Worcestershire Acute Hospitals NHS Trust

# Guideline for the acute management of sickle cell disease in children <18 years presenting to Riverbank Ward or the Children's Emergency Department at WRH (Full Version)

This guidance does not override the individual responsibility of health professionals to make appropriate decision according to the circumstances of the individual patient in consultation with the patient and /or carer. Health care professionals must be prepared to justify any deviation from this guidance.

## Introduction

Sickle cell disease (SCD) is an inherited haemoglobinopathy that is characterised by anaemia and a wide range of pathology secondary to intermittent small vessel occlusion. Children can present with chronic anaemia and/or hand foot syndrome (dactylitis) in the first few years of life. It may also be picked up during newborn screening. Children with SCD presenting acutely represent a unique challenge. Vaso-occlusive crisis (VOC) is the most common presentation but there is a much higher incidence of potentially life-threatening pulmonary, central nervous system (CNS), gastrointestinal (GI) and infectious complications in children.

The most common cause of death in children with SCD age <5 years are undetected splenic sequestration, acute anaemia and pneumococcal sepsis.

All patients with SCD have open access to Riverbank Ward. Patients who are not known to the local haematology team, or who are very unwell, may need to be seen in Children's ED.

## This guideline is for use by the following staff groups :

All healthcare professionals who are involved in assessing and managing patients <18 years old with Sickle Cell Disease on Riverbank ward or in the Emergency Department. Paediatric doctors, Paediatric nurses, Emergency department doctors, Emergency department nurses, Pharmacists.

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*This is the most current document and should be used until a revised version is in place*

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## 1.0 Key clinical assessments of the acutely unwell sickle cell patient

All children presenting to ED or PAU with sickle cell disease (SCD) should be triaged Amber and seen within 30 minutes of arrival.

**High risk are:**

- Children under 2 years (Low threshold for admission in children under 5 years)
- Acute sequestration (splenic or hepatic)
- Abnormal neurology
- Severe anaemia (>20g/L below baseline)
- Respiratory disease requiring O2 / any chest or respiratory signs
- Acute priapism > 2 hours
- Pyrexia > 38C

The most common death in SCD children under 5 years are undetected splenic sequestration/acute anaemia and pneumococcal sepsis.

**History**

• Note recent clinic letters, found on CLIP or Bluespier

• Symptoms – Pain, dyspnoea, fever, pallor, lethargy

• Focal symptoms e.g. chest, abdomen, jaundice, neurological

• Pain

o Site, severity (age appropriate pain score), recent pain relief dosage and times.

o Most patients will be able to tell you this is pain similar to previous episodes

o Provoking factors (fever, travel, procedures, recent hospital admission or transfusion)

• Past history and sickle-related complications. Particularly of chest crisis and management (PICU admission / ventilation).

• Ask if spleen normally enlarged.

• Usual sickle cell treatment – chronic transfusion programme, Hydroxycarbamide

• Compliance with penicillin/Immunisation history

• History of recent transfusion

• Travel history

• Co-morbidities – e.g. asthma

• Regard the patient (and/or their carer) as an expert in their condition, listen to their views and discuss with them the planned treatment regimen for the episode treatment received during previous episodes and any concerns they may have about the current episode

**Assessment**

• PEWS score

• Complete set of observations must include BP, pulse-oximetry and a pain score

• General examination including looking for pallor and jaundice

• Hydration status

• Signs of infection

• Full cardiovascular, respiratory and abdominal systems examination

  o Note spleen and liver size and compare to baseline

• Pain assessment with age appropriate pain scoring tool

• Record pain scores each time patient is reassessed (every 30 minutes until patient comfortable, then a minimum of 4 hourly)

• Assess all patients with sickle cell disease who present with acute pain to determine whether their pain is being caused by a vaso-occlusive crisis (VOC) or whether an alternative diagnosis is possible, particularly if pain is reported as atypical by the patient

• Full neurological examination (there is an increased risk of stroke during aplastic crisis)

**Investigations**

• FBC and reticulocyte count. Check the current Hb against the normal baseline of the patient. A fall of >20g/L is clinically significant.

• Blood group and save. Cross match if worsening anaemia.

• U+E, LFT and LDH. Amylase if abdominal pain.

• CRP

• Infection screen if febrile or signs of infection- blood/urine cultures, respiratory viral swab, blood gas and throat swab.

• If respiratory symptoms:

  o Combined nose and throat swab (respiratory viral screen)

  o CXR if respiratory symptoms particularly low oxygen saturations or chest pain.

  o Sputum culture if productive cough

  o Consider blood gas if oxygen saturations in air < 90%

• Erythrovirus screen (serology) in aplastic crisis

• Malaria screen if recent travel to a high-risk country. The gold standard test is three thick and thin blood films, with rapid diagnostic antigen testing. This is performed by the haematology lab. PCR can be performed if there is a high suspicion, following discussion with the malaria reference lab. If malaria is suspected, a Viral Haemorrhagic Fever risk assessment should always be performed, as the presentations can be similar.

## 1.1 Key points

SCD children are at risk of life threatening / emergency complications:

**Painful vaso-occlusive crisis (VOC)**

- The most common acute presentation in SCD
- Pain relief must be offered within 30 minutes of arrival
- Pain crisis can herald other complications (e.g., acute chest crisis, splenic sequestration, acute anaemia)
- Most patients rarely attend hospital and successfully manage their pain at home and will only attend when home options (e.g. NSAIDs/ paracetamol) have been ineffective
- High frequency users have more severe disease and have more complications

- SCD children can also present with pain due to common non-sickle problems

**Acute chest crisis (ACS)**

- Signs of acute chest crisis can be indistinguishable from a chest infection

**Acute anaemia**

- A fall in Hb of 20g/L from baseline is clinically significant anaemia
- Consider the cause: splenic sequestration, chest crisis, increased haemolysis and marrow red cell aplasia (usually erythrovovirus which will usually show a low reticulocyte count).

**Acute splenic enlargement**

- Can cause an acute fall in Hb

**Sepsis**

- SCD children are at risk of over-whelming sepsis due to functional hyposplenism
- Acute bacterial sepsis from encapsulated organisms including pneumococcal, salmonella, meningococcal, haemophilus
- Clinical signs may not be apparent

**Acute stroke**

- This is very rare but requires an emergency transfusion

**Priapism**

- Prolonged priapism (> 3 hours) is a urological emergency

**Dehydration**

- SCD children have reduced ability to concentrate their urine and so are more prone to dehydration.

**Osteomyelitis**

- If sickle cell patients are reporting of pain in joints (including their back) with a persistent fever, especially when unresponsive to IV antibiotics consider if there could be osteomyelitis and order an urgent x-ray. Refer to section 7.0 for further management.

## 2.0 Vaso-occlusive crisis

Vasso-occlusive crisis (VOC) are the most common complication of SCD. They can be precipitated by exposure to cold, infection, stress or dehydration. They may also rebound after incomplete resolution of a previous VOC episode. Most VOC symptoms can be managed at home with oral medications, and parents / patients are educated on how to do this. More severe pain requires clinical assessment and consideration of hospital admissions for parenteral opioids. The aim of VOC treatment is to minimise pain and prevent complications such as acute chest syndrome (ACS).

See **flowchart below** for initial management.

