 20mg/kg (max 750mg) 8 hourly
- If the patient is taking hydroxyurea, it should be continued unless the patient is nil-by-mouth, the blood tests show evidence of toxicity (neutrophils $< 1.0 \times 10^9/\text{l}$, platelets $< 80 \times 10^9/\text{l}$, reticulocytes $< 80 \times 10^9/\text{l}$, $\geq 50\%$ increase in serum creatinine, ALT $> 110 \text{ IU/l}$).
- Incentive spirometry should be started according to ward protocols.

Further Investigations

The following investigations may be appropriate depending on the initial findings:

- Ultrasound of the liver, gall bladder, pancreas or spleen if there is evidence of cholecystitis, pancreatitis, hepatic enlargement or tenderness, splenic enlargement or tenderness.
- Chest X-ray if there is chest pain, hypoxia, or chest signs, or an acute abdomen.

- Abominal X-ray if acute abdomen (following discussion with paediatric surgeons). This may also show evidence of constipation.
- Ultrasound of kidneys, ureters and bladder if there is haematuria, renal colic or severe lower abdominal pain.
- Parvovirus B19 serology if the reticulocyte count is $<100 \times 10^9/l$.
- Hepatitis serology if the ALT is >110 IU/l

Further Specific Management of Common Causes

This will depend on the initial diagnosis.

Acute vaso-occlusion

- Children with abdominal sickling are usually in severe pain and may require admission to HDU for adequate amounts of analgesia to be given safely. This will often require patient- or nurse-controlled analgesia, with appropriate monitoring for efficacy and sedation.
- The patient should be re-examined frequently to ensure that the pain is being controlled and that another underlying cause of the pain is not emerging i.e. acute surgical conditions. This should include chest examination to ensure that acute chest syndrome is not developing. Examination should be hourly for the first 6 hours and 6-12 hourly thereafter depending on progress.
- The paediatric surgical team should be informed of the patient's admission, although surgery should not be undertaken without discussion with consultant paediatric surgeons, paediatric haematologist and general paediatrician. In general conservative management is followed unless there is a definite surgical diagnosis such as acute appendicitis or perforation.
- Blood tests should be performed daily to monitor renal and hepatic function and full blood count. If the haemoglobin falls more than 2g/dl below the steady-state, or below 5g/dl, top-up blood transfusion may be necessary.

Constipation

- Typically the pain is moderate or mild, with an intermittent or colicky nature. Usually there is a history of infrequent or painful defaecation.
- Appropriate analgesia should be given, depending on the severity of the pain. Paracetamol and ibuprofen are typically sufficient, although opiates are sometimes needed.
- The child should be encouraged to drink.
- Lactulose and senna should be started.
 - Lactulose: 1 month – 1 year: 2.5ml bd, 1-5 years 5ml bd, 5-10 years 10ml bd, 10-18 years 15ml bd.
 - Senna syrup: 1 month – 2 years: 0.5ml/kg (max 2.5ml), 2-6 years 2.5-5ml od, 6-12 years 5-10ml od, 12-18 years 10-20ml od
- An enema may be appropriate.

Cholecystitis

- This is usually diagnosed with a combination of typical symptoms and signs, increased bilirubin and gallstones with a thickened gall bladder on ultrasound examination.
- The paediatric surgical team should be involved in the care of the patient at an early stage.

- If vomiting is severe a nasogastric tube may be necessary in addition to nil-by-mouth, intravenous fluids and cefuroxime.
- Management is typically conservative.
- At discharge it should be ensured that the patient has a follow-up appointment with the paediatric surgeons. Further investigations such as ERCP will be organised by the paediatric surgery team.
- After one episode of cholecystitis, an elective cholecystectomy will typically be performed depending on surgical considerations and the views of the patient/family.

Other diagnoses

- Other diagnoses should be managed according to standard protocols.
- Acute vaso-occlusive pain and acute chest syndrome can develop during and complicate any admission for unrelated reasons, such as gastroenteritis, or urinary tract infection.
- In general, children with sickle cell disease and abdominal pain should be monitored closely, with particular attention to fluid balance, early treatment of possible infection, and early investigation of any respiratory or neurological symptoms or signs.

References

Ahmed S, Shahid RK, Russo LA. Unusual causes of abdominal pain: sickle cell anemia. *Best Practice and Research Clinical Gastroenterology* 2005; 19, 297-310.

Bunn HF. Pathogenesis and treatment of sickle cell disease. *N Engl J Med* 1997; **337**: 762-69.