Sickle Cell Disease and Thalassaemia:
Acute Complications of Sickle Cell Disease

Profile	
Version:	*V3.0*
Author:	*Dr Elizabeth Rhodes, Consultant Haematologist* *Dr Julia Sikorska, Consultant Haematologist*
Executive/Divisional sponsor:	*Dr Lisa Pickering, Divisional Chair*
Applies to:	*All staff involved in the care of patients with Sickle Cell Disease*
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Approval	
Approval person/Committee:	*MedCard Divisional Governance Board*
Date:	*14th February 2019*

Contents

Policy Gateway

Please complete the checklist and tables below to provide assurance around the policy review process.

☒ I have involved everyone who should be consulted about this policy/guidance

☒ I have identified the target audience for this policy/guidance

☒ I have completed the correct template fully and properly

☒ I have identified the correct approval route for this policy/guidance

☒ I have saved a word version of this policy/guidance for future reviews and reference

Please set out what makes you an appropriate person to conduct this review:
Lead consultant for adult haemoglobinopathy service

Please set out the legislation, guidance and best practice you consulted for this review:

- West Midlands Review Service – Quality Standards: Health Services for People with Haemoglobin Disorders (2018/19)
- Standards for the Clinical Care ofAdults with Sickle Cell Disease in the UK 2018 (Sickle Cell Society) https://www.sicklecellsociety.org/wp-content/uploads/2018/04/Web-version-FINAL-SCS-Standards-GSM-6.4.18.pdf
- British Society Haematology : guidelines Red Cell Transfusion in Sickle Cell Disease Part I and Part II (7.11.17 and 18.11.18) https://b-s-h.org.uk/guidelines/guidelines/red-cell-transfusion-in-sickle-cell-disease-part-I/
- NICE guidelines for Sickle Cell Acute Painful Episodes (CG143) (2012) https://www.nice.org.uk/guidance/cg143
- BCSH guidelines for Acute Chest Syndrome (2015) http://www.bcshguidelines.com/documents/Acute_Chest_Crisis.pdf

Please identify the key people you involved in reviewing this policy why, and when:
Dr Mark Haden (February 2019) – consultant physician in Emergency Department (link consultant for haematology) Dr Anthony Preira (2015) – link consultant for neurology and stroke Dr Joyce Popoola (2015) – link consultant for renal and nephrology Mr Nick Watt-Coote (2015) – link consultant for urology Miss Cathy Egan (2015) – link consultant for ophthalmology

Summarise the key changes you have made and why:
Minor updates in line with the UK standards of care for SCD Updated contact details

Executive Summary

Patients with Sickle Cell Disease (SCD) are at risk of emergency presentations and admissions with an unpredictable nature. This is predominantly due to painful crises which need prompt and appropriate management. However they are also at risk of life threatening complications.

This guideline contains up to date advice based on both evidence and expert opinion to support the management of these patients.

1. Introduction

Sickle cell disease (SCD) is a life-long condition characterised by chronic red cell haemolysis leading to multi-organ dysfunction. Patients may also suffer from acute events often referred to as crises. At St George's, there are over 200 admissions a year for sickle cell disease related problems.

The most common cause of an acute crisis is a painful veno-occlusive crisis ("bony crisis") which causes patients to present with severe, unrelenting pain which requires **prompt and affective pain relief** (usually opioid) and regular review. It is important always to consider non-sickle causes for the pain.

There are Emergency Department (ED) and "Grey Book" guidelines for the emergency management of these patients which reflect the 2012 NICE guidelines and these are available on the intranet. Many patients who present regularly to St George's will have their own individual care plan (or protocol or proforma) available in a paper folder in the ED. These protocols are also uploaded on the EPR electronic documents section. Please refer to the most recent document.

These guidelines discuss the management, not only of an acute painful crisis, but also of other sickle cell related emergencies. These should be used in conjunction with the ED guidelines and Grey Book.

