# Sickle Cell Disease:
# Chronic Complications (Ophthalmology)

| **Profile** | |
|---|---|
| **Version:** | *V3.0* |
| **Author:** | *Dr Elizabeth Rhodes, Consultant Haematologist*<br>*Dr Julia Sikorska, Consultant Haematologist* |
| **Executive/Divisional sponsor:** | *Dr Lisa Pickering, Divisional Chair*<br>*DGB February 2019* |
| **Applies to:** | *All staff involved in the care of patients with Sickle Cell Disease* |
| **Date issued:** | *February 2019* |
| **Review date:** | *February 2022* |
| **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)<br><br>• 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 |

| Please identify the key people you involved in reviewing this policy why, and when: |
|---|
| Miss C Egan – Link Consultant Ophthalmologist (2015)<br>Mr G Holder - Consultant Ophthalmologist (2015) |

| Summarise the key changes you have made and why: |
|---|
| Updates in line with the UK standards of care for SCD<br>Updated contact details |

## Executive Summary

The most common ophthalmic complication of sickle cell disease (SCD) is sickle retinopathy, caused by vaso-occlusion leading to retinal ischaemia and infarction. This guideline contains advice to support the management of patients with SCD with retinopathy and on screening for retinotoxicity in those on iron chelation.

## 1. Introduction

Sickle retinopathy is the most common ophthalmic complication of SCD and can be subclassified into non-proliferative and proliferative forms. It is characterised by mechanical obstruction of the retinal capillaries by sickling although endothelial damage may also contribute.

Patients with early stage retinopathy are generally asymptomatic but it can lead to vitreous haemorrhage and retinal detachment resulting in loss of vision. Drug-induced retinopathy can arise from iron chelating agents and so the detection of retinal toxicity is particularly important through retinopathy screening.

## 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 lifelong condition due to abnormal haemoglobin variant.

## 4. Scope

This guideline is relevant to the care of patients with SCD requiring elective and emergency surgery 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 to 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.

## 6.0 Content

**6.1 Ophthalmological complications**
**6.2 Opthalmological review when on iron chelation**

### 6.1 OPHTHALMOLOGICAL COMPLICATIONS

SCD can cause many problems within the visual system from vascular occlusions to problems in both the posterior and anterior segments. Patients with sickle cell/haemoglobin C compound heterozygosity have a higher risk of sickle retinopathy than patients with homozygous sickle cell anaemia. All patients with SCD should have baseline retinopathy screening.

Non proliferative disease includes conjunctival vascular occlusions, iris atrophy, retinal haemorrhages, retinal pigmentary changes and other retinal macula, choroid and optic disc problems. These may often leave little visual consequence.

Proliferative disease can be more significant and lead to neovascularisation and risk of vitreous haemorrhage and retinal detachment. For these reasons patients with SCD should be assessed clinically for signs of retinopathy – blurred vision, new floaters and should be referred urgently if there are new signs and for review if there are old signs to the ophthalmology department. Intervention, which may include laser treatment or surgery at this stage, can prevent deterioration.

There is no clear evidence base for the benefit of strict annual screening but patients should be asked about visual symptoms at each clinic appointment and given advice to seek urgent medical attention via the eye casualty department or the emergency department.

Patients undergoing significant ophthalmic surgery should be discussed with the haematology team so as to be able to plan for the procedure.

### 6.2 OPHTHALMOLOGICAL REVIEW WHEN ON IRON CHELATION

**Desferrioxamine**
Asymptomatic patients when starting desferrioxamine should have a baseline review including electrophysiology and retinal imaging prefably within 6 weeks of starting. They will also be seen in the medical retina clinic after the EDD for documentation of any retinopathy. These patients should have yearly electrophysiology and psychophysic investigations.

Symptomatic patients should be discussed with ophthalmology urgently as desferrioxamine can be retinotoxic. Symptoms are usually after a spell of high dose intravenous therapy. They will require urgent electrophysiology and psychophysic investigations with a full ophthalmological assessment for any abnormalities found.

**Deferasirox and Deferiprone**
There is less clear evidence of any retinotoxicity and so these patients should be monitored clinically and discussed with ophthalmology if there are concerns of any symptoms.

**Hospital Contact Details**

| | | |
|---|---|---|
| Dr Elizabeth Rhodes | Haematology Consultant | ext 0885 |
| Dr Julia Sikorska | Haematology Consultant<br>Haematology Registrar | ext 0885<br>bleep: 7080 |
| Carol Rose | Sickle Cell CNS | mobile: 07500835735 |

**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.*<br><br>*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 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