**Assessment**: See section 1.0 for key clinical assessments of the acutely unwell sickle cell patient.

**VOC should be treated as a medical emergency.**
**Analgesia should be administered within 30 minutes of arrival to hospital**

- Oral analgesia should be given prior to formal clinical assessment i.e. by the triage or admitting nurse
- Apply ametop for younger / anxious patients

**Think about possible acute complications:**
**Acute chest syndrome**
**Splenic sequestration**
**Sepsis**

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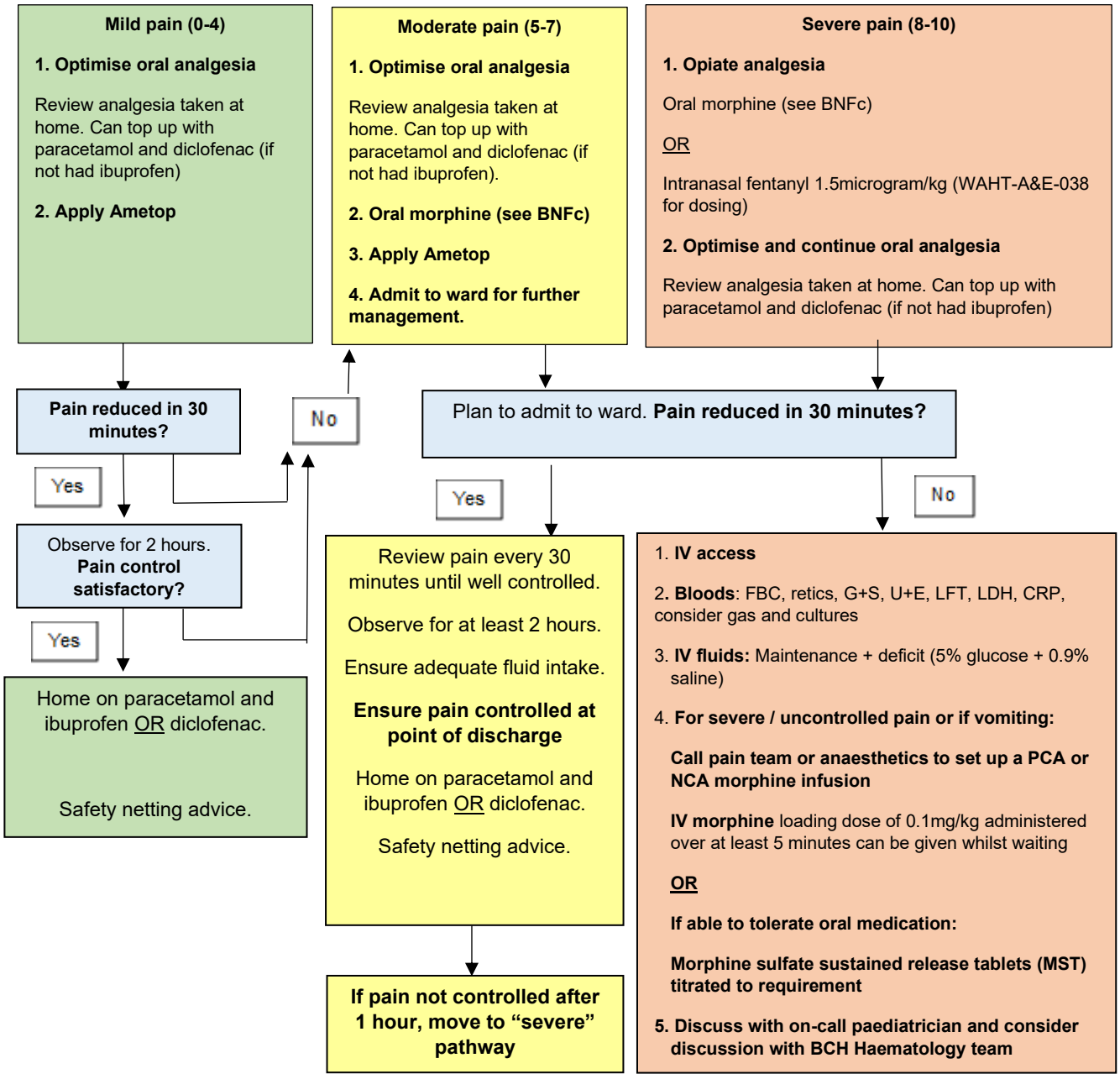
## Flowchart for the initial triage and management of Sickle Cell Disease patients <18 years presenting to PAU / CED with a vaso-occlusive crisis

**Triage / admitting nurse: If analgesia is required that you are unable to PGD, please hand over to nurse-in-charge to request prescription from a doctor. Analgesia should be given within 30 minutes of arrival.**

**All SCD patients seen in ED should be discussed with the paediatric consultant / registrar on call via bleep 676, even if you think they do not need admission.**




**If admitting the patient, please prescribe:**

1. Naloxone PRN if on IV morphine or large doses of oral morphine
2. Lactulose or Movicol/Laxido PRN
3. Ondansetron PRN
4. Continue REGULAR paracetamol and diclofenac/ibuprofen
5. Consider omeprazole for gastric protection
6. For breakthrough pain:

   Oral morphine (as per BNFc) 4 hourly PRN
7. Write up patient's usual folic acid and prophylactic antibiotics (usually Penicillin V).
8. Hydroxycarbamide to continue unless neutrophils <1.0 or platelets <100

## 2.1 Management of Vaso-occlusive (acute pain) crisis

1. **IV access**

   Avoid cannulation attempts of veins in legs, ankles and feet as this can lead to increased risk of venous thrombosis and leg ulceration.

2. **Bloods**

   Compare to patient's baseline. If there is a drop in Hb >20g/L, consider why this is. Blood transfusions should be used as sparingly as possible. Transfusion won't reduce the severity or duration of uncomplicated painful crisis. If concerned, please discuss with on-call paediatrician (or BCH Haematology team if admitted).

3. **IV fluids**

   Patients with SCD have reduced tubular concentrating ability. Ongoing fluid loss without adequate replacement causes reduction in plasma volume with increased viscosity and aggravation of sickling.

   Use IV fluids in children with severe pain, abdominal symptoms and inadequate oral intake.

   **Monitor fluid balance carefully.**

   Administer fluids at 100% maintenance plus correction for deficit unless at risk of acute chest syndrome. Over-hydration can cause fluid overload in patients with acute chest syndrome. For patients at risk of developing ACS (chest pain, chest infection, low O2 saturations), encourage oral fluids and maintain at 100% maintenance.

   Use maintenance fluids 0.9% saline + 5% glucose.

   Stop IV fluids as soon as patient is pain free and taking adequate oral fluids.

4. **Pain management**

   See flowchart.

   Pain in SCD is due to vaso-occlusion and may be severe. Medical and nursing staff often underestimate the severity of pain and therefore deal with it inadequately.

Severity may be difficult to assess, but if in doubt it's better to over-estimate and then reduce analgesia afterward. Aim to fully control pain as quickly as possible.

Use a pain scoring system to assess and monitor pain.

Pain relief must be offered within 30 minutes of arrival (as per NICE guidance).

Reassess pain every 30 minutes until pain improving.

If there is localised pain, consider further investigations (e.g. CXR for chest pain).

Offer all patients who are taking an opioid:

- Regular laxatives
- Anti-emetics PRN
- Antipruritic PRN

**Morphine**

| **Do NOT use MST and IV Morphine infusion together** |
|---|

Pain team (in hours) or anaesthetic registrar (bleep 700) can be contacted to set up a PCA (or consider nurse controlled in younger children) if pain is severe.

An IV bolus dose of 0.1mg/kg of morphine sulfate can be given whilst waiting. This will need to be administered slowly, over at least 5 minutes, to achieve adequate analgesia.

Breakthrough pain – prescribe Oral morphine (as per BNFc) every 4 hours PRN.

**Naloxone**

Must be prescribed as PRN alongside morphine. Use if concern about adequacy of respiration due to morphine use.

**Oral morphine for discharge/TTOs**

Do not give more than 3 days' supply on discharge.

Discuss and document indications for use and warning signs for overdose.

**See separate guideline WAHT-A&E-038**: Intranasal fentanyl for the management of acute pain in children

## 5. Antibiotics

**Please refer to Trust Paediatric antimicrobial guideline when prescribing antibiotics, especially for penicillin allergic patients**

Many VOC episodes are precipitated by acute infection.

