

Clinical Guidance

Management of patients with Sickle Cell disease in the haematology day unit:

The Nurse Led Acute Pain Service and Elective Transfusions Service Standard Operational Procedure

Summary

The Sickle Cell Day Unit service is an integral part of the new Haematology and Sickle Day Unit at Guy's Hospital. Its role is multi-faceted and includes: a) The immediate management of patients with Sickle Cell Disease (SCD) who present with acute complications of SCD including acute painful crises b) Blood transfusion (either exchange or top-up) of patients with Sickle Cell Disease (SCD), Thalassaemia and other inherited anaemias c) The review of patients with SCD and Thalassaemia who present with other (non-emergency) medical issues The service is nurse-led with medical support from the sickle cell team and is part of the routine work of the Day Unit staff. It is overseen by the sickle cell nursing and medical team, who support, monitor and aid development of the service. The majority of patients attending the Haemoglobinopathy service at this trust have SCD, but there is a small number with thalassaemia who primarily attend the service for transfusion support.

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Definitions/Abbreviations

SCD	Sickle cell disease	EMU	Clinical Decision Unit
CNS	Clinical nurse specialist	A&E	Accident and Emergency Department
MDT	Multi disciplinary team	SC	Subcutaneously
HbS	Sickle haemoglobin	GP	General Practitioner
RBC	Red blood cell	IV	Intravenous

Haematology and Sickle Day Unit Opening Hours:

09.00 – 1800hrs Monday to Friday

Patients wishing to access the Acute Pain Management Service must attend by 4pm

Patients requiring treatment outside of these hours, must attend A+E

Important phone numbers:

Adult sickle cell team -_Department of Haematology, 4th Floor, Guys Hospital.

Consultant Haematologist Dr Jo Howard ext 812471

Dr Rachel Kesse-Adu ext 812471

Sickle Team Specialist Registrar bleep 0248

Sickle Team Foundation year trainee bleep 2283

Advanced Nurse Practitioner - Mr Neill Westerdale – Bleep 1843/Ext 8271

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Acute Pain Service and Admission Protocol

These guidelines are designed to provide trained nursing staff involved in the direct care of patients with sickle cell disease, with a tool for assessing and implementing safe and effective clinical management.

1.0 Introduction

The nurse led pain service in the in Haematology day unit at Guy's Hospital has a multi-faceted role which and includes:

- a) The immediate management of patients with Sickle Cell Disease (SCD) presenting with acute painful crises including those who may require admission.
- b) Blood transfusion (either exchange or top-up) for patients with SCD, Thalassaemia and other inherited anaemias.
- c) The review of patients with SCD and Thalassaemia who present with other (non-emergency) medical issues.

This is intended to be a nurse-led service which is part of the routine work of the Day Unit nursing staff managed with support from the Sickle Cell Nurse Specialists (ANP or CNS) and the medical team.

The majority of patients attending the Haemoglobinopathy service at this Trust have SCD; however there are also a small number with thalassaemia. For ease of understanding, in particular in the lay population, the service will be referred to as the nurse led sickle cell pain service.

2.0 Background

SCD is the most common inherited condition in the UK, Lambeth, Southwark and Lewisham have the highest prevalence in the UK. It is characterised by a lifelong anaemia and intermittent, unpredictable episodes of acute pain, which may be severe enough to require hospital attendance for opiate analgesia. Currently, a SCD patient with an episode of severe pain will attend the A&E department, but this is not always ideal as the patients may not receive effective analgesia in good time and they may be seen by staff unfamiliar to them, or with the management of SCD.

The use of a Day Unit approach in managing sickle cell pain is important as it should provide swift assessment and adequate analgesia which may curtail a serious crisis and enable the patient to avoid a hospital admission this (is consistent with long term condition care and good disease management (DoH, 2007). There is good evidence of the efficacy of this approach, both in the UK and internationally. (Wright *et al*, 2004, Benjamin *et al*, 2000, Ware *et al*, 1999). In addition UK Guidelines have stated that all patients presenting with acute sickle pain should receive initial analgesia within 30 minutes and have achieved good pain relief within 2 hours. They have suggested that the Day Unit model is an excellent way to achieve these aims. (Nice, 2012, UK Standards 2008, Rees *et al*, 2003).

3.0 Objectives

- a) Patients with SCD who present with acute sickle pain will be assessed and managed effectively, rapidly, and safely, by staff who understand the complications of SCD
- b) The early and effective treatment of simple uncomplicated painful episodes will aim to reduce admissions.
- c) The nurse led pain service will facilitate admission if necessary
- d) Patients, who fall within the exclusion criteria (see 4.2) e.g. pregnant patients, will continue to use the Accident & Emergency department .
- e) The day case management facility will be Nurse led with support from the sickle medical team.
- f) Patients receiving regular blood transfusion will have these administered in a timely and safe manner. (The continuing need for transfusions, its complications and need for iron chelation therapy will be reviewed by the consultant in charge of their care regularly.)
- g) To identify the frequent users of the service with complex needs early and refer them for comprehensive support involving the health psychology team in a timely fashion.