All patients presenting with significant chest signs, neurological signs, priapism or shock should be discussed urgently with the haematology SpR for sickle cell (bleep 7080) or, if out of hours, the on call haematology SpR (via switchboard.)

2. Status and Purpose

This document is part of the Haematology Department's guidelines on the management of patients with SCD and is applicable to all staff involved in the care of these patients.

3. Definitions

Sickle Cell Disease – inherited life long condition due to abnormal haemoglobin varient

4. Scope

This guideline is relevant to the care of patients with SCD presenting to the Emergency Department at St George's site and being admitted to a ward at St George's

5. Roles and Responsibilities

5.1 Haemoglobinopathy team (Consultant haematologists, Clinical Nurse Specialist and Clinical Health Psychologist) – Responsible for the care of these patients, developing and updating guidelines to be reflective of good practice and to deliver the training to ensure good safe care

5.2 Medical staff involved in the care of patients with SCD. Responsible with the oversight of the haemoglobinopathy team to deliver the care of these patients in line with guidelines where possible.

5.3 Nursing staff and allied health professionals involved in the care of patients with SCD on wards, day unit and other areas of St George's Responsible with the oversight of the haemoglobinopathy team to deliver the care of these patients in line with guidelines where possible.

6. Content

6.1 Painful Veno-Occlusive Crises (VOC)

Painful bony crises can occur with or without a trigger.

Pain in a VOC is due to tissue (bone) hypoxia, which is a consequence of the rigidity of the red blood cells in these patients. These crises can occur without warning but well recognised triggers include cold weather, infection, hypoxia, dehydration, pregnancy and menstruation and these should be considered and asked about.

The frequency and severity of pain crises can and does vary between patients.

The mainstay of treatment is supportive care with prompt and adequate analgesia, hydration, warmth, oxygen and close monitoring for other complications.

On presentation to the ED the patients should be treated along the ED guidelines **as below**

- Observations should be taken
- Pain level assessed and documented and triaged as per ED guidelines
    - Severe pain (i.e. pain score 7 or above), triage as **RED** and patients should be managed in resus for parenteral opiates
    - Moderate pain (i.e. pain score 4-6) despite taking analgesia prior to arrival, triage as **ORANGE** and managed in resus for parenteral opiates
    - Moderate pain without having taken any analgesia, triage as **YELLOW** and managed in majors
- **Appropriate** analgesia within **30 minutes** of arriving at the ED – using individual protocol if available or by the ED guidelines for morphine dosing.
    - Appropriate analgesia means taking into account guidelines, individual protocols, the severity of the pain, triage as above and what the patient has already taken to try and manage the pain before attending the ED. Analgaesia decisions should be discussed with patients where appropriate
    - If individual protocol available then prescribe analgesia as per protocol
    - If no individual protocol available then:
        - Red or Orange triage: Prescribe morphine 5-10mg s/c if acute painful sickle
        - Yellow Triage: prescribe 30-60mg Dihydrocodeine or codeine
    - For all patients prescribe, where appropriate paracetamol 1 gram PO and ibuprofen 400mg PO. (be aware of patients with nephropathy)
    - If morphine allergy or if unsure, discuss with Haemtology Registrar
    - Prescribe second dose of analgesia in the event that pain is not controlled within 30 mins of first dose
- Do NOT use pethidine.
- Entonox should not be used after leaving the ambulance
- Anti-pruritics, antiemetics, laxatives as needed (avoid parenteral cyclizine)
- Contact and discuss with the Haematology Registrar for sickle cell (or on call registrar if out of hours)
- Contact and discuss with the CNS for sickle cell disease if within working hours
- Review pain levels within 30 minutes and every 30 minutes with top up doses of analgesia as necessary until pain relief is achieved and then at least every 4 hours
- Hourly observations to monitor pain, sedation, vital signs, respiratory rate and oxygen saturation for the first 6 hours and 4 hourly thereafter until they leave hospital or the episode has ended. Staff should be alert to potential risk of opiate toxicity and act on any concerning observations
- Pain management on the ward varies between patients – for patients who do not have an individual pain protocol then the following can be considered but should be discussed with haematology registrar for sickle cell or the on-call registrar if out of hours

- Those with severe pain and requiring parenteral opiates are usually managed with regular 2 hourly subcutaneous morphine (or oxycodone) injections. Breakthrough injections should not be prescribed and if pain control is not achieved then the dose of the 2 hourly injections should be increased
- Some patients with moderate pain may require PRN to allow them to reduce or manage their analgesia
- Some patients on very high doses of parenteral opiates may require continuous subcutaneous infusions – these patients should have individual pain protocols.

