Sickle Cell Disease: Emergency Management

Guidelines for Shared Care Centres and Community Staff

Reference: CG1411
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Approved: September 2015
Approved by D&TC: 10th July 2015
Review Due: September 2018

Intended Audience

This document contains information and clinical guidelines for management of children registered at the Haematology department at Sheffield Childrens NHS Foundation Trust presenting to designated shared care centres or in the community. It is to be used by staff within shared care centres and community services for information whenever they are caring for these children either in hospital or at home.

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1. Background

Children with sickle cell disease are prone to a number of health problems. Their parents are asked to seek medical advice urgently if they are concerned about their child, particularly if they are feverish, unusually pale, complaining of abdominal pain, having difficulty breathing or have neurological symptoms such as fits, drowsiness, confusion or weakness. Please take all symptoms seriously and listen to parental concerns. Children with sickle cell disease can have all the conditions and accidents that a normal child presents with but in addition may also have the problems listed above.

2. Painful Crisis (Vaso-Occlusive Crisis)

This is the most common reason for a child with sickle cell disease to seek medical help. It can occur in any part of the body – muscles, bones, abdomen, chest etc.

Dactylitis is a painful swelling of the fingers and hands or toes and feet. It is more common in babies and may be the first complication a child with sickle cell disease suffers from. It can occur in older children but is rare in adults.

It is important to remember that potentially serious complications of sickle cell disease can present without pain (stroke, aplastic crisis).

Management of pain is primarily supportive as there is no specific treatment available. As well as dealing with the pain it is important to assess thoroughly for signs of infection, and follow up any positive findings in the history or examination. Check an oxygen saturation. Give analgesia, hydrate if clinically required and treat any associated infection. There is no benefit in giving oxygen unless a low saturation has been found or the person is thought to be shocked.

Pain relief

There are no objective criteria by which to assess the severity of a painful crisis, so the patient’s or parent’s account of the level of pain should be accepted and the analgesia targeted to that level from the start. Pain charts should be used to help children express their pain over time. If in doubt err towards an over, rather than an underestimate of the severity of the pain. The objective should be complete pain relief as quickly as possible (within 30 minutes of presentation). Do not be anxious about the use of opiates if necessary. Once the child is pain free you will have gained their confidence, they will be able to relax and are more likely to allow you to examine them properly.

Mild – moderate pain

Parents are likely to have tried simple analgesia at home prior to bringing the child. Check the doses they have been using and that they have been using regular analgesia, rather than waiting for the pain to return. It may be that all the family needs is dosing advice and a new supply of analgesia – they may only need to stay for a few hours to make sure the pain is under control.
Suggested oral pain relief in children > 6 months of age:

Paracetamol in combination with Ibuprofen OR Diclofenac – regular 4-6hrly dosing with the addition of oral morphine if the above is inadequate.

<table>
<thead>
<tr>
<th>Drug</th>
<th>Dose (age 6 months to 18 yrs)</th>
<th>Comments</th>
</tr>
</thead>
<tbody>
<tr>
<td>Paracetamol</td>
<td>15 to 20 mg/kg/dose (max 1g) 4 – 6 hrly but Max 75 mg/kg/24 hrs Do not exceed 4 g per 24 hrs</td>
<td>Age &gt; 6 months</td>
</tr>
<tr>
<td>Ibuprofen OR</td>
<td>5 – 7.5 mg/kg/dose 6 hrly (max 30 mg/kg/24 hrs) Do not exceed 2.4g per 24 hrs</td>
<td>Age &gt;6 months Caution with severe asthma, coagulation abnormalities and renal dysfunction</td>
</tr>
<tr>
<td>Diclofenac</td>
<td>1 – 1.5 mg/kg/dose (max 50mg) 8 -12 hrly Max 3 mg/kg/24 hrs Do not exceed 150 mg per 24 hrs</td>
<td>Age &gt; 6 months Caution with severe asthma, coagulation abnormalities and renal dysfunction</td>
</tr>
<tr>
<td>Oral Morphine</td>
<td>200 micrograms/kg 4 hrly 200 – 300 micrograms/kg 4 hrly 5 – 10 mg 4hrly</td>
<td>Age 6 – 12 months Age 1 – 12 yrs Age 12 – 18 yrs Monitor regularly for signs of sedation/ respiratory depression</td>
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</tbody>
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If still in pain with this treatment, treat as severe.

Severe pain

Start a morphine infusion after an initial slow bolus

Doses: Morphine bolus 200microgram/kg

Morphine infusion **20microgram/kg/hr as a starting dose**1mg/kg in 50ml of diluent for children less than 50kg, 20microgram/kg/hour is 1ml/hour. This can be titrated to effect. Over 50kg, prescribe as 50mg/50ml rate adjusted according to body weight.

When prescribing opiate analgesia always prescribe laxatives, anti pruritics and anti emetics at the same time.
Assess adequacy of pain relief regularly and frequently and increase or reduce analgesia as appropriate. Monitor regularly for signs of sedation/respiratory depression. Record pain scores and vital signs observations.

If a child is requiring intravenous opiate analgesia please discuss as soon as possible with haematology staff at Sheffield Children’s Hospital - there is a Consultant Haematologist available for advice at all times.

