General Management of Children with Sickle Cell Disease including Pain and Infection

Reference: CG1419
Written by: Dr Jenny Welch
Peer reviewer: Dr Jeanette Payne
Approved: August 2018
Approved by D&TC: 9th June 2018
Review Due: August 2021

Intended Audience

This document contains information and clinical guidelines for management of children attending the Sheffield Children’s Hospital Haematology department and Ward. It is to be used by staff within the Trust whenever they are caring for children with Sickle Cell Disease.

Table of contents

1 Background
2 Painful Crisis
3 Possible Acute Complications
4 Discharge Information
5 Hyposplenism and Susceptibility to Infection
6 Jaundice
7 References
General Management of Children with Sickle Cell Disease including Pain and Infection

1 Background

Children with sickle cell disease (SCD) 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. The most common presentation is with pain. Children with SCD can also have all the conditions and accidents that a non SCD child presents with.

It is important to remember that potentially serious complications of sickle cell disease can present without pain (stroke, aplastic crisis). These problems are covered in separate Haematology & Oncology Unit guidelines.

All the families are given verbal and written information about when and how to contact the Haematology & Oncology Unit should they need advice or admission. They are told that they do not have to go through the Emergency Department (ED) or the Acute Admissions unit (AAU). Some families still access services this way and if referred a known patient via ED or AAU the haematology team should be ready to take over their care immediately. This allows staff who have specialist knowledge and training in the management of Sickle Crisis to assess and treat the child with confidence. It may be the child will be able to be discharged the same day.

However staff in the ED also have access to all Sickle Protocols via the intranet. Those in shared care hospitals have 24/7 access to a Consultant Paediatric Haematologist from the specialist team and internet access to ‘Sickle Cell Disease: Emergency Management Guidelines for Shared Care Centres and Community Staff’

Access to the service is a topic suitable for audit

Sickle pain should be treated as an acute medical emergency and adequate pain relief offered within 30 minutes of presentation. This is a NICE requirement and is the subject of regular audit.
General Management of Children with Sickle Cell Disease including Pain and Infection

2. Painful Crisis (vaso-occlusive crisis)

There is a Sickle Cell painful crisis admission proforma (H&O/Form1/1419) which is available in a folder above the doctors work area on the Haematology & Oncology Unit. Please use this to document a painful crisis admission as it guides you through the assessment and management process.

Painful crisis is the most common reason for a child with SCD to seek medical help. It can occur in any part of the body – limbs, muscles, bones, abdomen, chest. It is important to consider other causes of pain especially if the child feels the pain is not typical of a sickle crisis.

Remember to ask about any analgesia already taken by the patient before presenting to the hospital.

**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 SCD suffers. It can occur in older children but is rare in adults.

**Abdominal pain** is common and often due to sickle crisis. It can mimic other conditions but it is important to be guided by the clinical picture. It may be necessary to exclude cholecystitis, pancreatitis, appendicitis and urinary tract infection all of which can occur in SCD.

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. The child only needs supplementary oxygen if their saturations are low. There is no benefit in giving oxygen unless a low saturation has been found or the child is shocked.

All patients presenting with pain should be clinically assessed and monitored:

- Blood pressure
- Oxygen saturation
- Pulse
- Respiratory rate
- Temperature

Give analgesia, hydrate if clinically required and treat any associated infection.

**Pain relief**
The patient/carer is usually an expert in their condition. Discuss the planned treatment for this episode of pain with them. Include in the history treatments received if there have been similar episodes and how effective that was.

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.
General Management of Children with Sickle Cell Disease including Pain and Infection

**Pain charts** should be used to help children express their pain over time. Sheffield Childrens Hospital use a 0-10 visual analogue chart using Wong-Baker Analogue or Faces Pain Rating Scale for children and FLACC (Face, Limbs, Activity, Cry, Consolability) for infants. This system should be used throughout the Trust. 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 mins). 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.

Check if there are any particular concerns they have about the current episode (school work, holidays, parental absence for example). Discuss if any psychological or social support is needed (if appropriate).

Encourage the child to use their own coping mechanisms (for example, relaxation techniques or distraction with computer games or DVDs) in addition to the pharmacological strategies.

Ensure that the drug, dose and route of administration are all suitable for the severity of pain and age of the child. It may be helpful to review the patient’s notes to see what has been used successfully in the past.