4.0 Acute Pain Management Service

This policy should be used in conjunction with the main GSTFT sickle cell disease adult guidelines on the Trust intranet. Patient assessment should be clearly documented using the Day Unit pain assessment forms which should be filed in the outpatient section of the medical notes (Appendix 1) All patients attending the GSTFT Sickle Cell OPD will have an analgesia protocol on EPR (or a more detailed written care plans for the more complex patients which are available on the Day unit, A&E majors and EMU, these provide a guide for analgesia and individualised care) and all patients who have used the day unit acute pain service will have a signed drug chart with the administered analgesia prescribed on it. When a patient is admitted this prescription chart will follow the patient to the ward. Patients who are not known to GSFT SCD service or have not attended the sickle OPD for over 1 year must be medically reviewed prior to administration of any opiate analgesia.

The Day Unit staff are primarily responsible for initial management of patients with acute pain, including the initial administration of opiates, but the service will be covered at all times by a SCD specialist nurse, the Day Unit doctors or the sickle team FY2 or SpR .

The aims of treatment are:

- To provide rapid, effective analgesia and to assess patients for complications of SCD.
- To break the vicious cycle of sickling → hypoxia and acidosis → more sickling, all exacerbated by dehydration.

4.1 Source of referrals

The majority of patients will self-refer. In order to triage patients effectively and manage patient flows, all patients should telephone the SCD team (via the designated mobile number) before attending. Patients may also be referred from clinic, the community nurse or their GP. All patients requiring pain management outside the hours of 9am-4pm must attend A&E. Un-announced walk in patients who we are unable to review or manage in the HDU due to lack of space or pregnancy must also be triaged before transfer to St Thomas A+E.

4.2 Exclusion criteria

If a patient telephones and during triage assessment they are found to have any of the following clinical issues;

- Pregnancy < 18 weeks advise to attend the A+E department
- Pregnancy > 18 weeks advise to attend the Antenatal day assessment unit or birth centre
- Retinal problems e.g. visual loss or blurring, advise to attend Eye Casualty at St Thomas' hospital
- New neurological symptoms advise to call an ambulance to attend A&E department immediately.

4.3 History taking (Use Pain assessment documentation, Appendix 1 and 2)

Pain characteristics

- Onset and duration of pain
- Site of pain
- Nature of pain: whether similar to sickle type pain or other. Pain that is not due to a sickle cell crisis or chronic bone pain (AVN) should be referred to medical staff for evaluation
- Severity of pain: record pain and mood score
- Precipitating factors for example infection, cold weather or stress
- Analgesia/other agents used for pain relief prior to presenting to the centre

4.4 Observations

Take baseline observations of:

- Blood Pressure,
- Pulse,
- Temperature,
- Oxygen Saturations, (on air)
- Respiratory rate,
- Mood score, (see appendix 2)
- Pain Score (0 -10) initially (see appendix 2)

Observations should be repeated 2hrly when administering analgesia or more frequently if indicated. For patients on parenteral opiates, the respiratory rate must be checked every 20 minutes until the pain is controlled and then every 2 hours. The pain assessment chart (appendix 2) should also be used.

4.5 Administration of Analgesia

- Analgesia should be given within 30 minutes of the patient presenting. Document on the Day Care Assessment form (appendix 1)
- Efficacy of analgesia should be assessed initially 30 – 40 minutes after administration, using pain assessment chart (appendix 2)
- If the pain does not improve after the 2nd dose, analgesia dose might need titrating – patient will require a medical review.
- Analgesic efficacy is improved with the addition of Paracetamol and/or oral ibuprofen if no contraindications (nephropathy/gastric ulceration).
- The last dose of parenteral analgesia should not be administered later than 2 hours prior to the Day Unit closing time for those patients not requiring admission. This will allow sufficient time to ensure adequacy of a response to treatment and to monitor for complications such as respiratory suppression following opiate analgesia. Patients requiring admission can have analgesia up to the point they are transferred onto the ward.

4.6 Medical review criteria

If a patient's telephone triage assessment or initial observations on arrival to the day unit reveals any of the following signs and symptoms (see list below) a member of the medical team should be informed to review the patient immediately after arrival to the unit and a medical bed booked if appropriate. Medical reviews will be undertaken by haematology day unit doctor or sickle FY2 (bleep 2283) with supervision from the Sickle registrar or Consultants.

Patients with ANY of the below should be reviewed by the medical team on arrival

- Chest pain
- Shortness of breath
- Hypoxia (oxygen saturations <94%)
- Fever/rigors (Temp >38°C)
- Hypotension (BP <90/60)
- Tachycardia > 100 (even after pain has settled following analgesia)
- Raised respiratory rate of > 20 (even after pain has settled following analgesia)
- Abdominal pain
- Priapism (persistent erection)
- New neurological signs, confusion, numbness or weakness of the limbs
- Patients Hb dropped to 20g/L below baseline or less than 60g/L
- PAR scoring > 4
- Pregnancy (in cases of walk in)
- Concerns from the nursing team about the patient's clinical condition

4.7 Fluids

- Adequate oral fluid intake is essential.
- Encourage patient to continue oral fluids aiming for a total intake of at least 3 litres per 24 hours.
- Maintain adequate fluid balance chart recording strict input and output

If the patient is not able to drink adequately or is feeling nauseous, nursing staff should commence fluid replacement by intravenous route according to adult SCD guidelines.

4.8 Oxygen

- Administer humidified oxygen at 2-4 L/min by mask or nasal cannulae if oxygen saturation on air is less than 94%.
- If oxygen requirements are increasing to maintain oxygen saturation the patient will require a medical review.
- Arterial blood gases should be undertaken if oxygen saturations are below 94% on air and refer patient for medical review.