Routine investigations often needed on reviewing a patient with a painful crisis include

- FBC ,reticulocyte count, Group and Save sample (marked 'sickle cell patient')
- Renal and liver biochemistry
- Baseline pulse oximetry on air
- If signs of infection: blood, urine and sputum cultures
- If signs of chest involvement: CXR
- If reduced SpaO2 levels then consider arterial blood gas sampling if less than 94% or significantly less than baseline
- Plain XR of bones during crises are rarely indicated

Transfusions are rarely required to help manage simple painful crises.
Patients with sickle cell disease should not be transfused without discussion with the haematology team first.

If requesting blood it is imperative that the blood transfusion laboratory is aware that the receiving patient has sickle cell disease.

6.2 Acute Chest Syndrome

This is one of the leading causes of mortality in patients with SCD and 50% of all patients will have at least one episode of acute chest syndrome (ACS) in their lifetime.

ACS is usually described as a syndrome of new pulmonary infiltrates with respiratory signs or symptoms. The cause is often unclear and is often multifactorial including infection, pulmonary sickling and sequestration and fat embolism (secondary to bone infarction).It can be difficult to distinguish from pneumonia, pulmonary embolism or Adult Respiratory Distress Syndrome (ARDS). As an ACS progresses patients can demonstrate falling haemoglobin levels and a progressive thrombocytopenia. ACS can often develop in patients admitted with a painful crisis and no chest signs at presentation. Patients with back pain, rib pain and chest pain should be offered incentive spirometry via physiotherapy on wards (See Guidelines on Incentive Spirometry in SCD).

Signs and Symptoms

- Hypoxia
- Chest pain
- Signs of consolidation – usually basal, can be bilateral
- Fever
- Tachypnoea
- Tachycardia
- Shortness of breath
- *CXR changes may lag behind development of signs and symptoms*

Essential investigations:

- FBC, Group and Save, Renal and Liver biochemistry and a CRP
- Arterial Blood Gas sampling if saturations less than 94%
- CXR
- Blood and sputum serology and atypical serology, Naso-pharyngeal aspirates for viral testing

Management

- Inform the haematology team urgently
- Intravenous fluids,
  (but avoid hyper-hydration as can predispose to pulmonary oedema)
- Intravenous antibiotics if concerns of infection
- Adequate pain relief whilst monitoring for opiate toxicity
- Regular, frequent observations monitoring SpaO2, RR and O2 requirements
- NIV may be required, early involvement of critical care team is important
- Liaise with chest physiotherapy for any support which may be helpful
- **Urgent blood transfusion (which may need to be an exchange transfusion)** should be considered depending on hypoxia, Hb and clinical course. The haematology team will decide and arrange
- VTE prophylaxis if not contraindicated - It can be difficult to clinically exclude a pulmonary embolism and so initially therapeutic anticoagulation may be indicated until this can be excluded.

Patients with an ACS should be managed on a haematology/sickle ward (Ruth Myles or Gordon-Smith) and exchange transfusions can occur there but out of hours emergency exchanges should take place HDU. There should be close liaison with the critical care team as many patients will need to be managed in a level -2 or even level-2 critical care environment. The clinical condition of the patient may deteriorate and patients may occasionally require invasive ventilation. Patients with ACS should not be managed on outlying wards.

