Sickle Cell Disease and Thalassaemia: Outpatient Clinic Review and Monitoring

Profile	
Version:	*V3.0*
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Executive/Divisional sponsor:	*Dr Lisa Pickering, Divisional Chair* *DGB February 2019*
Applies to:	*All staff involved in the care of patients with Sickle Cell Disease and thalassaemia*
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Approval	
Approval person/Committee:	*MedCard Divisional Governance Board*
Date:	*February 14th 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 of Adults 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

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

Sickle cell disease (SCD) and thalassaemia are lifelong conditions with multiorgan involvement. Outpatient visits provide a key opportunity for assessment and monitoring and enabling self-management.

1. Introduction

Sickle cell disease (SCD) and thalassaemia are lifelong conditions with multiorgan involvement. Outpatient visits provide a key opportunity for assessment and monitoring and enabling self-management. This guidelines provides the guidance to ensure appropriate review of patients new to the service and their annual review. For specific guidance on organ involvement, chelation and hydroxycarbamide or management during pregnancy please see the relevant guidelines

2. Status and Purpose

Delete as appropriate:

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 Clinic Processes

- There is a weekly multidisciplinary clinic for haemoglobinopathy with medical nursing and psychology staff. This is held every Wednesday afternoon from 2-5pm
- Additional psychology clinic lists are held during week and psychology book their own appointments
- Referrals for medical clinics come via GP, other hospital providers, or as a follow up from an inpatient stay. They will be triaged via the haematology department and booked in appropriately. There is an SOS slot available at 1.45 which can be booked in discussion with the haematology consultant
- Changing appointments by SGH staff can be done by contacting the haem-onc schedulers to ask for rearrangements: haem.oncschedulers@stgeorges.nhs.uk
- On arrival all patients should be asked to confirm their contact details and address
- All patients with have weight, blood pressure, oxygen saturations measured and be asked to provide a urine sample which is dipsticked. Results of all of these are written in medical notes by clinic nursing staff.
- Each week there is a pre clinic meeting to discuss each patient – identify specific requirements (eg new patients, those post transition) and identify those who will need annual review including psychology.
- Patients should be offered the opportunity to sign up to the National Haemoglobinopathy register if they haven't already and be offered patient information on that.

6.2 New Patients

When a patient attends their first appointment in the adult services (assuming not via the transition process) that clinic appointment is longer and the following points should be covered and can be considered as a check list for helping the consultation. Patients with SCD or thalassaemia have a lifelong condition and should therefore remain under review at a specialist haemoglobinopathy centre. They should be offered an annual review as a minimum.

- Introduce the clinical team (Consultant, CNS, Clinical Psychologist)
- Confirm diagnosis if needed with Hb electrophoresis HbS, A2 and F estimation. Check also FBC, reticulocyte count, renal and liver biochemistry and LDH. The blood group, red cell phenotype and G6PD level should also be checked.
- Take full personal and family history (including medical, surgical and transfusion history)
- Check patient's understanding of their diagnosis, explain further and answer any questions
- Check immunisation status: especially pneumovax, HiB vaccine, meningococcal vaccine, hepatitis B vaccine and annual influenza vaccine – advise on vaccinations that are now required.
- Review the need for any up to date screening investigations to be done: blood pressure and oxygen saturations, echocardiogram, urinalysis for protein, and any ophthalmological symptoms requiring review.
- Discuss potential acute complications and how to seek medical attention and help
- Discuss implications of travel (air flight, anti-malarials)
- Discuss any family planning issues and offer genetic counselling as required
- Advise GP to prescribe penicillin V 250mg *bd* (if not allergic) and folic acid 5 mg *od* on repeat prescription
- Ensure has an appointment in the haemoglobinopathy clinic for their annual review
- Ensure patients and their families have contact details for the hospital, the day unit, the clinical nurse specialist, their community team and the consultant's secretary as needed.
- Check that the patient is known to a GP
- Inform the community team of the patient and arrange follow up in the community if appropriate
- Patient to be discussed at haemoglobinopathy MDT

6.3 Sickle Cell Disease, Annual Review

Patients with sickle cell disease (HbSS, HbSC and other compound heterozygotes causing a sickling disorder) should be reviewed in a specialist sickle cell clinic at a minimum of once a year for an annual review. Many patients will be seen more frequently depending on organ involvement, pain issues, pregnancy plans and social issues.

