Sickle Cell Disease:
Chronic Complications (Urology)

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:
Mr Nick Watkin (2015) – link Consultant Urologist Mr B Ayres (2015) - Consultant Urologist Dr L Seal (2015) - Consultant Endocrinologist Mr T Yap (2015) - Consultant Urologist

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 associated with a range of chronic complications as a result of end-organ damage. This guideline provides advice on the management of urological complications in patients with SCD.

1. Introduction

Sickle cell disease is associated with a high incidence of urological complications. Priapism has a lifetime incidence of up to 35-90% in male patients with SCD and can cause significant embarrassment and discomfort for patients but is often poorly discussed. Patient education on steps that can be taken to prevent episodes is important (exercise, staying warm and hydrated) and also when to seek emergency treatment is important.

Haematuria is also common in sickle cell disease and has many causes including papillary necrosis, renal stones and infection and should be investigated regardless of age to exclude malignancy.

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 PRIAPISM

For the management of acute priapism episodes see guidelines on the acute management of patients with SCD.

Stuttering priapism (recurrent episodes of an unwanted, painful erection that lasts for less than 2 hours) is relatively common in male patients with sickle cell disease and can start in adolescence. It is often thought to be due to sickle 'sludging' in the corposa cavernosa but it may well be multifactorial.

Patients may not disclose problems with priapism and so should be asked directly in their annual review.

Patients can often manage these episodes at home with encouragement of taking on some minor exercise, trying to pass urine, taking some pain relief if there are sickle cell pains or taking a warm shower. They must be advised to attend the emergency department if an episode lasts for longer than 2 hours.

Referral to Urology for further investigations (such as penile dopplers) and management advice (Alpha-agonists and anti-androgens usually as first line) should be considered if:

- Recurrent disruptive episodes
- Medication is required ongoing, and for discussion of long term side effects of these medications such erectile dysfunction or infertility
- There are concerns about erectile dysfunction or sub-fertility

There is no evidence for the use of regular transfusions for the management of persistant priapism though anecdotally some patients have found this a benefit and it may be considered on an individual basis.

6.2 HAEMATURIA

Haematuria is common in sickle cell disease and has many causes including infection, stones and papillary necrosis. It may also be secondary to sickle cell syndrome, malignancy and is a potential marker for progressive renal failure. Urine dipstick is sufficient (ie there is no need to confirm the presence of red cells with microscopy.)

It may be transient and there should be two documented samples of 1+ blood on urinalysis for further investigation.

- Exclude infection (sample sent for microscopy, culture and sensitivities)
- Ensure sample not taken during menstruation or heavy exercise
- Screen for diabetes and calculate eGFR and measure BP

- Microscopy for casts
- Assess for proteinuria (with a urinary PCR)
- Ultrasound with a full bladder to gain good views of bladder wall and kidneys. (+/- CT if no cause found on USS or if on-going loin pain and / or visible haematuria.)

Consider referral to urologists if:

- visible haematuria
- persistent non visible haematuria
- abnormalities on imaging
- loin pain or infection in males
- recurrent infection females (or infection if post-menopausal)

Cystoscopy is recommended if over 40 years old and / or smoker and / or visible haematuria

If patient has reduced GFR and proteinuria and or hypertension consider referral to nephrology (see Renal Guidelines) particularly considering the possibility of IgA nephropathy.

If concerns regarding papillary necrosis and obstruction then discuss with urology and haematology urgently

Possible Causes of Haematuria

Renal Papillary Necrosis: this process occurs due to medullary infarction and is seen in patients with both sickle cell disease and sickle cell trait. The haematuria seen in papillary necrosis can be of significant volumes and with clots risking ureteric obstruction with consequential renal failure. This can, on occasion, be life threatening. Treatment encompasses fluid replacement and drive to encourage high urinary flow, supportive blood transfusion as necessary and in some cases surgical intervention is required.
Medullary Carcinoma: this is a rare, aggressive renal tumour seen almost entirely in patients with sickle cell trait and occasionally in patients with sickle cell disease. Haematuria is the most common presentation of this disease.
Renal TB: can also present with haematuria and should be considered in the list of differential diagnoses whilst investigating these patients.
Non-Sickle Cell Causes of haematuria

6.3 MALE SEXUAL DYSFUNCTION AND INFERTILITY

Patients can suffer from male sexual dysfunction and infertility. The underlying problems can be multifactorial and can include one or more of the following:

- Repeated untreated episodes of priapism
- Medication
- Psycho-sexual dysfunction
- Transfusional iron overload can affect the testes, hypothalamus and the pituitary gland causing dysfunction and hypogonadism

Patients should be asked about any concerns at their annual review and patients with difficulties should have a full pituitary axis screen including a hormonal profile of

testosterone, SHBG, Gonadotrphin and prolactin. They can then be referred to the complex medical andrology service. Patients can either be discussed in an MDT setting or seen by Dr Seal in endocrinology with input from urology as needed.

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

Guidelines on the acute management of patients with SCD.

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