On discharge patients should be encouraged to take their penicillin prophylaxis, update any vaccinations that are needed and be offered the option of hydroxycarbamide in discussion with the

haematology consultant. If hydroxycarbamide has not been successful or is not acceptable to the patient then a regular transfusion programme to prevent recurrent episodes of ACS can be considered.

Involvement of physiotherapy during a paitents admission can alleviate the severity.

In patients who are blood refusersm the early use of physiotherapy and NIV is imperative.

6.3 Fever and overwhelming infection

Patients with sickle cell disease are advised to seek medical attention early if they have any signs of infection (especially a fever >38^{o}C). These patients are often hyposplenic, especially those who have HbSS disease who have usually auto-infarcted their spleen in early childhood.

They are particularly susceptible to encapsulated organisms (*H. influenzae*, *S. pneumoniae*, and *Neisseria meningitidis*.) and primary prevention is encouraged by offering vaccinations and regular penicillin V prophylaxis.

They are also at high risk of osteomyelitis due to bone infarcts and this often involves salmonella species.

If presenting with signs of sepsis then prompt treatment is required to

a) Prevent development of a full sepsis syndrome
b) To try and prevent infection triggering a sickle cell

Initial management

- Vital sign observations
- Blood tests including:FBC, reticulocyte count, renal & liver biochemistry, CRP
- Septic screen – urine, blood, sputum cultures, CXR if indicated
- Antibiotics – empirical treatment (if no clear source obvious)
  Co-amoxiclav with clarithromycin or
  levofloxacin alone if penicillin allergic
- Intravenous fluids and oxygen therapy
- Analgesia if concomitant pain crises
- Imaging – CXR for suspected pneumonia, USS abdomen may be indicated if concerns of biliary involvement

If chest pains, sign of pulmonary involvement and / or hypoxia consider diagnosis of acute chest syndrome and discuss urgent management

Osteomyelitis can be difficult to distinguish from severe bone crisis or bone infarct. All can present with swollen, tender, erythematous areas but VOC are 50 times more common than osteomyelitis. Radiological, orthopaedic and microbiological opinions should be sought to consider possibility of getting any diagnostic material and ensuring antibiotics cover salmonella.

Malaria: a travel history should be taken and malaria considered if recent travel to an at-risk country and chemoprophylaxis not strictly adhered to. Patients with SCD can contract malaria even though many patients may think they are immune and malaria should be investigated if there is clinical suspicion.

ParvoB19: this viral infection produces a transient red cell aplasia and should be suspected in infections with anaemia and reticulocytopenia.

6.4 Acute Abdominal Pain

The most common cause of abdominal pain in SCD is a result of veno-occlusion of the mesenteric circulation and is often referred to as girdle syndrome which can result in a symptomatic ileus.

- **Girdle syndrome**: management is usually supportive ensuring the patient is warm, well hydrated and well oxygenated. Surgical review maybe indicated and other causes of abdominal pain should be considered.

- **Cholelithiasis and Cholecystitis / Choleangitis**: Pigment gallstones are common in paitents with SCD (30% of children, 70% of adults.) Associated infections are not infrequently seen. General management of biliary sepsis is as in the non-sickle cell population (usually with antibiotics in line with St George's Healthcare NHS Trust antibiotic policy) with the addition of monitoring for sickling. Elective cholecystectomy is often indicated after the acute event and surgical plans should be developed in discussion with the haematologists.

- **Splenic Infarction**: This can occur in patients with residual spleens, most commonly seen in those with HbSC sickle cell disease. This is usually identified on ultrasound scanning and the treatment is supportive.

- **Splenic and Hepatic Sequestration**: These conditions are more common in children though hepatic sequestration can still occur in adults. The pathology is the trapping for the red blood cells within the liver and patients present with acutely tender hepatomegaly, marked anaemia with a reticulocytosis and a conjugated hyperbilirubinaemia. Management is usually supportive including top up red cell transfusion. There can be coexisting infections including salmonella species and broad spectrum appropriate antibiotics should be considered.

