Sickle Cell Disease:
Chronic Complications (Cardiology and Respiratory Disease including Pulmonary Hypertension)

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*
Date issued:	*February 2019*
Review date:	*February 2022*
Approval	
Approval person/Committee:	*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 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
- American Thoracic Society guidelines on Pulmonary Hypertension in SCD http://www.thoracic.org/statements/resources/pvd/sickle-cell-disease.pdf

Please identify the key people you involved in reviewing this policy why, and when:
Professor Brendan Madden (2015) - Consultant Cardiothoracic Physician

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

Sickle cell disease (SCD) is the most commonly inherited disease in the UK. It is an autosomal recessive disorder that results in the production of abnormal haemoglobin that in turn leads to an abnormal shape of the red blood cells and chronic haemolysis. Clinically these patients suffer from progressive end-organ damage, including cardiopulmonary complications. This guideline contains advice on how to manage these complications in patients with SCD.

1. Introduction

Both the heart and lungs are affected by chronic sickling and need monitoring in the outpatient setting.

A previous isolated episode or recurrent episodes of acute chest crises can predispose patients to cardiac and pulmonary damage but it is clear now that some patients will develop problems due to background chronic sickling and haemolysis.

This guideline provides advice on the management of chronic cardiopulmonary complications in patients with SCD.

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 CHRONIC SICKLE LUNG DISEASE

Chronic sickle lung disease is often divided into 4 stages though it is not clear if an individual patient will progress through all 4 stages or when intervention or treatment is best. Patients with sickle cell disease will often complain of chest pain and shortness of breath and varying degrees of hypoxaemia are often found. There are a variety of different diagnoses, both acute and chronic, to be considered
Pulmonary and Cardiac monitoring and investigations:
Patients should be monitored and have documented baseline pulse oximetry at annual review as minimum with hypoxaeia confirmed on arrterial blood gas sampling if oximetry levels are low
If there is evidence or concern regarding hypoxaemia then the following investigations should be considered:

- Pulmonary function tests looking for restrictive picture seen in sickle lung
- HRCT to look for changes of interstitial lung disease
- Consider CTPA if concerns re thromboembolic disease
- Consider referral for sleep studies if concerns regarding nocturnal hypoxia

There is no clear, evidence based treatment plan for patients with sickle lung disease. General measures include:

- Good surveillance and avoidance of infections
- Smoking cessation support
- Involvement of respiratory / cardiac physician familiar with sickle cell disease
- Disease modification to be considered in the form of hydroxycarbamide or red cell transfusions

6.2 PULMONARY HYPERTENSION

The prevalence of pulmonary hypertension in patients with sickle cell varies in studies with a range in numbers seen from 5-30%. Moderate – severe pulmonary hypertension is associated with an increased risk of death. Screening and diagnosis are therefore important so that with close liaison with the pulmonary hypertension team treatment plans can be made.
Patients should be screened with an echocardiogram as a minimum every 2 years (and documented in their annual review paperwork.) The following actions should then be considered

TRV and RSVP	Symptoms?	Risk Stratification	Action
TRV <2.5ms and RSVP< 25mmHg	No	Low	Repeat echocardiogram in 1-2 years
TRV 2.5-2.9ms or RSVP 25-29mmHg	No	Low - Intermediate	Repeat echocardiogram in 1 year
	Yes	High – Intermediate	Refer to Pulmonary Hypertension team
TRV >3ms or RSVP >30mmHg	No	High – Intermediate	Repeat echocardiogram in 3-6 months. Consider referral to Pulmonary Hypertension team
	Yes	High	Refer to pulmonary hypertension team

The Pulmonary Hypertension team will investigate and perform right heart catheterisation (RHC)for further evaluation as needed as well as other investigations such as HRCT scans.
Echocardiography is only a screening tool and there will be patients who have raised pulmonary artery pressures PAP) on echocardiogram and who will then have no evidence of pulmonary hypertension (PH) on RHC. The RHC is an important part of the diagnostic pathway.

Once a diagnosis of PH has been made the patients will be managed between both the PH team and the haematologists to consider treatment. These may include regular transfusion, hydroxycarbamide and or targeted therapies and anticoagulation.

In practice shortness of breath, chest pains and reduced oxygen saturations will be investigated with a combination of all modalities mentioned and referral paths and treatments guided as above.

Other respiratory conditions that should be considered include

- VTE
- Obstructive sleep apnoea
- High cardiac output states due to low Hb

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

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

American Thoracic Society guidelines on Pulmonary Hypertension in SCD
http://www.thoracic.org/statements/resources/pvd/sickle-cell-disease.pdf

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