If a child is unwell, fever >38, or a sequestration syndrome:

- Take blood cultures before starting antibiotics
- Look for a focus of infection
- Take urine culture
- Take throat and viral swabs – as atypical infections are more common, such as Mycoplasma
- If no clear focus: IV co-amoxiclav or ceftriaxone if clinically unwell
- If concerns about septic arthritis: ceftriaxone
- Caution when prescribing ceftriaxone in patients with sickle cell due to risk of immune haemolytic anaemia and biliary sludge

Chest infections should be treated with co-amoxiclav + clarithromycin, as atypical infections are often found in patients with SCD. Ask for a full respiratory viral panel, indicating that the child has SCD and is therefore immunocompromised.

Patients should be continued on their routine prophylactic Penicillin, unless started on a broad-spectrum antibiotic.

If in doubt, it's better to give broad spectrum antibiotics, especially in children < 5 years.

Patients on desferroxamine / deferiprone / deferasirox who have diarrhoea should be have these drugs stopped, due to risk of Yersinia infection. Send stool for culture – please ask specifically on the request form for Yersinia testing as it isn't done routinely. Discuss these cases with microbiology.

Ensure fever free for 24-48 hours prior to discharge, even if the focus for infection is clear. If continuing to spike fevers, consider re-culturing and stepping up antibiotics – discussion with microbiology may be useful.

## 3.0 Acute Anaemia

Steady state haemoglobin levels in children with SCD vary widely. This anaemia is exacerbated during periods of increased blood cell destruction, aplasia or acute sequestration. Spleen and liver size also need to be compared to steady state size. The spleen is usually palpable in children aged 1 to 3 years with SCD, and then typically becomes progressively smaller after that. Some children have persistent splenomegaly, which can be massive. Some children will have had splenectomies.

Four major types of crises are recognised in sickle cell anaemia:

• Aplastic Crisis

- Onset of profound anaemia over 1-3 days without sequestration.
- Acute fall in Hb (>20g/l beyond steady state level) AND reduced (<1%) or absent reticulocytes in the peripheral blood.
- Total WBC or PLT may or may not be affected.
- In addition, there is no significant increase in the unconjugated fraction of serum bilirubin.
- This is nearly always caused by Erythrovirus B19 infection, although other viruses may cause this to a lesser extent (e.g. influenza).
- Immunity appears to be lifelong.

• Acute sequestration crisis

- Acute fall in Hb accompanied by a rapidly enlarging spleen or liver (>2 cm above the steady state) and an elevated reticulocyte count
- Signs of acute circulatory insufficiency such as tachypnoea, tachycardia, and hypotension may or may not be present.
- Splenic sequestration typically occurs under 3 years of age and is often precipitated by infection

- Liver sequestration can occur at any age and treatment is similar to splenic sequestration. Rapid enlargement and shock are less common but deteriorating liver function and very rarely acute liver failure can occur.
- See section 5.0 for detailed management of sequestration crisis.

• Hyper-haemolytic crisis

- Acute fall in Hb associated with jaundice, marked reticulocytosis, and polychromasia on the blood film, increased hyperbilirubinaemia.
- Reticulocytopenia may also be present in hyperhaemolysis syndrome.
- Rarely acute renal failure may occur
- This is nearly always triggered by acute infection and may coincide with VOC
- It is important to determine if there has been a recent administration of a red cell transfusion in the previous 2 weeks as differential includes a delayed haemolytic transfusion reaction or hyperhaemolysis syndrome.
- Further transfusion can be life threatening and needs to be differentiated from a delayed haemolytic transfusion reaction.

• Vaso-occlusive crisis (VOC)

- See section 2.0 for detailed management of VOC.

### 3.1 Assessment

See section 1.0 for key clinical assessments of the acutely unwell sickle cell patient.

### 3.2 Management of aplastic crisis

*Discuss with on call consultant, and BCH Haematology team*

| **Shock present:** |
| --- |
| Resuscitate with IV fluids and emergency blood (O Rh D negative according to EPALS algorithm) |

If drop in Hb greater than 20 g/l from baseline, then transfuse packed red cells, usually maximum 15 ml/kg, see section 12.0 for more details.

**Shock not present:**

Send Erythrovirus (parvovirus) serology

Await FBC and cross-matched blood

Aim post transfusion Hb not greater than 100 g/L

If Erythrovirus infection is thought likely, the patient should be isolated in a side-room. Particular care should be taken to keep the patient isolated from patients who might be immunosuppressed. The patient should not receive nursing or medical care from staff who are, or might be, pregnant.

Interpretation of erythrovirus (parvovirus) serology:

IgM / IgG both negative. No evidence of recent erythrovirus infection. Susceptible to infection.

IgM positive / IgG negative. May be in keeping with recent erythrovirus infection but may also be a false positive IgM. Blood for erythrovirus viral load testing will be automatically referred by the laboratory.

IgM positive / IgG positive. Consistent with recent erythrovirus infection. Blood will be automatically referred to confirm by the laboratory.

IgM negative / IgG positive. No evidence of recent erthrovirus infection. Patient is immune to erythrovirus.

The family should be warned that other family members/contacts with haemolytic anaemias might also develop significant anaemia, and blood tests arranged if appropriate. Similarly, pregnant relatives should be warned to avoid the patient.

**Subsequent management**

Repeat FBC + reticulocytes daily

Treat complications e.g. Pain

### 3.3 Management of acute sequestration crisis

See section 5.0.

### 3.4 Management of hyper-haemolytic crisis

Discuss with on call paediatric consultant, **and** on call BCH Haematologist

**Shock present:**

Resuscitate with IV fluids and emergency blood (O Rh D negative according to EPALS algorithm)

**Shock not present:**

Await FBC and Cross matched blood

Blood bank should be informed if suspicion of delayed haemolytic transfusion reaction

Further treatment may involve observation, IV methylprednisolone 10mg/kg for 3 days, IV immunoglobulin 1gram/kg for 2 days, EPO and avoidance of red cell transfusion. Discuss with on-call BCH haematology team first.

**Subsequent management**

Repeat FBC, reticulocytes daily.

### 3.5 Management of vaso-occlusive crisis

See section 2.0.

## 4.0 Acute Chest Syndrome

Acute chest syndrome (ACS) is a leading cause of morbidity and mortality in SCD, and patients with frequent episodes of ACS and VIC have shorter lifespans. Severe episodes have also been associated with neurological damage.

Potential causes are infarction, infection and pulmonary fat embolism. Infectious causes are found in around 30% - most commonly chlamydia, mycoplasma and viruses.

**Risk factors**

- Over half of the episodes are preceded by admission with VOC episode. VOC involving the trunk cause splinting and decreased tidal volume are more likely to precipitate episodes.
- Post surgery / anaesthesia
- Asthma
- HbSS phenotype
- Low baseline Hb,
- Low Hb F%
- High WBC
- Can be worsened by fluid overload.

**Protective factors**

- Hydroxycarbamide
- Chronic transfusion programme
- Use of incentive spirometry has been shown to decrease incidence of ACS in patients hospitalised with VOC

### 4.1 Assessment

See section 1.0 for key clinical assessments of the acutely unwell sickle cell patient.

Typical presentation of ACS is:

A new infiltrate on CXR, excluding atelectasis, plus one or more of the following:

- Tachypnoea
- Fever > 38.5°C

- Chest pain – classically “T-shirt” distribution
- Cough - is a late symptom.
- Wheezing
- Hypoxaemia

CXR shows signs of lung consolidation – often bilateral and starting at the bases.

Physical signs may precede x-ray changes by up to 12 hours. However, chest x-ray changes can also precede signs. Consolidation in the upper or middle lobes without basal changes is suggestive of chest infection consistent with acute chest syndrome.

Falling Hb without evidence of splenic or hepatic sequestration is an indication for chest x-ray.

**Differential diagnosis in ACS**

Pulmonary embolism = Common symptoms are chest pain, dyspnoea and hypoxia. D-dimers are unhelpful in SCD as levels are usually elevated. If there is a high clinical suspicion of pulmonary embolism (i.e. sudden onset unilateral pleuritic pain that is not typical of sickle pain) treat for both conditions pending a computerized tomography pulmonary angiogram (CTPA). ACS may be complicated by pulmonary embolism or occur simultaneously.