**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 pain relief:

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

Do not use corticosteroids in the management of an uncomplicated acute painful sickle episode.
## General Management of Children with Sickle Cell Disease including Pain and Infection

<table>
<thead>
<tr>
<th>Drug</th>
<th>Dose by mouth (age 6 months to 18 yrs)</th>
<th>Comments</th>
</tr>
</thead>
<tbody>
<tr>
<td>Paracetamol</td>
<td>15mg/kg/dose (max 1g) 4 – 6 hrly Do not exceed 4g per 24 hrs</td>
<td>max 4 doses in 24 hours</td>
</tr>
<tr>
<td>Ibuprofen</td>
<td>5 – 7.5mg/kg/dose 6 hrly (max 30mg/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 – 1.5mg/kg/dose (max 50mg) 8 -12 hrly Max 3mg/kg/24 hrs Do not exceed 150mg 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 –10mg 4 hrly</td>
<td>Age 6 – 12 months Age 1 – 12 yrs Age 12 – 18 yrs Monitor regularly for signs of sedation/ respiratory depression</td>
</tr>
</tbody>
</table>

Some children registered with SCH have been using Dihydrocodeine for painful crises at home for several years. If they find this effective this can be continued, or it can be substituted for oral morphine as above.

**If still in pain with this treatment, (or pain already assessed as severe) treat as below ‘severe’.

**Severe pain and moderate pain already self medicated**

Do not use corticosteroids in the management of an uncomplicated acute painful sickle episode

Consider intranasal diamorphine as an immediate treatment for children over 10kg

**Dose:**

0.1mg/kg (100micrograms/kg) diamorphine hydrochloride.

Onset of action 5 minutes

Relatively short action (approx 2 hours)
**General Management of Children with Sickle Cell Disease including Pain and Infection**

**Administration:**

1. In a 1ml syringe, using a 5mg vial of diamorphine powder, make up to 0.5ml with 0.5ml of water for injection.
2. Discard the excess to leave 0.01ml/kg ie for a 15kg child 0.15ml (see table for examples).
3. Attach the syringe to the mucosal atomization device (MAD) firmly.
4. Spray the solution into the clearer nostril, on one side only if <0.2ml, if >0.2ml spray half the volume into each nostril.
5. The dose may be given by dripping the solution into the nostril(s) with the head tipped back, and the child sniffing, if no MAD is available.

**Note:** For small volumes it is worthwhile trapping a bubble of air between the plunger of the syringe and drug to act as a flush to clear the dead space of the atomizer. Speed of onset is usually around five minutes. The same cautions/contraindications apply to nasal administration as for other routes of opiate administration.

**Intranasal Diamorphine Dilution Chart**

Indication: Acute severe pain.

Dose: 0.1mg/kg (100 micrograms/kg) diamorphine intranasally

To prepare the solution dilute 5mg diamorphine in 0.5ml injection in a 1ml syringe (with 0.5ml water for injection) and administer the correct volume as per table below.

Discard the excess from the syringe prior to administration

<table>
<thead>
<tr>
<th>Weight (kg)</th>
<th>Administered volume (ml)</th>
<th>Dose of diamorphine (mg)</th>
</tr>
</thead>
<tbody>
<tr>
<td>10 kg</td>
<td>0.1mls</td>
<td>1mg</td>
</tr>
<tr>
<td>15 kg</td>
<td>0.15mls</td>
<td>1.5mg</td>
</tr>
<tr>
<td>20 kg</td>
<td>0.2mls</td>
<td>2mg</td>
</tr>
<tr>
<td>25 kg</td>
<td>0.25mls</td>
<td>2.5mg</td>
</tr>
<tr>
<td>30 kg</td>
<td>0.3mls</td>
<td>3mg</td>
</tr>
<tr>
<td>35 kg</td>
<td>0.35mls</td>
<td>3.5mg</td>
</tr>
<tr>
<td>40 kg</td>
<td>0.4mls</td>
<td>4mg</td>
</tr>
<tr>
<td>45 kg</td>
<td>0.45mls</td>
<td>4.5mg</td>
</tr>
<tr>
<td>50 kg</td>
<td>0.5mls</td>
<td>5mg</td>
</tr>
</tbody>
</table>

Record pain score before and after intervention.
General Management of Children with Sickle Cell Disease including Pain and Infection

Using intranasal diamorphine gives you the opportunity to site IV access and carry out necessary investigations with the child more comfortable. Plan to start an infusion before the intranasal dose wears off.

Alternatively start a morphine infusion after an initial slow IV bolus

Doses:

Morphine bolus 200microgram/kg (offer 2nd bolus after 30 minutes if still in severe pain on reassessment)

Morphine infusion (titrated to effect. For children <50kg, syringes are made up with 1mg/kg of morphine in 50ml of diluent. 1 ml/hr = 20microgram/kg/hr. For children >50kg, Morphine 50mg/50ml syringes are used and rate adjusted.