The annual review template is available on the intranet website and in paper form in clinic and at the appendix of this chapter

The annual review should be a documented clinic meeting that assesses effects on the following aspects of life with sickle cell disease.

- Baseline bloods and markers of haemolysis
- Blood pressure
- Echocardiogram and pulse oximetry screening for pulmonary hypertension
- Urinalysis to screen for proteinuria and underlying sickle nephropathy
- Visual symptoms and any ophthalmological concerns
- Transfusion history, antibody levels, ferritin levels, relevant imaging – to screen and monitor for iron overload and chelation plans
- Admission history
- Episodes of acute pain
- Chronic pain and it's management at home
- Analgeisia regimens
- How pain is affecting daily life – eg missing work / college / effect on relationships
- Family planning and sexual health review
- Vaccination and antibiotic use
- Psychological and mental health issues
- Cognitive or memory concerns

All patients attending for medical annual review will be offered a psychology annual review – this entails

- Completing a questionnaire based on anxiety and health concerns
- Meeting with psychologist
- Written report as part of the annual review to the patient and GP with results and plans for follow up if needed

The annual review should be documented on the pro-forma or in the notes or both and clinic letters. Patients receive a copy of their annual review and this forms their written annual care plan.

Where appropriate their emergency department care plan should also be updated and the patient offered a paper copy of that.

Further monitoring or investigation of abnormal results flagged up at the annual review should be referred through to specific consultants and specialities who have an interest in sickle cell related issues.

There is an annual review template available on the intranet website, on the L drive and in paper form in clinic.

6.4 Patients with TDT : Outpatient monitoring and Annual Review

By definition all patients with TDT will be transfusion dependent from early childhood, often needing transfusion every 2-4 weeks.

The mainstay of monitoring is 2 fold

i) To ensure the pre-transfusion Hb is above 95g/L to ensure adequate growth, bone health and to reduce extramedullary haemopoiesis

iI) To deliver good chelation – compliance and monitoring to prevent complications from the side effects of regular transfusions

All patients with TDT (β Thalassaemia major) should have regular outpatient clinic and an annual review to

- discuss the transfusion programme and any problems with the programme
    - access and blood tests
    - pre transfusion Hb and frequency of transfusions
    - red blood cell consumption
    - any suggestion of extramedullary haematopoiesis
- discuss ferritin and chelation
- ensure cardiac and endocrinology and rheumatology follow up are as needed
- any family planning issues are discussed
- all imaging and tests are up to date
- psychological and mental health issues are identified and addressed
- offered a psychology annual review
- discussion of trial options

Dedicated thalassaemia clinics are held every 6 months. One of these clinics is the annual review clinic held jointly with cardiology for a cardiac annual review at the same time.

There is a formal TDT annual review template available on the intranet website, on the L drive and in paper form in clinic.

6.5 NTDT Outpatient Monitoring.

Patients with thalassameia intermedia / HbEβthalassaemia may not need regular blood transfusions but still need regular review, monitoring of end organs and special considerations including transfusion at certain periods of their lives including surgery and pregnancy. They are also still at risk of iron overload due to their ineffective erythropoeisis

They should be monitored through outpatients for

- Adequate growth and development and fatigue levels
- Baseline Hb and state of haemolysis
- Spleen size and evidence of hypersplenism - Splenectomy should only be considered in patients with severe cases of massive splenomegaly or hypersplenism – increased risk of thrombosis and pulmonary hypertension post splenectomy
- Prophylactic antibiotics and vaccinations in those who have been splenectomised
- Monitoring for non-transfusion iron overload including T2* weighted MRIs (every 2- 5 years or more frequently if abnormal) and establishing a chelation plan if evidence of iron overload
- Screening for pulmonary hypertension (echocardiograms every 2 years unless abnormal)
- Bone health: vitamin D monitoring and supplementation, DEXA scans (every 5 years)
- Family planning issues and difficulties
- Cardiology and endocrinology assessments as needed
- Consideration of a transfusion programme (regular transfusions may be less of a concern as chelation is now more effective at reducing iron overload) – Indications for transfusion include:
    - Severe anaemia
    - Reduced exercise tolerance
    - Hypersplenism
    - Evidence of extramedullary haematopoiesis
    - Reduced growth and development
    - Prior to surgery (possibly for weeks-months before surgery)
    - Pregnancy (anaemia, symptoms, reduced fetal growth)
    - Pulmonary Hypertension risk reduction
    - Thrombotic risk reduction

Patients not only should be monitored for iron overload but they should be made aware of the possibility and need for chelation despite infrequent transfusions.