Fluid overload = Careful attention to fluid balance with strict input and output monitoring is required. Acute deterioration in a patient after blood transfusion should prompt consideration of this complication or transfusion-related acute lung injury (TRALI).

Opiate toxicity = Monitoring of respiratory rate, sedation and pain scores should be in place, with opiate narcosis being associated with a falling respiratory rate. Dose modification or discontinuation may be necessary, and naloxone may be required if there is evidence of opiate toxicity. Opiate narcosis may trigger or worsen ACS.

Alveolar hypoventilation secondary to pain = optimising pain relief and encouraging incentive spirometry.

## 4.2 Management of Acute Chest Syndrome

**Patients with ACS can deteriorate rapidly and require close monitoring.**

Discuss with on call paediatric consultant, and on call BCH Haematologist.

Most patients will need transfer to BCH / PICU for exchange transfusion.

Contact KIDS / ITU registrar if patient is unstable / increasing oxygen requirement as ventilatory support is often needed.

**Deterioration is suggested by:**

• decreasing level of consciousness

• decreasing oxygen saturations in air.

• increasing oxygen requirements, or failure of oxygen to correct saturations.

• increasing tachypnoea.

• increasing pain.

• increasing shadowing on chest x-ray.

• falling haemoglobin, increasing white cell count.

**Respiratory:**

- Aim for sats 99-100%
- Airvo may be helpful, especially if splinting or low oxygen sats
- Encourage mobility
- Sit upright
- Chest physio if tolerated
- Nebulised bronchodilators
    - For active wheezing
    - Any patient with history of obstructive airway disease
    - As a trial for any patient with ACS, continue if clinical response

<u>Considerations in worsening Respiratory Failure – early discussion with KIDS team</u>

- Possibility of fluid overload
- Possibility of opioid toxicity
- Further top-up transfusion (see below)
- Exchange transfusion
- Conventional mechanical ventilation
- High-frequency oscillatory ventilation
- Nitric Oxide (NO) via mask or endotrachial tube

- Dexamethasone 0.3 mg/kg 12 hourly for a total of 4 doses; this may be helpful in rapidly deteriorating patients who seem likely to need ventilation. The steroids must not be stopped suddenly to avoid rebound pain

Further investigations:

- Daily CXR if lack of clinical improvement; more often if clinically worsening
- Daily FBC with differential and reticulocyte count to detect fall in Hb
- Microbiology for respiratory pathogens and consider serology for atypical organisms (e.g. urine for legionella and pneumococcus)

**Fluid management:**

- Strict input / output monitoring
- Maintain 100% maintenance fluids: Don't over-hydrate or rapidly hydrate as fluid overload may contribute to respiratory distress.
    - If patient is generally well apart from lung consolidation on CXR and is able to drink adequately, IV fluids may not be necessary
- Consider diuretics if fluid overloaded

**Pain management:**

- Optimize pain control to decrease chest splinting and prevent atelectasis
- Avoid hypoventilation from over-sedation
- Use NSAIDs alone or in conjunction with opioids
- Also see section 2.0 for management of painful VOC.

**Medication:**

- Anti-microbials:
    - Broad spectrum (IV ceftriaxone)
    - Plus macrolide (clarithromycin), IV if unable to tolerate oral
    - Consider Oseltamivir if Influenza positive
- Steroids:
    - Not routine therapy
    - Discuss with haematology on call at BCH
- Hydroxycarbamide:
    - Continue pre-existing hydroxycarbamide unless evidence of bone marrow suppression (Neutrophils <1, platelets <100)
- Bronchodilators

- There is no clinical evidence of bronchodilators in ACS. However, for those patients with a history of asthma and signs of bronchospasm, this could be considered

## 4.3 Transfusion Considerations in ACS

Transfuse if Hb falling or deteriorating clinical condition

Exchange transfusion preferred – discuss with consultant haematologist at BCH

Top up transfusion

Goal is to improve oxygenation and prevent progression to respiratory failure

- For patients with acute anemia (>20g/l fall from steady state)
- For patients who are symptomatic, but not in impending respiratory failure
- For patients with clinical or radiological progression
- Try not to exceed Hct of 30% or Hb >100g/dL post transfusion due to the risk of hyperviscosity

Exchange transfusion

- May be required in ACS
- This is usually done in Birmingham Children's Hospital, so early discussion with KIDS / Haematology team is essential
- See section 12.0 for more details.

**Complications of ACS**

- Rapid deterioration and death – patients should be monitored closely to allow the timely use of blood transfusions and PICU support.
- Neurological complications – seizures, silent cerebral infarcts, cerebral haemorrhage, strokes and posterior reversible leukoencephalopathy syndrome (PRES) are all common following severe ACS. Neurological complications are associated with hypertension and top-up transfusion increasing the

haematocrit to >0.35. If neurological symptoms develop, urgent neurological assessment and CT/MRI should be arranged.

- Chronic chest syndrome – repeated episodes of acute chest syndrome can result in a chronic, restrictive lung deficit. In general patients should have full pulmonary tests following recovery from an episode of ACS 6-8 weeks later. Oxygen saturations should be recorded when fully recovered in clinic, and it may be appropriate to organise overnight home monitoring of oxygen saturation levels.
- Children who are treated with dexamethasone may develop rebound symptoms such as acute pain if steroids are stopped suddenly. In general the steroids should be stopped gradually with reducing doses of oral prednisolone tailing-off over 5 days.

## 5.0 Splenic Sequestration

Splenic sequestration remains a leading cause of death in children with SCD. In most cases the first episodes in SCD patients occur between 3 months and 5 years but can occur in children of any age and are often precipitated by viral and bacterial infections. Recurrent splenic sequestration events are common, occurring in approximately 50 percent of those who survive the first episode, and the mortality rate in these patients can be high. Acute chest syndrome occurred in 20% in one series. It is caused by intra-splenic trapping of red cells which causes a precipitous fall in haemoglobin level and the potential for hypoxic shock.

**Defined as:**

- A Hb decrease of at least 20 g/L from the baseline
- Evidence of increased erythropoiesis such as an elevated reticulocyte count
- An acutely enlarging spleen:
    - Transfuse blood without delay (if in extremis uncross-matched O RhD negative)
    - Blood transfusion ideally by exchange transfusion – discuss appropriate treatment with consultant haematologist at BCH but don't delay top-up blood transfusion locally if in extremis
    - Do not infuse other fluids while waiting blood as this will exacerbate heart failure
    - Broad spectrum antibiotics to cover Pneumococcus and Haemophilus – co-amoxiclav or ceftriaxone.

### 5.1 Assessment

See section 1.0 for key clinical assessments of the acutely unwell sickle cell patient**.**

**Prompt recognition is essential**

Characterised by sudden onset of tachypnoea, pallor, tachycardia and abdominal pain and splenic enlargement. May be precipitated by fever, dehydration and hypoxia. Rapid sequestration of red cells leads to sudden anaemia and can result in death from hypoxic cardiac failure with pulmonary oedema.

**Investigations:**

**IV venous access:** Send bloods for urgent FBC, retics, urgent cross-match, blood culture, U&E, LFT, store serum for Virology.

