Sickle Cell Disease and Thalassaemia: Chronic Complications (Liver Disease)

Profile	
Version:	*V3.0*
Author:	*Dr Elizabeth Rhodes, Consultant Haematologist* *Dr Julia Sikorska, Consultant Haematologist*
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*
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

Please identify the key people you involved in reviewing this policy why, and when:

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 hepatic complications. Thalassaemia patients may develop liver disease as a result of iron overload or infection. This guideline contains advice on how to manage hepatic complications in patients with haemoglobinopathies.

1. Introduction

Hepatobiliary complications are common in those with sickle cell disease (SCD) and can be caused by direct sickling, vaso-occlusion or iron overload. Patients may develop coexistant viral or autoimmune hepatitis. Liver function test abnormalities are common with a range of possible aetiologies and referral to the Hepatology team may be indicated.

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 SICKLE CELL DISEASE

Liver dysfunction in patients with sickle cell disease varies from mild abnormal liver enzymes to frank liver failure. Isolated hyperbillirubinaemia is usually unconjugated and due to the red cell haemolysis. If there are changes in the liver enzymes then conjugated bilirubin levels should be checked.

There are many causes of abnormal liver function tests and these include

- Direct hepatic sickling
- Medication – including antibiotics, chelation therapies, hydroxycarbamide
- Iron overload from repeated transfusions
- Transfusion transmitted diseases such as hepatitis B and C
- Acute but transient response to infections and periods of being unwell

Patients should have abnormal liver function tests repeated and if persistent a liver screen should be performed including liver ultrasounds, autoimmune studies, ferritin levels and virology studies.

Patients with unexplained or worsening liver function tests should be discussed or referred to hepatology team for review and consideration of further investigations including a fibroscan. In progressive liver disease, treatment may include an exchange transfusion programme – this should be a joint decision by the Haematology and Hepatology teams.

In patients where iron overload is considered to be likely a T2* weighted MRI should be performed to quantify iron loading in the liver. Liver biopsies should be avoided where possible. Iron chelation should be instigated if iron overload confirmed.

- Liver function tests should be monitored at least annually
- Simple transfusion should be considered for those with acute hepatic sequestration associated with anaemia
- Symptomatic gallstones should be treated with laparoscopic cholecystectomy due to shorter hospital stay and fewer complications
- **Emergency exchange transfusion should be considered in those presenting with intrahepatic cholestasis – patients suspected to have this diagnosis should be discussed with the Haematology SpR/Consultant immediately** (this is an uncommon but severe form of acute sickle hepatopathy with patients presenting with sever right upper quadrant pain, hepatomegaly, extreme conjugated hyperbilirubinaemia but moderate transaminitis and coagulopathy with some patients progressing to acute hepatic failure).
- If a liver biopsy is to be performed, this should be done via the trans-jugular route to minimise bleeding risk but should only be performed in patients with SCD if it is felt to be the only diagnostic test possible.

6.2 THALASSAEMIA

In thalassaemia abnormal liver tests are usually due to iron overload with or without underlying viral infections. All patients with abnormal liver function tests must have viral hepatitis screens performed (hep A, hep B, hep C and if thought relevant CMV and EBV as well)

All patients on regular transfusions should have virology testing every year and be tested for immunity to hepatisis B if titres do not show sufficient immune response.

They should be assessed for other causes, iron overload treated and levels monitored

Patients with worsening liver function despite chelation, patients with hepatitis or patients with symptomatic liver dysfunction should be referred to the hepatology team. Imaging for iron overload T2* MRI is available at St George's (see guidelines on managing iron overload and transfusion) and fibroscan to assess fibrosis is also available through hepatology.

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

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