Sickle Cell Disease:
Chronic Complications (Neurological Conditions including Headaches)

Profile	
Version:	*V3.0*
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Executive/Divisional sponsor:	*Dr Lisa Pickering, Divisional Chair* *DGB Feb 2019*
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*
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Contents

Paragraph		Page
	Executive Summary	
	Policy Gateway	
1	Introduction	
2	Purpose	
3	Definitions	
4	Scope	
5	Roles and responsibilities	
6	Other headings as appropriate	
7	Implementation and dissemination	
8	Monitoring compliance	
9	Associated documents	
10	References	

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

Please identify the key people you involved in reviewing this policy why, and when:
Dr A Pereira (2015) - link Consultant for Neurology

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

Executive Summary

This document offers guidance on the management of chronic neurological complications of sickle cell disease. For guidance on acute neurological complications **see separate guideline for management of acute complications in sickle cell disease.**

1. Introduction

Sickle cell disease is an inherited red cell disorder that is characterized by chronic haemolysis and vascular occlusion, causing pain and end-organ damage. SCD has been associated with impaired neurocognitive function and may be related to small vessel vasculopathy and silent cerebral ischaemia. This guideline offers advice regarding when to seek advice from the neurology team and consider referral to neuropsychology.

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 Neurological complications
6.2 Headaches

6.1 NEUROLOGICAL COMPLICATIONS

Previous strokes, headaches, seizures and moya-moya disease are all neurological complications seen in patients with SCD. MRI/MRAngiogram (MRA) imaging is a useful tool to look for silent strokes and CT angiogram (CTA) for aneurysm formation. These patients should be managed in conjunction with the neurologists. In the event of an acute stroke or neurological event **see separate guideline for management of acute complications in sickle cell disease.**

Neurocognitive decline is recognised in patients with SCD and may be related to silent cerebral ischaemia and small vessel vasculopathy. Memory loss is not uncommon in these patients. Patients should be discussed at MDTs for possible referral to neuropsychology for full neuro-cognitive assessments.

Mood disturbance may or may not be related to SCD and after exclusion of organic causes these patients should be discussed at MDTs to ensure appropriate medical and psychological follow up and possible referral to neuropsychology for full neuro-cognitive assessment and advice on adaptation and adjustment, as appropriate.

6.2 Headaches

Most headaches in patients with SCD will be due to migraines or chronic daily headaches often associated with analgesia use (particularly codeine containing products).

Refractory headaches can be referred to the headache service at St George's University Hospitals NHS Foundation Trust. Medications that might be useful include Maxalt Melts for migraines, amitryptyline (low dose, 10mg, nocte) or topiramate (50mg bd) for daily headaches.

Sudden onset severe headaches, new severe headaches or those with associated clinical signs should be managed urgently and as for patients without sickle cell disease.

Hospital Contact Details

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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.* *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

Management of Acute Complications in Adult patients with Sickle Cell Disease

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