## 5.2 Management of splenic sequestration

**Patients with severe splenic sequestration can deteriorate rapidly and die within hours.**

Discuss with on call paediatric consultant, and on call BCH Haematologist

**Initial management:**

- Correction of anaemia +/- hypovolemia with red cell transfusion.
- As severe disease can be fatal within a few hours, it is important to consider transfusion if the Hb has fallen by 20g/L from baseline.
- Acute shock present - resuscitate with intravenous fluids and emergency blood –
    - O RhD negative according to APLS algorithm.
- Acute shock absent - await results, cross-matched blood transfusion if the spleen is enlarging or Hb<50g/l, or Hb has fallen by 20g/L. Transfuse to increase Hb to about 80g/l.
- Start intravenous antibiotics – co-amoxiclav or ceftriaxone if severe. Antibiotics should be continued until the child is clinically better and may be adjusted depending on the results of blood cultures. Typically, the child will be discharged on their normal dose of penicillin prophylaxis, although a course of oral antibiotics may be given, depending on the clinical situation and the results of blood cultures.
- Treat other complications (e.g. acute chest syndrome section 4.0 or VOC section 2.0)

**Subsequent management**

- Monitor and record spleen size 12 hourly.
- Repeat FBC daily until stable, or more frequently if clinical deterioration.
- Ultrasound of the abdomen may be helpful but should not delay resuscitation and transfusion
- The rate of recurrent splenic sequestration is high
- Following red cell transfusion, the red cells sequestered in the spleen are remobilized and splenomegaly regresses. Haemoglobin level can increase, often to a level greater than predicted on the basis of the volume of red cells administered

## 6.0 Infections in SCD

All children with sickle cell disorders are at increased risk of infection. Underlying caused include hyposplenism, defects in opsonisation and in cell mediated immunity. The risk is highest for the HbSS genotype and in infants up to the age of 5. This is the time of particularly high risk for infection with encapsulated bacteria, *Haemophilus influenzae*, and pneumococcus. Patients should have prophylactic penicillin and additional Pneumovax at age of 2y and 5 yearly thereafter. Salmonella osteomyelitis, pneumonia due to typical and atypical organisms, and malaria, particularly in children returning from holidays in Africa, may also occur. The reason for the increased susceptibility to Salmonella osteomyelitis is not known. Other infections, such as urinary tract infection and acute cholecystitis, are common. Parvovirus B19 causes temporary red cell aplasia.

**Assessment**

See section 1.0 for key clinical assessments of the acutely unwell sickle cell patient.

It is common for a child with a simple acute painful episode to present with a fever and no obvious evidence of infection. Some young children present with painful, swollen joints or areas of swelling in a long bone. In these cases, it may be difficult to differentiate between acute bone infarction due to sickling and an osteomyelitis or septic arthritis.

- Infection screen (including blood cultures, urine cultures)
- Consider CXR, abdominal US, plain XR depending on symptoms
- Blood cultures should always be taken, and if there is a high level of suspicion of osteomyelitis (e.g. persistent high swinging fever, septic child, localized very tender swelling) further imaging should be done.

### 6.1 Management of infections in SCD

Prophylactic penicillin should always be continued in hospital if a different antibiotic is not prescribed to treat an acute infection.

IV Co-amoxiclav or cefotaxime are the first line broad spectrum antibiotics in the case of an unwell child with fever and no focus. If a localising infection is identified, then alternative antibiotics may be given – please refer to Paediatric Antimicrobial Guideline.

Local microbiology advice should always be sought in complex infections.

## 7.0 Osteomyelitis

### Background

- Patients with SCD are particularly susceptible to osteomyelitis
- Commonest organisms include Staphylococcus aureus and Gram negative bacilli (including *Salmonella*)
- Sickle cell vaso-occlusive crisis (VOC) can mimic osteomyelitis in the early stages as the presence of a hot tender joint can be found in VOC.

### 7.1 Assessment

See section 1.0 for key clinical assessments of the acutely unwell sickle cell patient

### 7.2 Diagnosis

- Largely clinical
- can be difficult to differentiate from vaso-occlusive crisis.
- Investigate if there are ongoing bony symptoms that do not settle after 1–2 days of standard VOC therapy
- Fever may be modest
- Positive cultures obtained from blood or bone obtained by aspiration or biopsy
- Findings on imaging are usually non-specific.

### Imaging for osteomyelitis

- Plain X-rays: early changes of osteomyelitis (periostitis and osteopenia) are also seen in VOC. Lucent areas seen later in course of infection
- Ultrasonography - periosteal elevation and subperiosteal fluid not specific. Fluid depths >4 mm highly associated with osteomyelitis.
- Radioisotope bone scanning and MRI may not differentiate infarction and infection but, in difficult cases, an opinion should be sought from a BCH radiologist who may have more experience of these situations

### 7.3 Treatment

Antibiotic choice

- Discuss with microbiology team
- IV Ceftriaxone if suspected septic arthritis
- Direct against any isolated organism

- Intravenous antibiotics should be given for at least 2 weeks. The patient can then finish the course with an oral antibiotic if there has been a good clinical response and serial CRP measurements show a downward trend.
- If there is no response to clinical parameters of infection - discuss with microbiologists (consider cover for Salmonella)

Malaria treatment

IV artesunate (or artemether with lumefantrine if less severe) for falciparum, chloroquine if non-falciparum – discuss with paediatric Infectious Diseases team if unsure.

**Surgery**

Consider surgical drainage if a collection is found on imaging. Be aware that swelling over long bones, muscles, and joint effusions are common with VOC. Decision for surgical drainage should be made in conjunction with the BCH haematology consultant.

## 8.0 Acute Stroke

11% of patients with sickle cell disease have a stroke by age 20 years. The peak age is 2-8 years. Risk depends upon genotype in Hb SS > Hb S-ß0 thalassemia > Hb S-ß+ thalassemia > Hb SC. It most commonly affects large arterial vessels.

Infarction is more prevalent than haemorrhagic stroke in the paediatric population.

Screening for primary stroke risk with transcranial Doppler (TCD) is now considered standard care for patients with Hb SS disease and HbS- ß0 thalassemia disease.

Primary stroke risk can be decreased with chronic transfusion therapy or hydroxycarbamide in patients with Hb SS disease and HbS- ß0 thalassemia disease that have repeated TCD velocities >200.

If left untreated 90% of children with sickle cell disease who have a stroke will have recurrent stroke. Patients with silent stroke have increased risk of subsequent infarctive stroke. About 20% of patients with sickle cell disease have silent stroke (2x the number who have an overt stroke).

**Risk factors:**

- Low baseline Hb
- High leukocyte count
- Prior TIA
- Frequency of acute chest syndrome
- Elevated blood pressure
- Stroke in sibling
- Dactylitis in first 2 years of life
- Absence of α-gene deletion

### 8.1 Assessment

See section 1.0 for key clinical assessments of the acutely unwell sickle cell patient.

The anterior cerebral circulation is most commonly involved, leading to headache, weakness, seizures, paraesthesiae, speech or behavioural abnormalities. Occasionally visual symptoms can occur in isolation and this should be borne in mind when assessing a patient.

Most children present with a classical weakness or dysphasia and are otherwise generally well. Less commonly, patients are clinically sick in association with a neurological event. This

should prompt additional investigation for other causes such as meningitis or encephalitis, fit or febrile convulsion, syncope, haemorrhage, cerebral malaria, trauma, fat embolism, hypoglycaemia, drugs, abscess, tumour.

**Examination**

- Thorough neurological exam including fundoscopy
- Do not forget vision, swallowing, speech, gait and conscious level (GCS)

**Investigations**

- Baseline observations including blood glucose
- Bloods – FBC (incl retic count and %HbS), Group and Save, U&Es, LFTs, CRP, coagulation screen, blood cultures and consider malaria screen
- Urgent neuro-imaging with CT scan to rule out acute bleed.
- Urgent ophthalmology referral if suspicion of retinal infarct
- MRI/MRA can be delayed until after treatment initiated.
- Other investigations to consider (if indicated) - Lumbar puncture, Viral serology - HSV, VZV, Urine & serum drug screen if altered mental status with no explanation

## 8.2 Management of acute stroke

**Discuss with on call paediatric consultant, and on call BCH Haematologist.**

Patients will need urgent transfer to BCH / PICU for exchange transfusion.