**Hydration**

Almost all patients requiring admission for a painful crisis will need intravenous fluids for a variable period. Use dextrose 5% Sodium Chloride 0.45% at at least maintenance rate in addition to oral intake. Monitor fluid balance carefully. People with Sickle Cell Disease need this large fluid intake to compensate for their poor renal urinary concentrating ability. Monitor U+Es as they may also be less able to excrete sodium. Continue until the patient is pain free and able to take normal amounts orally.

3. **Hyposplenism and Susceptibility to Infection**

Most sickle cell patients are hyposplenic and as such susceptible to a range of infections with encapsulated organisms such as pneumococcus and Haemophilus influenzae. All sickle cell patients should have their normal childhood vaccinations including pneumococcus and be taking penicillin prophylaxis. However this is only partially effective and compliance with penicillin is often poor.

Children over 6 months of age should be offered yearly influenza vaccination.

*Pneumococcal vaccine boosters with Pneumovax are required throughout life.*

All sickle cell disease patients who are pyrexial should be carefully assessed and have an infection screen including FBC, blood, urine, stool cultures, an NPA if clinically indicated and a chest X-ray. They should then be started either on IV Cefotaxime if unwell (and may need IV fluids) or oral amoxicillin if relatively well. If there are chest signs clarithromycin should be added (see BNFc for doses).

Other investigations should be guided by the clinical findings. Remember that osteomyelitis is more common in people with sickle cell disease than in the general population and may be due to Salmonella.

Once the child has been assessed and antibiotics started the case should be discussed with the clinical haematology staff at Sheffield Children’s Hospital – see contact numbers below.

4. **Aplastic Crisis**

This is Parvovirus associated red cell aplasia presenting with a dramatic fall in haemoglobin and a low reticulocyte count. There may be a preceding history of ‘slapped cheek’ syndrome. This is infectious and more than one family member may be affected.
Always check a FBC and reticulocyte count urgently in any sickle patient seen with a sudden onset of pallor or symptoms of acute anaemia. If the haemoglobin is low and the reticulocyte count high the likely cause is sequestration, where as if haemoglobin is low and reticulocyte count low aplasia is more likely.

It is obviously important to know the child’s baseline haemoglobin so that a comparison can be made, a drop of >20g/l would be considered significant. Treatment is simple top-up transfusion with phenotypically matched blood (after taking a sample for Parvovirus serology) to achieve the usual haemoglobin for that child. Over transfusion should be avoided.

Please contact the Clinical Haematology team at Sheffield Children’s Hospital to keep us informed if a transfusion is needed as it may be more appropriate to transfer the child to us.

5. Acute Splenic or Hepatic Sequestration

Splenic sequestration generally occurs in children less than 5 years of age and presents with sudden abdominal pain, splenic enlargement and a dramatic drop in haemoglobin (with a raised reticulocyte count, in contrast to aplastic crisis).

This is a medical emergency. The child will need an urgent red cell transfusion to deal with the fulminant cardiac failure caused by the acute anaemia. Sometimes the child is in extremis and it is necessary to use uncross-matched O negative blood. Do not use crystalloids until the patient has been transfused, to avoid exacerbating cardiac failure.

Please discuss urgently with a member of the clinical haematology team at Sheffield Children’s Hospital if you suspect this diagnosis.

Hepatic sequestration occurs in older patients whose spleen has atrophied and presents as above, but with hepatic enlargement.
6. **Chest Crisis (Acute Chest Syndrome)**

The pathophysiology of acute chest syndrome is complex and poorly understood, but leads to lung sequestration and presents with chest pain, fever, tachycardia, tachypnoea and hypoxia. Cough and x-ray signs can appear late. The child is likely to need ITU support.

**The clinical haematology staff at Sheffield Children’s Hospital should be contacted urgently**

Give

- Oxygen,
- Analgesia,
- Maintenance fluids
- Start on broad spectrum antibiotics (cefotaxime and clarithromycin) whilst transfer is arranged.

7. **Stroke (Cerebro-Vascular Syndrome)**

This usually presents as a stroke but occasionally as an encephalopathy.

**Urgent transfer of any sickle child with neurological signs should be arranged**

They will need exchange transfusion followed by a long term transfusion program to keep the % sickle haemoglobin low to prevent further stroke.

8. **Priapism (Painful Erection)**

Priapism is caused when sickled red cells block the blood vessels of the penis. It can occur at any age and is more common at night or early morning. A warm bath, analgesia and passing urine may help to relieve it, but if it persists for more than 2 hours the child should be seen at hospital where intravenous fluids and analgesia should be started.

**The child should then be discussed urgently with clinical haematology staff at Sheffield Children’s Hospital as exchange transfusion or penile aspiration may be required**

*Delay in treatment may cause more permanent damage to erectile tissue*

9. **Surgery**

Children with sickle cell disease may need elective surgery eg: for tonsillectomy or grommet insertion. Those with recurrent splenic sequestration will require splenectomy and those with symptomatic gall bladder disease will need cholecystectomy.

Pre operative management varies according to the type of operation and previous complications experienced by the child. All surgery should be carried out at a paediatric surgical centre with experience in managing children with sickle cell disease.
10. Contact Details

To obtain paediatric haematology advice telephone Sheffield Children’s Hospital switchboard (0114 271 7000) and ask to speak to the Haematology Consultant or SpR on call. There is a doctor available to give haematology advice at all times. The haematology/oncology ward is also always open (0114 271 7309) but there may not be an experienced haematologist immediately available on the ward, so bleeping the doctor through switchboard is usually the best option.

11. References

BNFc