6.5 Emergency Surgery

If emergency surgery is required then it should not be denied due to sickle cell disease but the haematology team must be involved at the earliest stage. Consideration must be given to the following.

- Pre-operative management: **Transfusion** may be indicated (either top up or exchange) if time allows depending on type of surgery and patients genotype and phenotype. The aim is to prevent postoperative sickling and to reduce the risk of acute chest syndrome, stroke and other complications. If transfusion is thought to be indicated and not able to be done pre-operatively then should be considered post operatively.
- Oxygenation: Patients should be well oxygenated before, during and after surgery
- HDU / Critical care bed post op and access to early non-invasive ventilation if needed
- Hydration (intravenously until able to eat and drink)
- Access to chest physiotherapy post operatively
- Good analgesia taking into account the patients previous experience with opiates – patients may either be opiate naive or tolerant
- VTE prophylaxis
- Low threshold for antibiotics

If emergency surgery is required and the sickle cell status of a patient is unknown then the patient can initially be screened with a sickle solubility test (‘sickle screen’ on iClip)

If this is positive then if the surgery is in hours and time allows an HPLC (high performance liquid chromatolography) to confirm the diagnosis of trait or disease can be done within 3 hours of receipt of an EDTA sample. Please do discuss with the haematology SpR to help coordinate the laboratory, anaesthetic and clinical pathway.

If out of hours or there is not time to wait then treat the patient as sickle cell disease and involve the haematology team. Keep patients warm, hydrated and well oxygenated during surgery and try to avoid acidosis. Decisions regarding transfusion and HDU can be made .

It is important to know the transfusion history of the patient to be able to interpret both the sickle solubility test and the HPLC

6.6 Acute Anaemia

Patients with sickle cell disease, especially homozygous HbSS disease, will have a baseline mild – moderate anaemia due to chronic haemolysis. This baseline value can vary markedly between individuals and so a diagnosis of acute anaemia needs to be made with the knowledge of the patient's baseline Hb. Patients will often know their own Hb values and patterns of anaemia.

Management of anaemia and transfusion decisions rely on the clinical symptoms and the drop in Hb. Often once the drop is greater than 20g/L then intervention is required or if Hb is <50g/L.

Causes of Acute Anaemia

- **Blood loss:** History, examination, causes and management should occur as in the non-sickle population. Transfusion support may be required.

- **Increased Haemolysis during Crisis:** This is usually associated with an acute crisis, increased jaundice and reticulocytosis. Transfusion of red cells may be indicated depending on the patient's clinical symptoms and fall in Hb. However most painful crises do not require transfusion.

- **Aplastic Crisis:** - This is a case of transient red cell aplasia usually due to Parvovirus B19 infection, which results in a halt of red cell production for approximately 4-8 days. Patients may present with a profound anaemia and reticulocytopenia which requires transfusion support until bone marrow production restarts. Patients may or may not report a viral prodromal scenario. This is a unique, self-limiting illness. Parvovirus B19 serology can be sent. Pregnant staff and visitors should avoid contact with patients with suspected infection. Pregnant patients with Parvovirus B19 infection should be discussed with the obstetric team and may require fetal medicine unit follow up.

- **Hepatic / Splenic Sequestration:** Patients present with tender organomegaly, anaemia, reticulocytosis and at extreme cardiovascular collapse. Supportive treatment including transfusion is usually required.

- **Other causes** of anaemia to consider include
    - G6PD deficiency triggered haemolysis
    - Hyperhaemolysis - a complex, severe but rare condition usually occurring after red cell transfusion where the recipient and donor red cells are destroyed. Future transfusions will need to be carefully considered.
    - Delayed haemolytic transfusion reactions.

6.7 Acute Renal Failure and Haematuria

Mild – moderate acute renal failure is relatively common in patients presenting with a sickle cell crisis and is often multifactorial.

Causes include

- NSAIDs and other nephrotoxic drugs
- Dehydration
- Sepsis
- Hypothermia

Acute papillary necrosis can also present with obstructive acute renal failure and haematuria.