Discuss top up transfusion if delay and starting Hb allows

(Target Hct </= 36% or Hb </= 12 and Hb S < 30%)

- Obtain IV access
- Ensure patient Nil By Mouth
- Assess and secure airway (if drowsy).
- IV antibiotics if suspicion of sepsis.
- Treatment for elevated ICP if present
- Anti-convulsants if seizures are present
- Maintain oxygenation to keep sats >95%

## 9.0 Priapism

Defined as a sustained, painful, and unwanted erection usually unrelated to sexual activity and classified into:

- Stuttering. Recurrent episodes that last from minutes up to 3 hours. They may herald a prolonged event
- Prolonged. Longer than 3 hours

**Prolonged priapism is a urological emergency that requires urgent urologic intervention.**

Up to 90% of males with SCD will have experienced one or more episodes by the age of 20 years. May occur in children as young as 3 years. Due to vaso-occlusion, which causes obstruction of the venous drainage of the penis. Triggers include sexual arousal/ prolonged intercourse, fever, cold exposure, nocturnal tumescence (REM sleep), full bladder, dehydration. Alcohol, cocaine and testosterone have all been implicated. Recurrent or prolonged priapism can result in corpus cavernosal fibrosis and erectile dysfunction, the incidence of which is inversely correlated to the duration of priapism. Venous stasis leads to further acidosis and a vicious circle of sickling and inflammation, which if not broken may lead to fibrosis and erectile dysfunction.

### 9.1 Assessment

Patients and families are advised to attend the emergency department if an episode of priapism lasts for > 2 hours.

**History**

- Time of onset
- Frequency and duration of current and previous episodes.
- Triggers (see above).
- Response to previous therapy.
- Associated symptoms: dehydration, pain elsewhere, trauma

**Examination**

- Hydration status
- Size and degree of penile tumescence/turgor

- Bi-corporeal (corpus cavernosa only) or tri-corporeal (includes corpora and glans)

**Investigations**

- FBC, reticulocyte count, urinalysis +/- culture. Group and save.

## 9.2 Management of priapism

Prolonged priapism (>3 hours) is a urological emergency

Aim of therapy is:

- Pain relief
- Abort the erection (detumescence)
- Preservation of erectile function.

**Immediate discussion with on call BCH Haematologist**

**Referral to Paediatric Urology team if no relief within 3 hours of the onset of symptoms. Goal of urology consult is penile aspiration/irrigation with epinephrine.**

- **Keep NBM if surgical intervention required**
- **IV hydration (10ml/Kg bolus and 100-150% maintenance )**
- **Analgesia (careful use of opioids to avoid urinary retention)**
- **Supplemental O2 to keep Sats > 95%**
- **Never use ice packs**
- **DO NOT TRANSFUSE RED CELLS IN THE ACUTE SITUATION**

**Urological management**

Surgical Intervention should not be delayed by either medical treatment or the provision of blood products

- Irrigate with epinephrine solution until detumescence.
- May need to repeat 3-4 times.
- Admit for observation.
- Without detumescence after 24hours of medical therapy (supportive care, aspiration, irrigation and transfusion), surgical intervention/shunting may need to be performed.
- Winter Shunt: a shunt is created between the corpus cavernosa and the glans penis allowing blood to drain into the uninvolved corpus spongiosa. Usually performed by

inserting needles longitudinally through the glans into the corpus cavernosa, thus creating a fenestration in the fibrous albuginea

- If percutaneous shunting fails, open shunts between the corpus cavernosa and the corpus spongiosum, dorsal vein or saphenous vein have been described.
- Complications of surgical intervention include infection, stricture, fistula and high risk of impotence.
- If impotence persists for 12 months, the patient may be referred to the Urology team for consideration of implantation of a semi-rigid penile prosthesis.

**Transfusion**

Top up or exchange transfusion may be appropriate only if:

- Failure of repeated aspiration
- Proceeding to a surgical shunt procedure.

There has been a reported association of SCA, priapism, exchange transfusion and neurologic events (headache, seizure, impairment of consciousness, or stroke) called ASPEN syndrome. Therefore, observe for neurological symptoms post-exchange transfusion.

**Inpatient Discharge Criteria**

- Able to void.
- Penis more flaccid (swelling and/or oedema may be present for several weeks)
- Pain controllable on oral analgesia.
- Consider addition of pseudoephedrine.

**Home Treatment (advice for families/patients)**

Families are advised to follow the following measures in order to prevent prolonged

priapism.

- Increased fluids
- Urination
- Oral pain medications
- Warm baths / soaks
- Gentle exercise
- Oral pseudoephedrine for patients with previous history of priapism.

**Recurrent Priapism**

- Patient/family education. Lack of awareness of this sickle related complication and embarrassment to discuss an acute event represent leading to under-recognition and under-treatment and are a significant hurdle to appropriate care. In order to raise awareness and decrease anxiety or embarrassment of the subject, priapism must be discussed at each outpatient visit. Boys and their parents must be educated that prolonged priapism is a urological emergency. If untreated priapism can result in impotence.
- Referral to Paediatric Urology team
- Recurrence can be prevented by the use of pseudoephedrine
- If recurrent and/or severe: short term transfusion therapy (6-12 months) has been recommended.

## 10.0 Acute abdomen

**IF AN OPERATION IS REQUIRED:**

Liaise with the haematology team at BCH promptly.

Pre-operative transfusion may be required.

Sickle pain episodes may present with pain in the abdomen and intra-abdominal pathology can precipitate pain crisis. This population has the usual problems which can present with abdominal pain, however, the incidence of gall stones with cholecystitis, peptic ulcer disease, and pyelonephritis is increased. Complications such as splenic or hepatic sequestration should always be considered. When in doubt admission for observation is almost mandatory. Possible causes of abdominal pain include (not exhaustive list):

**Common**

- Constipation
- Vaso-occlusive pain crisis (abdominal painful crisis, girdle syndrome): Pain is usually diffuse, steady, involves back and extremities, and is characteristic for patient. Bowel sounds can be present or absent. Diffuse tenderness without guarding or rebound is present. Liver and spleen size is unchanged. See section 2.0.
- Cholecystitis - see section 3.13
- Gastroenteritis
- Urinary tract infection
- Pneumonia (referred pain)

**Less Common**

- Splenic sequestration – see section 5.0
- Hepatic sequestration
- Acute pancreatitis
- Acute appendicitis
- Acute chest syndrome (referred pain)
- Gastritis/peptic ulcer disease

**Rare**

- Hepatitis
- Ischaemic colitis

### 10.1 Assessment

See section 1.0 for key clinical assessments of the acutely unwell sickle cell patient.

**History**

Define the onset, whether continuous, intermittent, new or recurrent, location, radiation, character (diffuse, localized, crampy, sharp, dull aching), duration, aggravating/ alleviating factors, nausea, vomiting, diarrohea, constipation, change in urine/stool colour, melena, hematemesis, increased jaundice, anorexia, distension, last menstrual period (date, duration, normality), illness in contacts.

Past Medical History. Document haemoglobin phenotype, general health, past surgery (appendectomy, cholecystectomy, splenectomy) dates, recent hospitalizations, present medications, drug allergies.

Systemic Symptoms. Define fever, weight change, urinary frequency, urgency, dysuria, sexual history, vaginal discharge, past menstrual problems.

**Investigations**

- FBC and retic count, U&Es, LFTs, amylase
- Urinalysis and culture
- Consider - pregnancy test, CXR, AXR, Abdo USS

**Patient and Parent Education**

The most important consideration in abdominal pain is that splenic sequestration be diagnosed early as above. Patients should also be taught to present for evaluation if they develop unusual abdominal pain, urinary or gynaecological symptoms. Patients with known gall stones need to be evaluated for changes in pain or jaundice.

## 11.0 Renal disease

The kidney in SCD exhibits numerous structural and functional abnormalities along the entire nephron. The environment of the renal medulla is characterized by hypoxia and acidosis which both promote HbS polymerisation and sickling. This area is especially susceptible to malfunction. The spectrum of sickle nephropathy ranges from hyposthenuria (poor concentration), haematuria, proteinuria, acute renal failure, infections and progressive loss of function to end stage renal disease (ESRD).

