

## Sickle Cell Disease Chronic Complications (Renal Disease)

Profile	
<b>Version:</b>	V3.0
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<b>Executive/Divisional sponsor:</b>	<i>Dr Lisa Pickering, Divisional Chair DGB February 2019</i>
<b>Applies to:</b>	<i>All staff involved in the care of patients with Sickle Cell Disease</i>
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Approval	
<b>Approval person/Committee:</b>	<i>Divisional Governance Board (MedCard)</i>
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## 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 Joyce Popoola (2015) – link consultant for renal and nephrology

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 renal complications. This guideline contains advice on how to manage renal complications in patients with SCD.

## 1. Introduction

Patients with SCD are susceptible to both anatomical and functional renal damage through several mechanisms. Chronic kidney disease (CKD) is relatively common and progressive. Structural and functional defects of the nephron cause a spectrum of conditions including painless haematuria, proteinuria and eventual loss of function resulting in end stage renal failure (ESRF) in some cases.

## 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 Hyposthenuria**

**6.2 Proteinuria**

**6.3 Chronic kidney disease**

**6.4 End stage renal failure**

**6.5 Dialysis & renal transplantation**

**6.6 BP optimisation**

## 6.1 HYPOSTHENURIA

Due to an increased glomerular filtration rate (GFR) in these patients from early childhood to young adulthood there is an increase in the proximal tubular function and an inability to concentrate the urine. This manifests clinically as a susceptibility to dehydration which can trigger crises.

Patients therefore must be encouraged to drink adequate volumes.

Patients may also have an incomplete form of distal renal tubular acidosis leading to acidosis and hyperkalemia this can usually be corrected with dietary potassium restriction and oral bicarbonate however this should be discussed with a nephrologist if persistent.

## 6.2 PROTEINURIA

Patients must have a urine dipstick assessment of their urine for urinary protein as an initial assessment at least once a year (usually at their annual review.). Urinalysis may also pick up haematuria and indicators of urinary tract infection. Positive measurements during menstruation or acute illness need to be repeated.

Protein result	Investigations	Action
Trace / none		Repeat annually
1+	<ul style="list-style-type: none"> <li>• Monitor and optimise BP - Aim BP <math>\leq</math>130/80</li> <li>• Screen for diabetes</li> <li>• Stop Nephrotoxic drugs</li> <li>• Exclude UTI</li> </ul>	If BP and GFR normal then repeat in 6 months IF BP high then refer to GP to treat (recommending ACE-inhibitor) and monitor proteinuria 3-6 months If reduced GFR refer to renal team
2+ or more	<ul style="list-style-type: none"> <li>• Send urine for PCR</li> <li>• Monitor and optimise BP - Aim BP 130/80</li> <li>• Screen for diabetes</li> <li>• Stop Nephrotoxic drugs</li> <li>• Exclude UTI</li> <li>• Request renal screen – autoimmune bloods and renal USS</li> </ul>	If urine PCR < 100 and BP within normal limits then repeat in 6 months If PCR > 100 or hypertensive refer to renal team for review and instigation of ACE-inhibitor as needed

### Discuss and/or Refer to Renal Medicine:

PCR > 100mg /mmol or ACR > 70mg/mmol

Reduced GFR with any level of proteinuria

Increasing or refractory proteinuria

If there is concern that this is not a sickle related renal problem (e.g abnormal ultrasound or autoimmune screen) a renal biopsy by a nephrologist may be necessary to make a definitive diagnosis.

Patients with diabetes may need earlier review.

**If urine PCR > 50 mg/mmol, patients should be considered for hydroxycarbamide treatment.**

### 6.3 CHRONIC KIDNEY DISEASE (CKD)

Renal function (as in the eGFR) should be monitored at least annually and a reduction in the eGFR warrants investigation and referral to the nephrologists. All patients with stage CKD 4 (eGFR <30) should be followed up by a nephrologist. Minimisation of nephrotoxic drugs such as NSAIDs and gentamicin and ensuring dosing of all medications is done according to their eGFR. Administration of contrast for radiological procedures and the like should be avoided where possible and appropriate precautions taken when essential. Patients may require treatment with ACE-inhibitors and may be considered for hydroxycarbamide therapy or transfusion therapy on an individual basis.

It is important to ensure patients are adequately hydrated at all times.

### 6.4 END STAGE RENAL FAILURE (ESRF)

This is managed by the nephrologists and should include early discussion of renal replacement therapy options including transplant and dialysis.

Patients with renal failure associated anaemia can be considered for erythropoietin therapy as in the non-sickle population. Patients with SCD on dialysis will require higher than the average dose when erythropoietin is used to correct anaemia. This should be done in discussion with the haematologists and nephrologist.

### 6.5 DIALYSIS & RENAL TRANSPLANTATION

Haemodialysis (HD) and peritoneal dialysis (PD) are both options for management of end-stage renal failure in the Sickle cell patient. Some precautions such as warming the PD fluid prior to insertion and ensuring the HD machine is kept at a reasonable temperature while the blood is flowing through the circuits.

When possible, renal transplantation provides sickle cell patients with the best quality of life.

Each patient being considered or on the list for a renal transplant should be discussed between haematology and nephrology to cover the following:

- Transfusion programme prior (increased blood transfusions can increase risk of antibody formation therefore needs to be with antigen matched blood)
- Surgical and anaesthetic plan for the procedure
- Post and peri-transplant management to try to protect new transplanted kidney. Most patients will be considered for a transfusion programme (this maybe exchange blood transfusion to reduce hyperviscosity) following the procedure.
- Special focus to fluid balance, haematocrit, peripheral perfusion, temperature and oxygenation peri-procedure and any venous access issues
- Likely need for more frequent than average follow-up in outpatients in relation to fluid balance, immunosuppressant levels, renal function, anaemia.
- Patients need to be counselled re: possibility of recurrence of sickle cell damage to transplanted organ



## 6.6 BP OPTIMISATION

Many patients with sickle cell will run a lower than average BP so higher pressures should be treated aggressively. In patients with a normal GFR and no proteinuria then a target of  $\leq 140/90$  is adequate whilst in patients with proteinuria (PCR>50 or ACR>30) the target should be to reduce BP to less than 130/80

### 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**

<b>Element/ Activity being monitored</b>	<b>Lead/role</b>	<b>Methodology to be used for monitoring</b>	<b>Frequency of monitoring and Reporting arrangements</b>	<b>Acting on recommendations and Leads</b>	<b>Change in practice and lessons to be shared</b>
<i>WMQRS peer review quality standards</i>	<i>Consultant haematologist</i>	<i>As required (every 2-3 year)</i>	<i>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.</i>	<i>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</i>	<i>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.</i>

## 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>