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

## Document Information

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<td>Responsible committee</td>
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### For Child Health Clinical Guidelines Groups’ use only

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| Dr Moira Dick  
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| Dr Sue Height  
(Consultant Paediatric Haematologist) | 2       | 30/12/13   |                           |
| Simon Broughton  
(Consultant Paediatrician) | 2       | 30/12/13   |                           |
| Professor Mark  
Davenport  
(Consultant paediatric surgeon) | 2       | 30/12/13   |                           |

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

Background
Acute abdominal pain is a common reason for admission to hospital in children with sickle cell disease (SCD). It may be related directly to vaso-occlusion, be a complication of treatment or unrelated to SCD. 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.

Contents of guideline
- Aim
- Causes of abdominal pain
- Urgent management and assessment
- Examination
- Initial investigations
- Initial management
- Further investigations
- Further specific management of common causes
  - Acute abdominal sickling
  - Constipation
  - Cholecystitis
  - Other diagnoses
The Management of Acute Abdominal Pain in Children with Sickle Cell Disease

Aim
To provide guidance on the management of children with sickle cell disease and abdominal pain admitted to the paediatric wards at King’s College Hospital NHS Foundation Trust. 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 with faecal impaction
- Acute abdominal sickling (also called abdominal painful crisis, girdle syndrome)
- Gastroenteritis
- Urinary tract infection

Less Common
- Cholecystitis
- Pancreatitis
- Parvovirus B19 infection
- Splenic sequestration
- Hepatic sequestration
- Acute appendicitis
- Acute chest syndrome
- Gastritis/peptic ulcer disease
- Septicaemia

Rare
- Hepatic infarction
- Viral hepatitis
- Ischaemic cholangiopathy
- Ischaemic colitis
- Renal papillary necrosis

Urgent 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 jaundiced increased?
• Are there symptoms of a urinary tract infection?
• Is there a history of splenomegaly or splenic sequestration?

**Examination**

Full examination should be performed, with particular reference to:

• 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°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: 90 ml/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°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 x 10⁹/l, platelets < 80 x 10⁹/l, reticulocytes < 80 x 10⁹/l, ≥ 50% increase in serum creatinine, ALT > 110 IU/l).
- Incentive spirometry should be started according to ward protocols.

**Further Investigations**

Further 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, 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 <100x10⁹/l.
• Hepatitis serology if the ALT is >110 IU/l

Further Specific Management of Common Causes
This will depend on the initial diagnosis.

Acute abdominal sickling
• 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 will usually 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.
• Movicol should be started, and lactulose and senna added as necessary.
  o Movicol Paediatric Plain: under 1 year: ½ - 1 sachet daily; 1-5 years (until constipation resolves): 2 sachets on first day, then 4 sachets daily for 2 days, then 6 sachets daily for 2 days, then 8 sachets daily; 5-12 years (until constipation resolves): 4 sachets on first day, then increased in steps of 2 sachets daily to maximum of 12 sachets daily.
  o Movicol: 12-18 years: 4 sachets on first day, then increased in steps of 2 sachets daily to maximum of 8 sachets daily
  o Lactulose: 1 month – 1 year: 2.5ml bd, 1-5 years: 5ml bd, 5-10 years: 10ml bd, 10-18 years 15ml bd.
  o 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


David Rees
28th December 2013