**Investigations**

- Ultrasound scan is the first line of investigation.
- MSU for microscopy, culture and sensitivity, and cytology.
- Intravenous urography may be necessary in some cases but request a renal opinion first as pre-hydration and acetylcysteine may be necessary.
- Treatment is with bed rest, maintenance of a high urinary output and iron replacement.
- Consider malignancy as a cause

### 11.1 Acute renal failure

May be precipitated by infection, dehydration, drugs or occur in the context of multi-organ failure. These patients must be managed jointly with the renal physicians.

**Investigation and management**

- evaluation of medications
- MSU
- urine dipstick
- abdominal ultrasound to check kidney size and look for obstructive causes of renal failure
- Management includes meticulous fluid balance.

### 11.2 Haematuria

- Microscopic haematuria is common

- Frank haematuria is due to renal papillary necrosis and ischaemic necrosis in the renal medulla. Occasionally it can be severe and can cause renal colic and ureteric blockage.
- Usually affects the left kidney.
- Can also occur in patients with sickle trait
- Infection can complicate papillary necrosis
- Consider urolithiasis
- Liaise with urologists

## 11.3 Urinary tract infections

- Common, particularly in females with SCD and especially during pregnancy.
- Should be vigorously treated to prevent serious deterioration in renal function.
- Antibiotics: avoid tetracyclines; monitor renal function. Antibiotic choice as per hospital policy
- Haematuria, secondary to papillary necrosis, can precipitate UTI, but other factors must be excluded.

## 11.4 Proteinuria

- 40% of Hb SS patients with nephrotic syndrome can develop end stage renal failure.
- Can be associated with a very rapid decline in renal function
- Screening should be annually with a urine dipstick:
    - If screen abnormal then request urinary protein:creatinine ratio and urine microalbumin (this measures the albumin:creatinine ratio)
- Referral to renal physician specialising in sickle cell disease for further investigations
- ACE inhibitors sometimes can be effective

## 11.5 Medullary carcinoma

- Increased frequency reported in patients with haemoglobinopathy - especially HbSC and sickle cell trait
- Presents with loin pain, haematuria, weight loss, and possibly a palpable renal mass.
- The disease is aggressive and is usually metastasized at presentation.
- Response to chemotherapy and radiotherapy is poor.

## 11.6 Chronic renal failure

- Occurs in around 4% of patients.
- A variety of pathologies have been described.
- Annual assessment: renal function, urine dipstick, BP measurement

**Investigations**

- Urea and electrolytes, creatinine with eGFR, calcium, phosphate, bicarbonate, liver function tests, blood glucose, lipids, urate.
- Renal immunology profile (CRP, ANA, ANCA, Protein electrophoresis, immunoglobulins and complement C3/C4) but only if there are persistent urinary abnormalities on dipstick testing.
- Full blood count and reticulocytes
- Urine for microscopy and culture
- Spot urine for urinary albumin/creatinine ratio.
- Ultrasound of kidneys and urinary tract

**Management**

- All patients should be referred to the renal team for evaluation
- Rigorous blood pressure control
- Prompt treatment of UTIs necessary
- Avoid long term NSAID
- Consider erythropoietin (eGFR <60ml/min and Hb <6.5g/dl and/or retics <150)
- Renal replacement therapy
- Renal transplantation may be considered

## 12.0 Transfusion in SCD

### General Principles:

- Blood transfusion should only be used for specific indications. There must be a clear indication for every transfusion
- Consider the cause carefully of any acute fall in Hb - splenic enlargement, acute chest syndrome, haemolytic episode, parvovirus
- Blood transfusion should be used as sparingly as possible.
- Transfusion are an essential and life-saving therapy for some acute complications.
- There is an increased risk of delayed haemolytic transfusion reactions in sickle cell disease. These can mimic sickle painful crisis and clinicians should have a high index of suspicion for investigating for the development of antibodies when painful episodes develop in the post-transfusion period.
- Hyperhaemolysis has also been described post-transfusion without the development of antibodies.
- Avoid hyperviscosity by ensuring final haematocrit < 0.35.
- Give sickle negative, CMV negative, phenotypically matched blood, always liaise with blood bank and indicate patient has sickle cell disease.
- Emergency exchange transfusion is rarely needed except in acute stroke or deteriorating acute chest syndrome
- Transfusion will NOT reduce the severity or duration of uncomplicated painful crises.
- Discuss blood transfusion prior to administration with on call consultant and/or BCH haematologist on-call (do not delay in emergency treatment for shock in splenic sequestration)

### Investigations:

For all transfusions in patients with sickle cell disease it is important to record the following information before and after the procedure:

- Hb, WCC, Platelets
- Hct and %HbS
- U&Es, Ca and LFTs

## 12.1 Top-up transfusion

**Indications:**

When a patient with SCD drops their Hb sufficient to cause, or risk, clinical compromise such as cardiac failure and hypovolaemic shock. It is rarely necessary to transfuse if the Hb is over 60 g/l, or <20g/l below steady state unless there is reticulocytopenia associated with the falling Hb or clinical evidence that the fall in Hb will continue.

Top up transfusion may also be indicated in patients with moderate to severe crises, where exchange transfusion is not felt necessary.

These circumstances may arise with:

- Splenic sequestration
- Hepatic sequestration
- Acute chest syndrome
- Mesenteric syndrome
- Aplastic crises (Erythrovirus B19)
- Blood loss
- Haemolytic crises

Other possible indications:

- Pre-operative
- Pre-flying

**Aims:**

To improve oxygen carrying capacity.

To dilute sickle cells and improve blood viscosity and flow.

Raise Hb to patient's steady state level.

Do not raise Hb > 100g/dl (or Hct > 0.35) so as to avoid increasing blood viscosity.

**Practical Points:**

**Volume of blood to be transfused (mls) = (desired Hb [g/L]– current Hb [g/L]) x 0.4 x weight (kg)**

**Transfuse slowly in the absence of shock.**

**Blood products:**

Should be ABO and Rh compatible, CMV negative and sickle negative.

**Complications of blood transfusion in sickle cell disease**

- **Hyperviscosity:** This may result in poor cerebral function, seizures and a worsening clinical condition.
  Check the Hct is <0.35.

- **Metabolic disturbance**: These include hyperkalaemia, hypocalcaemia, citrate toxicity, hypernatraemia, hypo- or hyperglycaemia and late onset alkalosis.
  Be aware of these complications and check biochemical profile after the procedure and correct as necessary.

- **Delayed haemolytic transfusion reactions**: can mimic episodes of VOC – high index of suspicion if patient has recently had a transfusion.

## 12.2 Exchange transfusion

**Practical Points:**

**Liaise with BCH haematology consultant +/- KIDS team early if you think an exchange transfusion may be necessary.**

**Indications:**

Exchange transfusion is generally reserved for the treatment of life or organ-threatening and other major complications of SCD. Exchange transfusions will be perfomed at Birmingham

Children's Hospital, so prompt discussion with the Haematology team and KIDS will be required to facilitate timely transfer.

Goal is to remove sickled cells (Hb S) and replace with Hb A .