The management of this process reflects the underlying causes

- Fluid resuscitation and fluid balance
- Exclude or treat infection (send MSU)
- Stop nephrotoxic drugs
- Liaise with nephrology early or urology for haematuria
- Further bloods (eg autoimmune screen) , imaging (eg USS, KUB) and other investigations such as cystoscopy, renal biopsy as guided by possible causes

6.8 Acute Stroke / Neurological Events

This is one of the leading causes of death in patients with sickle cell disease and can occur at any age. Ischaemic strokes are more common in children and older adults (over 30 years old) whilst intracerebral haemorrhages (related to ruptured aneurysms and moya moya formations) are more common in younger adults.

Patients who have had a previous stroke are at high risk (60-70%) of subsequent events including haemorrhagic conversions after ischaemic strokes if a secondary prevention transfusion programme is not undertaken

St George's is a hyperacute stroke unit that has on site vascular interventional neuroradiology, neurology and neurosurgery.

Management of Suspected Stroke

- **Urgent imaging**. At presentation an urgent CT and CT Angiogram (CTA) scan should be performed. MRI with digital subtraction angiography may be required to help confirm the diagnosis but should be performed after an exchange blood transfusion once HbS% is less than 30%.
- **Clinical stabilisation**. This includes careful fluid rehydration and oxygen therapy.
- **Transfusion**. Urgent exchange transfusion is indicated for an infarct but an initial top up transfusion may occur if the presenting Hb is < 6g/dL. The aim is to improve tissue oxygenation and to minimise damage occurred. The HbS% post transfusion should be <20%. A second procedure may be needed the 2nd or 3rd day to keep the S% less than 30% at all times.
- **Thrombolysis.** Currently there is no evidence of increased intracranial haemorrhage in adults with SCD and acute stroke who have received thrombolytic therapy and so patients should be considered for thrombolysis if there are no contraindications and they meet the criteria. If patients are being considered for **thrombolysis** along the stroke pathway then haematology should be involved with thrombolysis discussions. The definitive treatment for an ischaemic stroke that has occurred due to sickle cell disease should involve an exchange transfusion with urgent reduction of the HbS%.

Intracerebral Haemorrhage: If this is confirmed on imaging then it should be discussed with the neurosurgical team on call. Mainstay of treatment is as in the non-sickle population. The indication for an exchange blood transfusion is less clear but is usually suggested – particularly if neurosurgical intervention is required. This may need to happen post surgery due to time pressures.

Automated exchange transfusion is preferred due to less labiality with blood pressures. However it is important to be aware that automated exchange transfusions can result in a transient thrombocytopenia

Following a stroke the patient should be managed with the stroke team to enable access to physiotherapy, occupational therapy and neuropsychology for the optimal rehabilitation. Patients who have had an ischaemic stroke should continue on a regular exchange transfusion programme to maintain an S% of less than 30% at all times.

Other neurological events that may be seen in patients with SCD include seizures, venous sinus thromboses and behavioural changes. These should be managed jointly between haematology and neurology.

6.9 Priapism

Venoocclusion of the penile circulation results in a painful, unwanted and persistent erection. This affects 10-20% of all men with sickle cell disease usually first presenting in adolescence. Resolution should be achieved within 4 hours to preserve erectile function. For this reason patients in outpatients should be reminded to present to the ED if episodes of priapism are unresolved at 2 hours.

Stuttering Priapism. This is usually self limiting and patients often manage at home. Patients will often have recurrent episodes but may be reluctant to disclose these problems. Patients who do discuss their symptoms may describe fluctuating levels of erection over an hour or two or several events a day or a week. Simple techniques such as drinking fluids, gentle exercise (climbing stairs, jogging on the spot) trying to pass urine and keeping warm can all help. These patients should be referred for urological follow up and consideration of prophylactic treatment with etilephrine.

Fulminant Priapism. This is defined as a prolonged attack, lasting longer than 3 hours. This needs urgent treatment to prevent cavernosal fibrosis and impotence and should be treated as a medical emergency after 1 hour.