Remember that excess Opiotes can cause respiratory suppression. (Nalaxone is occasionally required.)

4.9 Investigations

If the patient is requiring admission or acute sickle cell pain is not resolving the following investigations should be initiated

- FBC, retics
- U&E's
- LFT's
- LDH
- CRP
- G&S

If a patient has a fever with temperature > 38° C refer patient for medical review, additionally send off:

- Blood cultures
- Dip stick urine MSU
- Throat and viral swabs (if appropriate)
- Stool cultures (if diarrhoea)
- Chest X ray (if appropriate)
- Pregnancy test if appropriate (+ result needs referral to medical staff)

5.0 Admission from the acute pain management service

Patients may require admission and medical review if they have:

- Unresolved pain (pain score >7) requiring opiate analgesia or any on-going symptoms or complications outlined in 4.6

The nurse caring for the patient should ensure

- The day unit doctor or sickle cell FY2 (bleep 2283) is informed the patient requires medical review and clerking prior to transfer to the wards.
- Bed managers are given adequate notice to secure a bed (bed manager bleep 1165). If a bed is not available on the Guy's site for a sickle patient admission this should be discussed with the Inpatient matron for Haematology and Oncology (bleep 1639) and the sickle cell consultants prior to transfer to St Thomas' site.
- If the patient is transferred to St Thomas' for any reason the appropriate bed Manager (bleep 0162), sickle cell doctors bleep 0248/2283/clinical nurse specialist (bleep 1843) on the St Thomas' site are informed of the admission. When transferring the patient a medical bed should be secured rather than transferring to the A&E department.
- Adequate handover is given to the ward nurses as well as the on call team particularly for those patients who are unwell and will require review
- Routine bloods and other investigations (outlined in 4.9) are completed.
- The patient has a cannula in situ if medically indicated.
- A copy of patient's care plan (if one in use) is placed within medical notes.
- That patient is reasonably comfortable prior to transfer.
- That the medical notes and drug chart are transferred to the ward with the patient.
- That where a transfer to St Thomas' is required this is done safely with nurse escort or paramedic crew if indicated.
- If a patients refuses admission for any reason clear documentation of the advice given to patient must be made in the medical notes (Follow procedure for discharge against medical advice.)

5.1 Walk-in patients

Patients with SCD and thalassaemia may attend the department for advice, such as a SCD patient with a fever or thalassaemic patient was reporting excessive fatigue. Such patients should be encouraged to telephone one of the sickle clinical nurse specialist team first so that space can be identified in the unit and the medical notes prepared prior to their arrival. If space is available the

patient should have a brief nursing assessment including basic observations, haematological investigations and Day unit SpR or sickle cell FY2 on bleep 2283 should review the patient if clinically indicated and prescribed treatment that may be required. When there is no suitable space to keep the patient in the unit they should be assessed and given initial management such as analgesia then advised to attend the A+E all such patients must be discussed with the sickle medical and nursing team.

5.2 Discharge from the Acute Pain Management Service

Patients with uncomplicated crises who are more comfortable following day case treatment (pain score <7) may be discharged by Day Unit Nursing Staff been trained by the Sickle Team to do so, the Day unit SpR or the sickle FY2/SpR with a limited prescription for oral analgesia such as Paracetamol and Dihydrocodeine for 2-3 days and provided with a discharge information leaflet. (Appendix 3)

A discharge summary must be completed and posted to the patient's GP and copy to Dr J. Howard/Dr R Kesse-Adu (Consultant Haematologists) detailing the day case treatment and any advice given. (Appendix 4).

Where a any patient is identified who may benefit from home follow up by the specialist nursing community team the South East London Sickle Cell and Thalassaemia Centre is made aware (refer to section 10.0 for details). Patients are referred to sickle health psychology team if appropriate (refer to section 9.0 for details).

Patient should be encouraged to make their own arrangements for transport home and aim to avoid public transport in the winter months. Hospital Transport is only available for patients who have limited mobility, which should not apply to any of the patients attending the Acute Day Unit pain service.

5.3 Advice on discharge

Patients must be advised to re-present to A&E (out of hours) if their pain subsequently worsens or return to the Day Unit the following day to be reassessed (between 09.00 – 16.00hrs).

Following day case treatment with Opiate analgesia, patients are at risk of sedation/drowsiness and should be advised NOT to drive.

Patients should be encouraged to access the health psychology service to gain additional support around developing effective coping strategies.

5.4 Prescriptions

A limited supply (up to 2-3 days) of oral analgesia such as Paracetamol and Dihydrocodeine or a NSAID (if no contra-indications) may be prescribed to take away following day case treatment; patients who require subsequent analgesia should be re-evaluated. Requests for re-supply of oral opiates should be discussed with a member of the medical sickle cell team.

Any changes to individualised opiate analgesia regime MUST be authorised by the Haematology consultant, documented in the patient notes and reflected in the analgesia plans and on the register.

All prescriptions issued from the Day Unit, either for day-case patients or patients who are reviewed in the clinic should be recorded in the prescription folder which is the central record kept in the Day unit.

All patients who receive regular prescriptions of strong Opiates MUST be reviewed in clinic every 8 weeks. Those who default should not be issued a repeat prescription unless reviewed in clinic: this is to ensure that there are no clinical changes which would impact on medications issued.

5.5 Follow-up

Please ensure all patients have follow up clinic appointments when discharged home. If they do not have an appointment, one should be booked on ext 82741.