**Indications:**

- For impending or actual respiratory failure
- For marked clinical deterioration with progressive radiological findings despite a simple top-up transfusion
- For patients with Hb >100g/dl or hct >30% to avoid the increased viscosity of a top-up transfusion
- Severe acute chest syndrome
- Mesenteric syndrome
- CVA
- Splenic sequestration
- Hepatic sequestration
- Fulminant hepatic failure
- Priapism unresponsive to therapy

## Monitoring

| Page/ Section of Key Document | Key control: | Checks to be carried out to confirm compliance with the Policy: | How often the check will be carried out: | Responsible for carrying out the check: | Results of check reported to: *(Responsible for also ensuring actions are developed to address any areas of non-compliance)* | Frequency of reporting: |
|---|---|---|---|---|---|---|
| | **WHAT?** | **HOW?** | **WHEN?** | **WHO?** | **WHERE?** | **WHEN?** |
| | These are the 'key' parts of the process that we are relying on to manage risk. We may not be able to monitor every part of the process, but we MUST monitor the key elements, otherwise we won't know whether we are keeping patients, visitors and/or staff safe. | What are we going to do to make sure the key parts of the process we have identified are being followed? (Some techniques to consider are; audits, spot-checks, analysis of incident trends, monitoring of attendance at training.) | Be realistic. Set achievable frequencies. Use terms such as '10 times a year' instead of 'monthly'. | Who is responsible for the check? Is it listed in the 'duties' section of the Policy? Is it in the job description? | Who will receive the monitoring results? Where this is a committee the committee's specific responsibility for monitoring the process must be described within its terms of reference. | Use terms such as '10 times a year' instead of 'monthly'. |
| | Assessment and administration of analgesia within 30 minutes of arrival | Audit | Every 3 years | Paediatric Haematology Team | Paediatric Audit Meeting | Every 3 years |

## References

*Guidelines for Management of Sickle Cell Disease* Birmingham Children's Hospital (updated 2024)

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Stroke in Childhood: Clinical guideline for diagnosis, management and rehabilitation. Published: May 2017. https://www.rcpch.ac.uk/sites/default/files/2021-02/Stroke%20guideline%2008.04.19%20updated%202021.pdf. [Accessed: 1st March 2024]

WAHT-A&E-038: Intranasal fentanyl for the management of acute pain in children within the emergency department. 2024.

## Contribution List

### Contribution List

This key document has been circulated to the following individuals for consultation;

| Designation |
|---|
| Paediatric consultants |
| Emergency department paediatric lead team |
| Paediatric educators |
| Pain team |
| Antimicrobial pharmacist |
| Paediatric pharmacist |

This key document has been circulated to the chair(s) of the following committee's / groups for comments;

| Committee |
|---|
| Medicines Safety Committee |
| Paediatric Governance Meeting |

Worcestershire Acute Hospitals NHS Trust

## Supporting Document 1 - Equality Impact Assessment Tool

## Equality and Health Inequalities Impact Assessment (EHIA) Tool

**Herefordshire & Worcestershire STP - Equality and Health Inequalities Impact Assessment (HEIA) Form**
**Please read HEIA guidelines when completing this form**

**Section 1 - Name of Organisation** (please tick)

| | | | | | |
|---|---|---|---|---|---|
| Herefordshire & Worcestershire STP | | Herefordshire Council | | Herefordshire CCG | |
| Worcestershire Acute Hospitals NHS Trust | X | Worcestershire County Council | | Worcestershire CCGs | |
| Worcestershire Health and Care NHS Trust | | Wye Valley NHS Trust | | Other (please state) | |

| | |
|---|---|
| **Name of Lead for Activity** | **Dr Charlotte Cragg** |

| | | | |
|---|---|---|---|
| **Details of individuals completing this assessment** | **Name** | **Job title** | **e-mail contact** |
| | **Dr Charlotte Cragg** | **Specialist Doctor** | **Charlotte.cragg2@nhs.net** |
| | | | |
| | | | |
| **Date assessment completed** | **04/06/2026** | | |

**Section 2**

| | | | | |
|---|---|---|---|---|
| Activity being assessed (e.g. policy/procedure, document, service redesign, policy, strategy etc.) | **Title: Guideline** | | | |
| What is the aim, purpose and/or intended outcomes of this Activity? | | | | |
| Who will be affected by the development & implementation of this activity? | ❑<br>☑<br>❑ | Service User<br>Patient<br>Carers<br>Visitors | ☑<br>❑<br>❑<br>❑ | Staff<br>Communities<br>Other _____ |
| Is this: | ☑Review of an existing activity<br>❑ New activity<br>❑ Planning to withdraw or reduce a service, activity or presence? | | | |

| | |
|---|---|
| What information and evidence have you reviewed to help inform this assessment? (Please name sources, eg demographic information for patients / services / staff groups affected, complaints etc. | |
| Summary of engagement or consultation undertaken (e.g. who and how have you engaged with, or why do you believe this is not required) | |
| Summary of relevant findings | |

## Section 3

Please consider the potential impact of this activity (during development & implementation) on each of the equality groups outlined below. **Please tick one or more impact box below for each Equality Group and explain your rationale**. Please note it is possible for the potential impact to be both positive and negative within the same equality group and this should be recorded. Remember to consider the impact on e.g. staff, public, patients, carers etc. in these equality groups.

| Equality Group | Potential positive impact | Potential neutral impact | Potential negative impact | Please explain your reasons for any potential positive, neutral or negative impact identified |
|---|---|---|---|---|
| **Age** | | x | | |
| **Disability** | | x | | |
| **Gender Reassignment** | | x | | |
| **Marriage & Civil Partnerships** | | x | | |
| **Pregnancy & Maternity** | | x | | |
| **Race including Traveling Communities** | | x | | |
| **Religion & Belief** | | x | | |
| **Sex** | | x | | |
| **Sexual Orientation** | | x | | |
| **Other Vulnerable and Disadvantaged Groups** (e.g. carers; | | x | | |

| Equality Group | Potential positive impact | Potential neutral impact | Potential negative impact | Please explain your reasons for any potential positive, neutral or negative impact identified |
|---|---|---|---|---|
| care leavers; homeless; Social/Economic deprivation, travelling communities etc.) | | | | |
| **Health Inequalities** (any preventable, unfair & unjust differences in health status between groups, populations or individuals that arise from the unequal distribution of social, environmental & economic conditions within societies) | | x | | |

**Section 4**

| What actions will you take to mitigate any potential negative impacts? | Risk identified | Actions required to reduce / eliminate negative impact | Who will lead on the action? | Timeframe |
|---|---|---|---|---|
| | | . | | |
| | | | | |
| | | | | |
| **How will you monitor these actions?** | | | | |
| **When will you review this EIA?** (e.g in a service redesign, this EIA should be revisited regularly throughout the design & implementation) | | | | |

**Section 5** - Please read and agree to the following Equality Statement

**1. Equality Statement**

1.1. All public bodies have a statutory duty under the Equality Act 2010 to set out arrangements to assess and consult on how their policies and functions impact on the 9 protected characteristics: Age; Disability; Gender Reassignment; Marriage & Civil Partnership; Pregnancy & Maternity; Race; Religion & Belief; Sex; Sexual Orientation

1.2. Our Organisations will challenge discrimination, promote equality, respect human rights, and aims to design and implement services, policies and measures that meet the diverse needs of our service, and population, ensuring that none are placed at a disadvantage over others.

1.3. All staff are expected to deliver services and provide services and care in a manner which respects the individuality of service users, patients, carer's etc, and as such treat them and members of the workforce respectfully, paying due regard to the 9 protected characteristics.

| | |
|---|---|
| **Signature of person completing EIA** | C Cragg |
| **Date signed** | 4/6/26 |
| **Comments:** | |
| **Signature of person the Leader Person for this activity** | |
| **Date signed** | |
| **Comments:** | |

## Supporting Document 2 – Financial Impact Assessment

To be completed by the key document author and attached to key document when submitted to the appropriate committee for consideration and approval.

| | Title of document: | Yes/No |
|---|---|---|
| **1.** | Does the implementation of this document require any additional Capital resources | No |
| **2.** | Does the implementation of this document require additional revenue | No |
| **3.** | Does the implementation of this document require additional manpower | No |
| **4.** | Does the implementation of this document release any manpower costs through a change in practice | No |
| **5.** | Are there additional staff training costs associated with implementing this document which cannot be delivered through current training programmes or allocated training times for staff | No |
| | Other comments: | |

If the response to any of the above is yes, please complete a business case and which is signed by your Finance Manager and Directorate Manager for consideration by the Accountable Director before progressing to the relevant committee for approval.