St George’s Healthcare NHS trust has a very small cohort of patients with thalassaemia intermedia and so referral to the quaternary service at the Whittington Hospital should be considered for review, joint management or advice where necessary or discussion of clinical trials.

6.6 Multi-Disciplinary Team Meetings

There are weekly multidisciplinary team meetings held. They are attended by haematology consultant, clinical nurse specialist, clinical psychologist, community nurse specialist for Wandsworth and representatives from the At specific times ward nursing staff attend to discuss specific patients.

Patients discussed

- Inpatients
- Pregnant patients
- Patients of interest or concern
- Follow up of patients previously discussed
- Referrals to psychology
- Referrals to red cell pain management service
- Patients going through transition
- New patients to the service
- Any patients who have needed a critical care admission
- Patients planning or undergoing surgery
- Any deaths

There are monthly MDTs held with the obstetric team with the medical haematology team to discuss all pregnant sickle cell patients

There are quarterly MDTs with renal and neurology and the apheresis service

There are MDT opportunities with the Whittington Hospital thalassaemia team

6.7 Patients who are being considered for Bone Marrow (HSC) Transplant (With Dr Mickey Koh)

Bone marrow or Haematopoeitic Stem Cell transplantation is not currently funded for adults with SCD though there is increasing experience and improving outcomes seen in adult patients now with non-myeloablative conditioning.

The principle is that patients who have suffered complications of sickle cell that mean they may benefit from the risk of a HSCT whilst being well enough for the procedure should be considered. Patient's whose complications would usually mean a lifelong transfusion programme may be appropriate to consider for transplant. There needs to be an available donor and this is usually required to be a matched sibling who does not suffer from SCD. Funding will need to be secured on individual cases

Haplo-identical donor and Matched Unrelated Donor stem cell transplants in adults with SCD should be undertaken as part of a clinical trial

Indications for consideration of transplant may include

- stroke, abnormal TCD, neurological involvement – including silent infarcts with cognitive sequelae
- recurrent ACS
- recurrent vaso-occlusive crises despite HC or unable to take HC
- pulmonary hypertension
- sickle nephropathy

Patients who have expressed an interest or who clinicians may feel may be appropriate should be

discussed first with the clinical lead for transplantation at St George's University Hospitals NHS Foundation Trust, Dr Mickey Koh. They should also be discussed at a regional MDT. He will then arrange for review of the case and clinic appointments as necessary

Throughout the process stringent psychological input will be required.

6.8 Patients Who Do Not Attend

Patients who do not attend either outpatient clinics or planned transfusions should be discussed with the consultant, usually at the Multi-Disciplinary Team meeting or in the post-clinic meeting. Follow up of these patients may include phone contact or home visit by the community team or liaison with GP

Taking into consideration the complexities of the nature of SCD and thalassaemia our DNA policy is as follow

- Paitents who have recurrent admissions but fail to attend their outpatient clinic appointment will receive a discharge letter after 1 DNA with details of how to be referred back to clinic
- Active patients who regularly have attended clinics within the last 2 years will receive a letter after their 2nd consecutive DNA explaining risk of discharge and importance of clinic attendance and we will attempt to call on phone. After their 3rd consecutive DNA they will be discussed at MDT and then they will be discharged with a letter to GP and patient
- inactive patients not seen in last 2 years (either in clinic or inpatient stay) will be discharged with a letter to GP and patient. (and discussed at MDT)

6.9 Patients Who Move Out of Area

UNIVERSITY

Many patients may move out of area temporarily for university. Discussion with the patient before they move usually occurs at the outpatient clinic.

The patient will be given a copy of the most recent annual review which will include details of admissions, complications and pain protocol. The patient will be encouraged to inform the local medical team on arrival of his or her condition. Contact between the St George's nursing team and the local team can be offered.

Patients on a transfusion programme or receiving medication such as hydroxycarbamide or chelation will be seen in clinic before moving and plans for monitoring and treatment whilst away will be made (either to return regularly to our centre or to be referred to local centre for review)

Patients will be given all contact details for St George's team so they can contact us whilst away if any concerns arise.

PERMANENT MOVE OUT OF AREA

The patient will be given a copy of their most recent annual review which will include details of admissions, complications and pain protocol.

The nursing team will liaise with the nursing team of the new area if one exists to ensure early contact is made.

A full clinical letter will be sent to the receiving team.

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

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**
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*	*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*

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 of Adults 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