All patients should be given details of their next Sickle outpatient clinic appointment following day case treatment.

Patient who frequently choose not to attend outpatient visits or those who have not attended within the last 12 months should be evaluated by the medical team at presentation to the Day Unit for pain management and prior to any treatment, blood investigations or prescriptions, however Nurse led evaluation and investigation should be continued in the interim.

5.6 Challenges/concerns

1. The nurse led acute pain service for sickle patients has been operating for over 2 years and has been well managed following extensive training and support of the nursing staff on the Haematology Day Unit.. In order to ensure continuing education for new staff and updates for nursing staff, on the acute management of SCD and its myriad complications the sickle cell team will continue to provide teaching sessions and regular updates. New members of the haematology nursing team should spend time with the sickle cell clinical nurse specialists during their induction so that they can be introduced to the condition, the pain service and admission processes.
2. As the day unit nurses assume responsibility for the initial assessment, investigations and analgesia administration for patients, they will continue to be supported by both the sickle cell clinical nurse specialists and the sickle cell medical team where advice is required or patient review sought to decide on future management.
3. As all patients attending the unit will be triaged by phone it is unlikely that a large number will attend simultaneously, however in this unlikely scenario: Any patients who have not been telephoned triaged can be asked to attend A&E *IF* no space is available on the unit appropriate for their needs and transfer is deemed safe (Please discuss all such patients with a member of the sickle CNS or Medical team before transfer). Paramedic ambulance crews are able to transfer these patients, to the A&E department if required.
4. Patients with unresolved pain (score >7 at 5pm) should have admission discussed with them and a bed organised via the bed manager. As patients will only be accepted on to the unit until 4pm, this should allow time for analgesia and reviewed for discharge by 6pm. Staff will be encouraged to make early decisions about whether or not a patient needs hospital admission to allow early liaison with the Bed Manager. All patients will be admitted to Guy's, but if there are no beds they will be transferred to an inpatient bed at St Thomas' via an ambulance. Paramedic ambulance crews are able to transfer these patients, even if they have received opiates.
5. Where there are concerns about a patient's behaviour or frequency of their visits this should be raised with the sickle cell team. Any patient attending for a third time within 2 weeks should be referred for medical review. In addition frequent attenders will be discussed at the monthly sickle team MDM.

Transfusions and Haemoglobinopathies

6.0 Blood Transfusions

There are about 50 patients with SCD, thalassaemia or other inherited anaemias who require regular blood transfusion therapy. In addition other patients may require ad hoc transfusion for particular medical indications e.g. pre-operative. Patients will only be started on regular transfusion after discussion with the sickle team in clinic. The blood transfusion protocols are usually nurse led but a medical review should be requested if there are any nursing concerns and in all cases if the patient has not attended the out-patients clinic for over 6 months. All blood transfusions will be prescribed by the day unit medical doctors or the sickle team doctors.

6.1 Procedure for transfusion

The patient will attend the Day Unit for a cross match and pre transfusion haemoglobin 24-48 hours before the planned date of transfusion. Patients new to the service will require extended

phenotyping as well as two group and save (G&S) samples taken at separate times and by different people. The nurse performing venepuncture should order blood units by requesting a crossmatch (CXM) on EPR (the number of units will be subject to haemoglobin result and transfusion type - see 6.2 and 6.3) and record the date of transfusion in the Day Unit diary.

Blood tests should also be taken as indicated for:

- Full blood count (HbS %) each transfusion (not thalassaemia patients)
- Renal function (each transfusion)
- Liver function (each transfusion)
- Bone profile (3 monthly)
- Ferritin (3 monthly)
- Virology (yearly: see sickle cell adult guidelines))
- Glucose (yearly)
- Thyroid function (yearly)
- Sex hormones (yearly: tests for males and females differ see adult sickle cell guidelines)

The nurse will check haemoglobin results prior to ordering the transfusion units from the blood bank, (for regularly transfused patients please refer to the individual patient transfusion folder for target haemoglobin to guide blood ordering if on top up transfusions, all ad hoc transfusions should have the number of units required clearly documented in the medical notes by the physician requesting the transfusion). Once the required numbers of units are confirmed to be ready with the blood bank, the nurse will complete and print the blood request form on EPR and collect the required number of blood units. Blood should be stored in cold boxes and the qualified nurse responsible, must ensure:

- the patient is appropriately consented,
- that the blood is correctly prescribed,
- the blood units are collected, administered, correctly
- should ensure that the patient is monitored as per GSTT Blood Transfusion Policy.

The details of the transfusion must be entered into the patient's notes (and the transfusion folder if the patient is on a transfusion program).

For cardiovascularly stable patients requiring transfusions, do not exceed the maximum infusion rate of 5 ml/kg/hr.

Extended phenotype matched (Full Rh and Kell typed), sickle negative blood should be used.

6.2 Simple Top up

Patients with B thalassaemia major or other inherited anaemias will have simple top up transfusions. They may also be required in patients with SCD when the haemoglobin falls more than 20g/l below baseline or patients have symptomatic anaemia. Each patient with B thalassaemia major has a folder which indicates the rationale for transfusion therapy, frequency of transfusion and haemoglobin targets, there is a section within the folder where results for each transfusion should be recorded by the nurse completing the episode of care.