Management

- Confirm priapism and encourage to pass urine (catheterisation may be required)
- Hydration – oral or intravenous
- Warmth – **DO NOT USE LOCALLY APPLIED ICE PACKS**
- Adequate analgesia
- Urgent discussion with urology for consideration of etilephrine (orally or intracorporal), aspiration or in severe cases surgical intervention.
- Routine blood tests including FBC, renal profile and group and save

There is no evidence for the use of emergency exchange transfusion to treat prolonged priapism. Top up transfusion would be indicated if Hb is <60g/L or after discussion with haematologists on call. EBT can be considered in refractory cases requiring surgery.

6.10 Acute Changes in Vision (With Miss C Egan, Consultant Ophthalmologist)

Patients with sickle cell disease, especially those with HbSC disease, can suffer from many ophthalmological conditions that can affect the optic disc, macula and the retina.

Sudden changes in vision needs urgent referral to eye clinic / eye casualty for formal ophthalmological assessment and ongoing haematological input.

Consideration should be given to a central nervous event (eg occipital stroke) in patients with loss of vision but normal ophthalmological investigations.

It is important to remember the retinal artery occlusions should be managed as a stroke as this is a central nervous artery.

6.11 Admissions and Discharge

Most patients presenting to the ED will need admission for pain relief or treatment of other complications. Some patients however may well be able to go home from the ED with follow up arranged. This maybe a review on the day unit, clinic appointment or a meeting with one of the community sickle cell Clinical Nurse Specialists (CNS) depending on which borough the patient lives in (we have links with sickle cell community CNSs in Wandsworth, and some other local boroughs)

Patients are admitted directly under the haematology team and a bed on Ruth Myles Ward (St James' Wing) or Gordon-Smith Ward (Lanesborough Wing) should be requested.

The haemoglobinopathy team consists of a Consultant Haematologist, Haematology Registrar, a Clinical Health Psychologist and a Clinical Nurse Specialist.

During admission patients will be seen regularly by different members of the teams. There are multidisciplinary (Consultant, CNS and Psychologist) ward rounds twice a week.

Specific things to consider

- Is pain relief sufficient? Are pain levels being reviewed regularly on the ward?
- Are pain assessments documented?
- Observations should be done 4 hourly
- Is there a plan for reducing analgesia?
- Constipation, mobility, VTE prophylaxis, antibiotic rationalisation and mood should all be considered daily
- Is fluid intake sufficient?
- Do work or college need to be informed of admission?
- Are there child care issues that need to be addressed?
- Are there any safeguarding or capacity issues?
- Would referral to Full Circle Therapy Team be appropriate to offer?
- Would individual nursing or psychology input be useful?
- Are there discharge plans in place including an estimated discharge date?

Our **Clinical Nurse Specialist** will see patients on regular ward rounds but can also see patients at separate times and support nursing staff on the ward if needed. She can be contacted by bleep or telephone (see contacts later)

Our **Clinical Health Psychologist** will see patients twice a week on the multidisciplinary ward round too and patients can ask to see her at other times and nursing staff or medical staff can also discuss issues with her. She can be contacted by telephone or email (see contacts later)

Discharge planning

This should include a planned date of discharge, consideration of whether further time will be needed off work or college, who will help look after or support the patient if further recuperation is required at home and what pain relief will be needed post discharge.

There should also be follow up plans discussed. Many patients may not need increased follow up other than their routine clinic appointment but other options include a day unit review, earlier clinic appointment with either the doctor or the CNS, clinic appointment with the Clinical Psychologist and or review by a community sickle cell nurse specialists.

Other points

Patients should not be moved between wards without discussion with the clinical team looking after them. In particular patients should not have multiple ward moves and if moves are essential this should be explained to the patient and not happen overnight if possible. Moves can worsen pain and delay discharges.

