Sickle Cell Disease: Chronic Complications (Avascular Necrosis)

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*
Date issued:	*February 2019*
Review date:	*February 2022*
Approval	
Approval person/Committee:	*Divisional Governance Board (MedCard)*
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

Avascular necrosis (AVN) is a common complication in patients with sickle cell disease (SCD). It may be asymptomatic in the early stages but can progress, often causing severe pain and disability. Treatment can be divided into conservative and surgical approaches.

1. Introduction

Avascular necrosis (AVN) affects up to 50% of patients with SCD is the death of bone tissue due to loss of blood supply, most commonly occurring in the femoral and humeral head but has been reported in the knees, feet and back. It can result in chronic pain, impaired mobility and disability. It should be considered in all patients presenting with sudden or progressive joint pain, particularly if affecting the hips or shoulders and should be managed using a multidisciplinary team (MDT) approach involving the haematology team and a specialist orthopaedic surgeon.

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 OSTEOMYELITIS

For guideline on osteomyelitis see Management of Acute Complications in Adult patients with Sickle Cell Disease.

6.2 AVASCULAR NECROSIS (AVN)

Osteonecrosis, Avascular Necrosis or AVN, occurs when vaso-occlusion results in the infarction of the articular surfaces and heads of the long bones. It is one of the most common orthopaedic complications in sickle cell disease though its prevalence is hard to quantify.

The femoral head, the head of the humerus and then the knee and small joints of the hands and feet are the most common sites where AVN is seen. Patients are also likely to have multiple joints affected: >50% of patients with an affected hip have bilateral disease and 74% of those with an affected shoulder will also have an affected femoral head.

At clinic reviews patients should be asked directly about pain and reduced function of joints. Patients with symptoms should be examined looking at pain, restricted movement and the affect on daily life (stairs, exercise etc) and then have appropriate imaging. Plain x-rays are the most appropriate initial investigation but have low sensitivity for early stage disease and so MRI should be considered if symptoms persist as it has a higher sensitivity and specificity for AVN. If the affects are severe they should be considered for referral to the orthopaedic consultant with a specialist interest in SCD.

Conservative management includes adequate treatment of pain and physiotherapy with appropriate mobility aids where needed. Surgical intervention is nearly always a joint replacement and so this decision needs to be taken in discussion with the patient with regards to timing and affect on their life.

The role of bisphosphonate therapy is not clear in AVN in sickle cell disease but can be considered in certain individuals.

Hospital Contact Details

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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