The pre-transfusion Hb, target Hb and transfusion frequency will be documented for each patient in their day unit folder and medical notes. For ad hoc one off transfusions this will be documented by the doctor making the transfusion decision in the patients' medical notes. The nurse undertaking the top up transfusion is responsible for recording blood investigation results in the patient transfusion folder and informing medical staff that targets set out within the folder are not met if this occurs.

6.3 Exchange Blood Transfusion

The majority of patients with SCD who are on long term transfusion regimens will have regular automated red cell exchanges. Indications are primary or secondary stroke prevention, recurrent pain or chest syndromes which do not respond to Hydroxycarbamide, and other indications as decided by the Haematology consultant. Frequency, target and duration of transfusion will be decided by the sickle team. Emergency exchange transfusions may sometimes be needed for

patients acute complications of SCD on admission (e.g. acute chest syndrome, a new CVA, fulminant priapism or other life-threatening complication). Elective exchange transfusion/top up transfusions may be required for major surgery and during complicated pregnancies. Pregnant sickle patient who are less than < 16 weeks gestation requiring planned exchanges will have them completed on the haematology day unit. However once the patient reaches greater > 16 weeks gestation they should be exchanged on the birth centre at St Thomas' hospital and the procedure coordinated by the sickle cell clinical nurse specialists

Each patient on a regular program of exchange or top up transfusions has a transfusion folder kept in the day unit, which indicates; the rationale for transfusion (top up or exchange), haemoglobin, haematocrit, sickle cell percentage (HbS%) target and frequency of transfusions. The nurse completing the transfusion is responsible for recording results such as pre and post haemoglobin and HbS% in the patients folder. If the patient does not achieve the desired haemoglobin, haematocrit or S% this should be communicated to a member of the sickle medical and nursing team for guidance and the plan recorded within medical notes.

6.3 Late patients and cancellations

Patient can have automated exchanges booked for 830 or 1230 slots Monday to Friday (excluding bank holidays) late attendance may result in postponement of the procedure if appropriate but the patient must be discussed first with a member of the medical and nursing team prior to cancellation and rebooking

6.4 ACCESS /CVAD /SEDATION

Exchanges can be completed by peripheral cannulae if venous access is suitable and has been assessed as so by senior member of the haematology medical nursing team. Patients without suitable access will require femoral vascath insertion and this should be organised with the oncology and haematology CVAD team. Some patients on exchanges have y double lumen Portacaths or renal fistulas in situ via which they can be exchanged; these MUST only ever be accessed by nurses who have been assessed as competent to do so. SCD patients with portacaths in situ should not have these devices accessed unless it is by a trained member of the day unit staff or one of the sickle clinical nurse specialists. Portacaths must be flushed post procedures to maintain patency with Heparin saline (please refer to GSTT policy for maintenance of patency of intravenous access devices).

For GSTT clinical guidance regarding sedation and insertion of femoral lines please refer to: Insertion of central venous access device: femoral lines by nurses in the oncology & haematology directorate.

No emergency exchange transfusion should be initiated without consultation with the Haematologist on call.

The Standard Operating Procedure for Exchange Blood transfusion and the Trust protocol for Manual Exchanges should be followed (both available on the intranet).

Patient information leaflets for top up and exchange transfusions are available on the intranet.

6.5 Follow up after transfusion therapy

Once the transfusion is complete, and the patient is stable, they should be discharged home. A discharge letter should be sent to the GP (Appendix 4). For regular transfusion patients the nurse undertaking the transfusion must discuss the date of the next transfusion with the patient prior to discharge and also book them into Haematology clinic if they have not been reviewed in clinic for 6 months. If a one-off transfusion, the nurse completing the procedure should confirm the patient has a follow up appointment in Haematology clinic. When a patient fails to attend for transfusion please inform the sickle CNS team, even if they call to reschedule,

7.0 Psychological Care

Patients with chronic illness such as sickle cell disease have emotional reactions, concerns and psychological difficulties that can have a huge impact on their health. SCD pain is

multidimensional in nature and therefore psychological variables such as perception of control, the meaning of the pain to the patient and dysphonic moods such as anxiety and depression influence the severity and frequency of pain. Psychological interventions such as cognitive behavioural therapy (CBT) are very effective adjunctive pain control therapies (Thomas *et al*, 1999). The sickle cell team offers a variety of health psychology support services for adolescents, adults and their families. These services are facilitated by health/clinical Psychologists who are trained to help people manage pain and emotional concerns. The Health Psychology services offer the following services to all patients with haemoglobinopathies:

Cognitive Behavioural Therapy

Appointments are offered for patients and family members.

Support Groups

The service offers to patients, families and carers affected by SCD the possibility of attending the SCD Support Groups, which are facilitated by a health Psychologist. The 1 ½ hour weekly sessions which are held every Tuesday evening, run throughout the year. The objectives of these groups are:

- To introduce psychological concepts and help participants manage stress
- To reduce fears, anxiety and a sense of isolation
- To link people suffering from SCD, and to share pertinent issues, ideas, information and experiences.

Drop-in Service

The psychologists also offer a drop-in service in which the patient can come in to see one of the psychologists without an appointment. The drop-in service runs every Wednesday between 2:00 – 3:00pm and is based on a first come first serve basis.

Neuropsychology Service

Patients with SCD have increased risk of stroke and silent stroke. Neuropsychological assessment can help to identify cognitive impairment and lead to specific recommendations for patients and staff in order to improve coping with daily life, communication, concordance with treatments, and making decisions about treatments.