Behavioural Challenges

The St George's Healthcare NHS Trust Zero Tolerance (Violence an Aggression) Policy should be adhered to at all times. Although pain, fear of illness and hospitals can be challenging for patients intimidating, violent or aggressive behaviour will be acted upon and not tolerated. If nursing staff become concerned about behaviour this should be discussed with a senior nursing member of staff – including the site managers out of hours and a member of the haemoglobinopathy team in hours.

Some patients who do find admission to hospital very difficult may have individual care plans that cover all aspects of care and admission (medication, other treatment options as well as discussions about behaviour) and these can be found uploaded on EPR. They are also available from members of the haemoglobinopathy team.

Concerns related to safeguarding, unacceptable or unusual behaviour should be raised with the Consultant Haematologist for Haemoglobinopathies or the Clinical Psychologist. Acute psychiatric concerns out of hours should be discussed with the Liaison Psychiatrist on call (bleep 6501)

All concerns regarding safeguarding or unacceptable behaviour should be recorded in the medical notes, and nursing staff should be encouraged to do the same.

Hospital Contact Details

Dr Elizabeth Rhodes	Haematology Consultant	ext 0885
Dr Julia Sikorska	Haematology Consultant Haematology Registrar	ext 0885 bleep: 7080
Carol Rose	Sickle Cell CNS	mobile: 07500835735
Full Circle	See referral form on haematology intranet or fullcircle intranet page	

Out of hours

Haematology Registrar	via switchboard
Haematology Consultant	via switchboard

Useful Numbers

Safeguarding Adults	David Flood	Bleep 8031
Liaison Psychiatry	On Call	Bleep 6501
Chaplain Contacts		ext 3285

7. Dissemination and implementation

7.1 Dissemination:

Guidelines will be available on the departmental intranet page and available in paper form in the junior doctor office in haematology.

7.2. Implementation

Guidelines will be promoted by the haemoglobinopathy team

8. Consequences of Breaching the Policy

Failing to follow this policy could lead to action under the Trust's disciplinary policy.

9. Monitoring compliance

The table below outlines the process for monitoring compliance with this document.

Monitoring compliance and effectiveness table					
Element/ Activity being monitored	**Lead/role**	**Methodology to be used for monitoring**	**Frequency of monitoring and Reporting arrangements**	**Acting on recommendations and Leads**	**Change in practice and lessons to be shared**
QSIS / SSD – quality dashboard – includes analgesia times in the ED	*Consultant haematologist linking with Emergency department consultant*	*Audit submitted annually*	*Audit will be submitted to quality dashboard and discussed at departmental clinical governance* *The lead or committee is expected to read and interrogate the report to identify deficiencies in the system and act upon them. Consider stating this responsibility in committee terms of reference.*	*Haematology and Emergency Department will undertake subsequent recommendations / action planning for any or all deficiencies and recommendations within reasonable timeframe?* *Required actions will be identified and completed in a specified timeframe. Consider stating this responsibility in committee terms of reference.*	*Required changes to practice will be identified and actioned within a specific timeframe. A lead member of the team will be identified to take each change forward where appropriate. Lessons will be shared with all the relevant stakeholders.*
WMQRS peer review quality standards	*Consultant haematologist*	*As required (every 2-3 year)*	*The lead or committee is expected to read and interrogate the report to identify deficiencies in the system and act upon them. Consider stating this responsibility in committee terms of reference.*	*Required actions will be identified and completed in a specified timeframe. Consider stating this responsibility in committee terms of reference.* *These will be discussed at Divisional governance board*	*Required changes to practice will be identified and actioned within a specific timeframe. A lead member of the team will be identified to take each change forward where appropriate. Lessons will be shared with all the relevant stakeholders.*

10. Associated documentation

11. References

- West Midlands Review Service – Quality Standards: Health Services for People with Haemoglobin Disorders (2018/19)

- Standards for the Clinical Care ofAdults with Sickle Cell Disease in the UK 2018 (Sickle Cell Society) https://www.sicklecellsociety.org/wp-content/uploads/2018/04/Web-version-FINAL-SCS-Standards-GSM-6.4.18.pdf

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