

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

Document Information			
Version:	3	Date:	10 th November 2015
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Responsible committee	Child Health Clinical Governance & Risk Committee		
Review date:	December 2017		
Target audience:	Paediatricians, paediatric haematologists, haematologists		
Stakeholders/ committees involved in guideline development :	General Paediatrics Paediatric haematology Haematology Pharmacy (Shereen Boreland) Microbiology (Amanda Fife)		

For Child Health Clinical Guidelines Groups' use only	
Assessed by:	Child Health Clinical Guidelines Group
Assessment date:	
Approved by :	Child Health Governance and Risk Committee
Approval Date	
Author notified:	

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Ratified by:	
Date ratified:	
Reference No.:	
Date when guideline comes into effect:	

Document History

Document replaces: 2

Consultation distribution (before ratification)

Sent to	Version	Date	Actions taken as a result
Dr Moira Dick (Consultant Community Paediatrician)	3	30/07/15	
Dr Sue Height (Consultant Paediatric Haematologist)	3	30/07/15	Codeine changed to dihydrocodeine for <12 yrs Fluid balance Addition of lactate to assessment Tazocin and gentamicin as antibiotics if absent bowel sounds
Simon Broughton (Consultant Paediatrician)	3	30/07/15	
Professor Mark Davenport (Consultant paediatric surgeon)	3	30/07/15	
Dr Subarna Chakravorty	3	24/08/15	
Dr Amanda Fife	3	6/11/15	Addition of antibiotic choices if Penicillin allergic (Vancomycin/Ciprofloxacin and Metronidazole)

Reviews and updates (including CGG comments)

Date	New version no.	Summary of Changes	Author of change/s
17/11/15	4	Add in Lactate/blood gas	Simon Broughton

Dissemination schedule (after ratification)

Target audience(s)	Method	Person responsible
Paediatricians		D Rees S Broughton
Paediatric Haematology		D Rees
Emergency Department		S Broughton

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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.

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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 recently using codeine (or dihydrocodeine if <12 years)?
- Has the child been taking non-steroidal anti-inflammatory drugs frequently?
- Is there diarrhoea or vomiting?

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- 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 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 – please compare to steady state values

- FBC, U&E's, reticulocyte count
- Blood Group and save
- CRP
- Blood gas and Lactate
- 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 if the pain is severe, there are no bowel sounds or if there is diarrhoea or vomiting. Normal full maintenance fluids should be started; however, if the child is assessed as being clinically dehydrated, the rate should be increased guided by frequent clinical assessment up to 150% maintenance, with careful monitoring and fluid balance. It is important to avoid fluid overload, especially if there is associated respiratory symptoms.
- Please see the fluid guideline for guidance on volume of fluids of normal maintenance.
- 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 $>38^{\circ}\text{C}$ or there are no bowel sounds; in which case intravenous antibiotics should be started:
 - Tazocin 90mg/kg (Maximum 4.5g) QDS if not allergic PLUS
 - Gentamicin 7mg/kg (max 350mg) OD
- If the child is Penicillin allergic then please start the following intravenous antibiotics for Sickle Abdomen (for ages outside these ranges, please consult the BNFC):
 - Vancomycin (Child 1 month–18 years 15 mg/kg every 8 hours (maximum daily dose 2 g), adjusted according to plasma concentration)
 - Ciprofloxacin (Child 1 month–18 years 10 mg/kg (max. 400 mg) every 8 hours)
 - Metronidazole (Child 2 months–18 years 7.5 mg/kg (max. 500 mg) every 8 hours)

- 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 $< 1.0 \times 10^9/l$, platelets $< 80 \times 10^9/l$, reticulocytes $< 80 \times 10^9/l$, $\geq 50\%$ 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.
- Abdominal 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 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 20g/l below the steady-state, or below 50g/l, 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.
 - Movicol Paediatric Plain: under 1 year: $\frac{1}{2}$ -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.
- Movicol: 12-18 years: 4 sachets on first day, then increased in steps of 2 sachets daily to maximum of 8 sachets daily
- 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 or bilious a nasogastric tube may be necessary in addition to nil-by-mouth, intravenous fluids and Tazocin and gentamicin.
- 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.

9th November 2015