Any patients identified as needing Psychology input must be referred to the Psychology department on 82725/82718 (Mon – Fri 9am-5pm).

Staff support

Psychologists offer support for staff on an individual or group basis. If a member of staff has had a difficult encounter with a SCD patient- or any haematological patient groups the psychology team are there to help you debrief from the situation and to help you consider effective solutions for moving forward.

You can contact the psychologists on 82725/82718

8.0 Community Referrals

Certain aspects of patient care are best addressed in the Community. Patients should be made aware to the relevant voluntary organisations and primary care specialist nurses and referred if they wish to be supported in the community. Community based support can include home visits, support with housing and other social issues.

The decision to refer a patient to the community team is made following patient assessment and after discussion with the patient. Verbal consent from the patient is necessary to pass their details on to the Community teams. A referral form is completed and sent to the community team who will contact the patient after discharge to arrange a home visit.

Contact details:

South East London Sickle Cell and Thalassaemia Centre, Wooden Spoon House
5 Dugard Way, Kennington, London, SE11 4TH
0203 049 5993/0203 049 5989/0203 049 6069/0203 049 6085

9.0 Multidisciplinary Meetings (MDM)

A monthly multidisciplinary team (MDT) meeting (a group of professionals from differing disciplines meeting to make decisions regarding treatment of individual patients) is held by the sickle team to facilitate patient. This Multidisciplinary meeting (MDMs) is an appropriate forum to discuss patients with complex or challenging issues aiming for an agreed management approach.

Composition of the Sickle cell MDT:

1. Consultant Haematologists and junior medical team
2. Clinical Nurse Specialists and Advanced Nurse Practitioner
3. Community sickle cell nurses
4. Psychologists
5. Day Unit nursing staff
6. Affiliated health care professional as required (e.g. pharmacist, chronic pain teams)
7. When relevant other medical consultants or patient's GP

The following are examples of issues that can be referred to the MDT from the day unit for discussion

- Frequent presentations to either the Day Unit or admissions with sickle cell crises
- Patients who have poor coping strategies when dealing with their disease
- Patients who comply poorly with treatment such as not attending for transfusion appointments
- Any other concerns.

This list is not exhaustive any member of the sickle cell team can refer a patient to the MDT provided the areas of concern/for discussion are clear.

When necessary and appropriate patients may be invited to the MDTs; in this case the format is that of a case conference. Minutes of a case conference and its outcomes should be documented in the patient notes, and a copy sent to all present at the MDT and the patient's GP.

10.0 Training and awareness

These guidelines apply to all qualified nursing staff providing direct patient care in the Day Unit. The contents of the guidelines will be discussed in team meetings and all new staff will be given the opportunity to familiarise themselves with the policy as part of their induction into the service. Nurses providing care to patients with haemoglobinopathies should have an awareness of the following documents.

- Standards for the clinical care of adults with sickle cell disease in the UK
- Standards for the clinical care of children and adults with thalassaemia in the UK
- A sickle cell crisis? A report of the National Confidential Enquiry into Patient Outcomes and Death
- Nice Guidance : Sickle cell acute painful episodes: management of an acute painful sickle cell episode in hospital
- Royal College of Nursing RCN Competences: Caring for people with sickle cell disease and thalassaemia syndromes: A framework for Nursing Staff

10.1 Monitoring

Compliance to the operational policy will be monitored by audit and record keeping. This can be identified with minutes of MDT meetings, prescription records, drug and blood monitoring tools.

10.2 Review

This policy will be reviewed in two years time. Earlier review may be required in response to exceptional circumstances, organisational change or relevant changes in legislation of guidance.

10.3 References

- Benjamin, L., Swinson, GI and Negal R. (2000) Sickle Cell Anaemia Day Unit: An approach for the management of uncomplicated painful crisis. *Blood*, Vol. 95, 4 (1130-1137).
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- Blood and Blood Products Transfusion Policy (GSTT).
- DOH (2007) Raising the Profile of Long term conditions care: A compendium of information
- GSSFT Medicines Administration Policy.
- GSTFT Insertion of central venous access device: femoral lines by nurses in the oncology & haematology directorate.
- (GSTFT). Sickle cell disease adult guidelines.
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- Nice (2012) Sickle cell acute painful episodes: management of an acute painful sickle cell episode in hospital, National Institute for Health and clinical Excellence 1-25.
- RCN (2004) Adolescent Transition Care. Guidance for Nursing staff.
- Royal College of Nursing Competences: (2011) Caring for people with sickle cell disease and thalassaemia syndromes: A framework for Nursing Staff.
- Standards for the clinical care of children and adults with thalassaemia in the UK (2008) United Kingdom Thalassaemia society.
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- Ware, M.A, Hambleton, I, Ochaya, I and Sergent G.R (1999) Day Care Management of Sickle cell painful crisis in Jamaica: A model applicable elsewhere? *BMJ* vol. 104 (93 -96).
- Wright J, Bareford D, Wright C, Augustine G, Olley C, Musumadi L, Dhanda C and Knight C (2004) Day case management of sickle pain: 3 years experience in a UK sickle cell unit. *British Journal of Haematology*. 126, 878-80.