For how to write/make up a morphine infusion see the Haematology & Oncology Unit pain guidelines

Assess adequacy of pain relief regularly and frequently (every 30 minutes) and record on PEWS chart. Increase or reduce analgesia as appropriate. Continue until adequate relief has been achieved and then reassess at least every 4 hours thereafter. Monitor vital signs and oxygen saturation. Be alert for opiate induced narcosis and hypoventilation and the development of further complications such as infection or chest crisis.

For some patients with fluctuating pain Patient Controlled Analgesia (PCA) may be appropriate. Contact the Acute Pain Team to facilitate setting up the PCA, which should be used and monitored in accordance with the Trust PCA protocol.

All patients taking strong opioids should be assessed hourly for the first 6 hours and at least 4 hourly thereafter.

The time it takes from admission to control of pain is a topic suitable for audit

All patients on opioid analgesia should be prescribed laxatives regularly, anti emetics as needed and anti pruritics as needed.

If the child does not respond to analgesia as expected reassess for the possibility of an alternative diagnosis

Hydration

Almost all patients requiring admission for a painful crisis will need intravenous fluids to maintain an intake of 1.5 litres/m²/day (total of oral and IV) . Use dextrose 5% with Sodium Chloride 0.45%. Sickle patients are less able to excrete sodium so monitor electrolytes carefully – it may be necessary to switch to 5% dextrose at times. Continue until the patient is pain free and able to take the required amount orally.

Discuss the child with the Haematology SpR or Consultant.
Sometimes it is helpful to convert to oral morphine if fluid intake is improving, the pain is under control and the drip tissues. Doses are as in the Haematology & Oncology Unit Pain Guidelines.

Oral analgesia with paracetamol and diclofenac should be continued concurrently with IV opiate analgesia if the patient is able to tolerate oral medication or should be restarted as the morphine is weaned when the patient is able to take oral medication again.

3. Possible Acute complications

Acute chest crisis can follow painful crisis. This should be suspected if any of the following are present at any time:

- Abnormal respiratory signs or symptoms
- Chest pain
- Fever
- Hypoxia or an escalating oxygen requirement
- Acute Chest crisis is covered in a separate guideline.
- Other possible complications include acute stroke, aplastic crisis, infection including osteomyelitis and acute splenic sequestration. Guidance is available below and on the intranet via the Haematology & Oncology Unit guidelines

4. Discharge Information

Before discharge the child and carers must be given information about how to continue to manage the current episode at home:

- How to continue to wean down the analgesia
- How to obtain support if any new concerns
- How to obtain additional medication
- How to manage possible side effects (eg constipation)
5. **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 Prevenar 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 (first dose given at 2 years and then every 5 years for life). The infection risk is the highest for the SS genotype and in children up to the age of 5 yrs, but severe sepsis can occur in all ages and all genotypes.

Be alert to Salmonella osteomyelitis, pneumonia due to typical and atypical organisms and malaria – particularly in children arriving or returning from holiday in Africa.

Diagnosis can be difficult as a child with a simple acute painful episode may be febrile, with no overt site of infection. Young children may present with painful swollen joints or swelling in long bones and it can be hard to differentiate between bone infarction due to sickling and an osteomyelitis or septic arthritis.

All sickle cell disease patients who are pyrexial should be carefully assessed and have an infection screen including FBC and reticulocyte count, blood, urine and stool cultures 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 and may include ultrasound, xray and MRI (discuss with radiologist after alerting the haematology team) of a swollen bone or a painful joint. The patient may deteriorate very rapidly so it is important they are reviewed regularly and that the nursing staff are made aware of the potential for sudden worsening and the need to be vigilant. Remember that osteomyelitis is commoner in people with sickle cell disease than in the general population.

Once the child has been assessed and antibiotics started the case should be discussed with the haematology SpR or Consultant.

**Immunisation status and compliance with penicillin prophylaxis are topics which must be audited in order to comply with national quality standards**

6 **Jaundice**

Some children with SCD may be constantly mildly jaundiced due to the increased haemolysis that is part of the condition. Others only become jaundiced with intercurrent infections and painful crises (reflecting increased haemolysis). Management is of the underlying acute complication. Obviously if jaundice is severe, or the only presenting symptom, it may warrant investigation.
General Management of Children with Sickle Cell Disease including Pain and Infection

7 References

