NHS SCOTLAND

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SCOTTISH PAEDIATRIC AND ADULT HAEMOGLOBINOPATHIES NETWORK

# Scottish Paediatric & Adult Haemoglobinopathy Network

## The Management of Acute Abdominal Pain in children with Sickle Cell disease

**NOTE**
This guideline is not intended to be construed or to serve as a standard of care. Standards of care are determined on the basis of all clinical data available for an individual case and are subject to change as scientific knowledge and technology advance and patterns of care evolve. Adherence to guideline recommendations will not ensure a successful outcome in every case, nor should they be construed as including all proper methods of care or excluding other acceptable methods of care aimed at the same results. The ultimate judgement must be made by the appropriate healthcare professional(s) responsible for clinical decisions regarding a particular clinical procedure or treatment plan. This judgement should only be arrived at following discussion of the options with the patient, covering the diagnostic and treatment choices available. It is advised, however, that significant departures from the national guideline or any local guidelines derived from it should be fully documented in the patient's case notes at the time the relevant decision is taken.

# 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 (VOC), be a complication of treatment or unrelated to SCD.

This guideline is aimed at all children in Scotland with sickle cell disease and acute abdominal pain.

# 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 (this can complicate or accompany other causes of abdominal pain)
- 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/ pneumonia
- Gastritis/peptic ulcer disease
- Septicaemia

## Rare

- Hepatic/splenic infarction
- Viral hepatitis
- Ischaemic cholangiopathy
- Ischaemic colitis
- Renal papillary necrosis
- Testicular torsion

## Urgent Management and Assessment

If the child is shocked or collapsed they should be urgently resuscitated according to standard resuscitation protocols.

If the child is in pain, analgesia should be given according to the Painful Crisis Guidelines available on the SPAH Paediatric Guideline website page.

## History

Should initially focus on identifying the more common/serious diagnoses including:

- Does pain feel like previous episodes of pain due to SCD?
- Has the child been constipated or recently using opiate analgesia?
- Has the child been taking non-steroidal anti-inflammatory drugs frequently?
- Is there diarrhoea, vomiting or fever?
- Is there a history of gall stones or cholecystitis?
- Has jaundice increased?
- Are there symptoms of a urinary tract infection?
- Is there a history of splenomegaly/hepatomegaly or splenic/hepatic 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 – please compare to steady state values

- FBC and reticulocyte count (malaria screen if recent travel to high risk area)
- Blood Group and save
- UE, LFT, Amylase, CRP
- Venous/capillary blood gas and Lactate
- Blood cultures if temperature >38C or diarrhoea/ vomiting
- Urine for dipstick, microscopy and culture
- Pulse oximetry on air
- Stool culture/virology if diarrhoea

## Initial Management

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

- 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. Consider NGT placement after discussion with Paediatric surgery.
- Penicillin V should be continued at the prophylactic dose unless the temperature is >38C or there are no bowel sounds; in which case intravenous antibiotics should be started: Antibiotics should be as per local guidelines for treatment of suspected intra-abdominal sepsis.
- If the patient is taking Hydroxycarbamide, it should be continued unless the patient is nil-by mouth or the blood tests show evidence of toxicity (neutrophils < 1.0 $x10^9$/l, platelets < $80x10^9$/l, reticulocytes <80 $x10^9$/l, > 50% increase in serum creatinine, ALT > 110 IU/l).
- Incentive spirometry should be started according to local protocols.

## Fluid management

Dehydration occurs readily in children with sickle cell disease due to impairment of renal concentrating ability and may aggravate to sickling due to increased blood viscosity. Diarrhoea and vomiting are thus of particular concern. Patients may also have cardiac or respiratory compromise and so fluid overload must also be avoided.

**Careful assessment of individual fluid status, administration of an appropriate hydration regimen and close monitoring of fluid balance is therefore imperative.**

An IV line should be established whenever parenteral opiates have been given, or if the patient is not taking oral fluids well. In the less ill patient who is able to drink the required amount, hydration can be given orally. As an alternative, consider a nasogastric tube in an alert patient.

## 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.
- Ultrasound of kidneys, ureters and bladder if there is haematuria, renal colic or severe lower abdominal pain.
- Chest X-ray if there is chest pain, hypoxia, chest signs, or an acute abdomen.
- Abdominal X-ray if acute abdomen (following discussion with paediatric surgeons). This may also show evidence of constipation.
- Parvovirus B19 serology if the reticulocyte count is < $100x10^9$/l.
- Hepatitis serology if the ALT is >110 IU/l

## Further Specific Management of Common Causes

This will depend on the initial diagnosis**.**

### Acute 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. In patients with limb girdle syndrome, lower back and pelvic pain commonly accompanies abdominal pain. This will often require patient or nurse-controlled analgesia, with appropriate monitoring for efficacy and sedation.
- **Note children with abdominal VOC and absent bowel sounds/ evidence of ileus are at risk of progressing to ischaemic colitis and bowel perforation. These patients require careful/ frequent monitoring.** 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, consultant paediatric haematologist/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 20g/l below the steady-state, or below 50g/l, top up blood transfusion will usually be necessary.
- Patients showing clinical deterioration/ lack of improvement despite optimum management with fluids, antibiotics, analgesia may require either top up or exchange transfusion.

## Constipation

- Typically the pain is moderate or mild, with an intermittent or colicky nature. Usually there is a history of infrequent or painful defecation.
- Appropriate analgesia should be given, depending on the severity of the pain. Paracetamol and ibuprofen are typically sufficient, although opiates are sometimes needed. Note that opiates are likely to exacerbate constipation and may exacerbate pain.
- The child should be encouraged to drink.
- The choice of laxatives should follow local guidelines.
- Patients prescribed regular opiate analgesia should be considered for regular laxative treatment to prevent constipation.

## Cholecystitis

- This is usually diagnosed with a combination of typical symptoms and signs, increased bilirubin ALP and GGT, and gallstones with a thickened gall bladder or dilated common bile duct on ultrasound examination +/- MRCP. Pancreatitis in sickle cell patients is often secondary to gallstones. An amylase/lipase should always be checked alongside LFT in patients with severe upper/ RUQ abdominal pain.
- The paediatric surgical/gastroenterology team should be involved in the care of the patient at an early stage.
- If vomiting is severe or bilious a nasogastric tube may be necessary in addition to nil-by mouth, intravenous fluids and IV antibiotics as per local guidelines.
- Management is typically conservative.
- At discharge it should be ensured that the patient has a follow-up appointment with the paediatric surgeons/gastroenterology. Further investigations such as MRCP/ERCP may be organised in discussion with the paediatric surgical/gastroenterology 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

Links to additional guidelines including the Acute Anaemia Guideline and Splenic and Hepatic Sequestration are available on the SPAH Paediatric Guideline website page.

# Note

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.