10.4 Appendices

- Appendix 1: Day case Pain assessment chart
- Appendix 2: Vital observation and pain assessment chart
- Appendix 3: Discharge information leaflet
- Appendix 4: Discharge summary to GP
- Appendix 5: Acute Episode Guideline Summary

Appendix 1:

NURESE LED PAIN SERVICE FOR SCD PATIENTS ASSESSMENT FORM

Patients Name:	Hosp #
Sex M / F (circle)	DOB

Date/Time of arrival to day unit:

<p><u>Site of pain:</u></p> <p>Description of pain: (tick) _</p> <ul style="list-style-type: none"> • Sharp • Burning • Throbbing • Shooting • Aching • Stabbing • Sore • Crushing • Other 	<p><u>Duration of pain:</u></p> <p>Precipitant/Triggers: (tick)</p> <table border="1"> <tr> <td> <ul style="list-style-type: none"> • Infection • Dehydration • Cold weather • Hot weather • Stress • Physical activity • Other </td> </tr> </table>	<ul style="list-style-type: none"> • Infection • Dehydration • Cold weather • Hot weather • Stress • Physical activity • Other
<ul style="list-style-type: none"> • Infection • Dehydration • Cold weather • Hot weather • Stress • Physical activity • Other 		

Analgesia taken in the last 8 hrs:.....

Exclusion Criteria: All patients must be telephone triaged and referred to A+E if appropriate. However, if they present unexpectedly to the Day Unit, they should have urgent medical review, and may require admission or transfer to St Thomas' dependent on clinical scenario. Patients with **ANY** of the below should be referred for medical review

- Chest pain, Shortness of breath, Hypoxia (oxygen saturations <94%)
- Fever/rigors (Temp >38°C), Hypotension (BP <90/60)
- Tachycardia > 100 (even after pain has settled following analgesia)
- Raised respiratory rate of > 20 (even after pain has settled following analgesia)
- New neurological symptoms: headache, confusion, numbness of limbs
- Abdominal pains
- Priapism (persistent erection)
- Pregnancy
- Visual loss or bleeding in the eye
- PAR > four
- Concerns from the nursing team about the patient's clinical condition

IMPORTANT: In the event of a clinical emergency: PAR score > 4 contact Acute Care Team on bleep 1162 or the CRASH team on 2222. Once decision is made to transfer to STH, call LAS (urgent escort) on 0207 827 4597

Pain assessment chart for Sickle Cell Patients

<u>Nurse's notes</u>	
<p>Time of 1st dose analgesia: Unit: (Re-assess pain after 30mins)</p>	<p>List of drugs given in the Day</p>
<p>2nd Assessment: (assess patient responses to analgesia, fluid in take etc)</p>	
<p>Investigations:</p> <p>FBC –retics <input type="checkbox"/></p> <p>LFT's <input type="checkbox"/></p> <p>U&E's <input type="checkbox"/></p> <p>LDH <input type="checkbox"/></p> <p>Other..... (Blood cultures CRP , Xray, X-match if indicated)</p> <p>Dipstick/MSU <input type="checkbox"/></p>	

<u>Doctor's notes</u>
<p>Time:</p>

Decision to admit/Discharge:
<p>To organise a non-urgent bed; Bleep 1165 for Guy's site practitioner or 0162 for St Thomas'</p>

Admission checklist

- Bed requested; yes/no
- Ward informed: yes/no
- Next of kin informed: Yes/no
- Medical notes/ xrays: Yes/no
- Blood samples: Yes/no
- IV access yes/no

Discharge checklist

- Medical review: yes/no
- TTO: yes/no
- GP letter: yes/no
- OPA made: yes/no
- Fax to Haem. Consultant yes/no
- Community referral yes/no
- Psychology referral: yes/no

Discharge information

Date.....

Dear Patient,

Following your treatment today,

- You will be given a small amount of tablets to take home to ensure that you are comfortable once you have been discharged. Your Day Unit nurse will go through the medications with you to ensure you understand how to take them.
- You are advised to continue taking your pain killers at home as required following the instructions given.
- If your pain worsens once you have left the Unit, you may return to A/E to be reassessed or return to the Day Unit the following day (Between 09.00 – 16.00hrs)
- You are encouraged to access the health psychology service to gain additional support around developing effective coping strategies which are often of great benefit to many patients. Speak to your Nurse during the course of your treatment on the day unit to arrange for Psychology service.
- For your own safety, you are strongly advised **NOT** to drive if you received Opiate pain killers today, such as Morphine, Oramorph Oxycodone or Oxynorm. These medicines cause sedation and drowsiness which may alter your perception and judgement while on the road. Please plan ahead and talk to the nurses to help to arrange for someone to collect you.

If you have any questions about the contents of this letter, please talk to the staff on the Day Unit who will be very happy to help.

Yours sincerely,

The Sickle Cell team

Appendix 4:

Discharge Summary for Day Case Treatment

GP Details	Pt's details
<p><u>Consultant</u> <u>Jo Howard / Rachel Kesse Adu</u></p>	<p>Seen by <u>nurse/senior house officer/registrar/other</u>..... Please circle as appropriate.</p>
<p><u>Date/s of treatment</u></p>	
<p><u>Past medical history</u></p>	<p><u>Presenting complaint/treatment</u></p>
<p><u>Treatment administered</u></p>	<p><u>Medication/dressings to take home</u></p>
<p><u>Outpatient appointment or follow up plan</u></p>	<p><u>Investigation results</u></p>
<p>Form completed by</p> <p>Copy form to GP ✓ Copy of form Dr Howard Secretary ✓</p>	<p>Any other information</p>

Acute Episode Guideline Summary

Initial Assessment

- Triage as high priority
- Measure O2 sats
- Give 28% O2

Quick assessment for presence of

- infection,
- dehydration,
- acute chest syndrome
- neurological events
- severe anaemia
- priapism
- splenic enlargement,

Care plans / Behavioural Contracts

- Patients with complex have individual care plans which are kept in the sickle cell folder in A&E Majors. *Please refer to these.*
- When transferring the patient to the ward please place a photocopy in the notes.

Complications and clinical issues in Sickle Cell Disease

Acute chest syndrome:

- LEADING CAUSE OF MORTALITY IN ADULTS WITH HbSS
- PLEASE CONTACT HAEMATOLOGY REGISTRAR / REGISTRAR ON CALL AND INFORM HDU/ ITU IF STRONG SUSPICION

• **Features:**

- Fever, Cough, Chest pain
- Respiratory distress/ hypoxemia
- New opacity on chest x-ray
- Worsening anaemia
- Bilirubin stained sputum
- Antecedent painful crisis

Please refer to the section on Acute chest syndrome for further details

• **Priapism:**

- This is Painful penile erection
- Urgent urological and haematological review is necessary.
- Ensure adequate hydration and analgesia
- Encourage patient to empty bladder
- Do not pack the penis in ice
- If Priapism resolves and patient is on Cyproterone please restart as prescribed

Abdominal pain / sickle girdle syndrome:

- sickle girdle syndrome: pain radiating from the vertebral column
- sickling in abdomen: tenderness, rigidity and ileus; can mimic peritonitis
- abdominal pain can also indicate cholecystitis, peritonitis, Obs&Gyn problems
- IMPORTANT: please contact haematology Registrar on call when

Pain Management

- Start analgesia within 30 mins of coming into A&E
- Ascertain amount and type of analgesia used at home
- Check individual health care plan with individual analgesia protocols kept in the Sickle Cell folder in A&E majors.

Managing Pain in Opioid-naive patients:

- Clinical assessment of severity of pain
- Oral analgesia according to the WHO 3 step ladder (see appendix)
- Give Morphine only if pain is severe and / or oral analgesia ineffective
- Morphine dose 5-10 mg po/sc loading dose and titrate against response
- Add Paracetamol, ibuprofen / diclofenac (CAUTION: sickle nephropathy)
- Monitor pain, sedation, vital signs, respiratory rate and O2 sats every 30 mins. Then every 2 hrs once pain is controlled and patient stable.
- CAUTION: if respiratory rate <12/min, omit morphine dose and give Naloxone 100 mcg IV and repeat if appropriate.
- Consider adjuncts:

antipruritics: Hydroxyzine 25 mg bid po
antiemetics: Cyclizine 50 mg tds

Investigations:

❖ *All patients need:*

- Full blood count
- Urea, creatinine & electrolytes

❖ *Other investigations to be considered:*

- Blood group and phenotype if patient requires transfusion
- Haemoglobin electrophoresis whenever a new sickle cell patient presents and the diagnosis is not known to GSTT

❖ *Non routine tests directed by specific clinical problems:*

- Chest X-ray (febrile, breathless, chest pain, reduced O2 sats)
- Arterial blood gases (O2 sats <85%, unexplained drowsiness)
- Liver function tests (jaundice, abdominal pain)
- Reticulocytes (Hb more than 1 gm lower than baseline)
- Blood and urine cultures (febrile)
- Ultrasound abdomen (abnormal LFTs, abdominal pain)
- Parvovirus B19 serology (reticulocytopenia)
- Brain imaging (seizure, TIA, stroke, severe headache)
- Limb X-rays (trauma, persistent unexplained swelling)

Supportive management:

- Folic acid: all patients should get it. 5 mg od po
- Hydration: encourage patients to drink 2-3 litres in 24 hours. Use IV fluids only if the patient is dehydrated or unable to drink orally (e.g. nil by mouth). Preferred solution is Hartmann's solution 3L/ day
- Oxygen: Give humidified oxygen at 2-4 l/min by mask or nasal cannula if O2 sats < 95%
- Thromboprophylaxis: Enoxaparin 40 mg sc od if patient immobile and being admitted

Neurological events:

- Transient ischaemic attacks or strokes occur in up to 25% of patients with SCD
- Please discuss with neurology and haematology teams
- Urgent neuro-imaging is needed because ischemic lesions may require urgent exchange transfusion

Infection and sepsis:

- Patients are particularly prone to pneumococcal & other infections
- If temp > 38°C take blood cultures, septic screen and start antibiotic therapy according to the likely focus of infection.
- If prescribing antibiotics at therapeutic doses discontinue regular prophylactic antibiotics
- If sickle patient is pyrexial and has recently travelled to a malaria region of the world then test blood for malaria
- If there is a drop in Hb of >2gm/L from baseline & reticulocyte count is low / low normal please request Parvo virus titres
- Hydroxycarbamide should be stopped if the patient is febrile or platelet count < 100 x 10⁹ /L, or neutrophils < 1 x 10⁹ /L

Pregnancy:

- Increased risk of pre-eclampsia, maternal and neonatal mortality and anaemia
- Check FBC & retics, U&Es and Group & save
- Contact haematology registrar on call
- Women >16 weeks of gestation are admitted on obstetric HDU

Day Unit Flow Chart (see appended word document)

