St Helens and Knowsley Teaching Hospitals NHS Trust

Acute and Emergency Management of Sickle Cell Disease in Adults

Version No: 1

Document Summary:

Document status	Approved	
Document type	Guideline	**Trust wide**
Document number	Document Control will provide document number if a new document	
Approving body	Drugs and Therapeutics Council	
Date approved	11/05/2023	
Date implemented	15/06/2023	
Review date	**11/05/2026**	
Accountable Director	Medical Director	
Policy Author	Dr Toby Nicholson, Consultant Haematologist	
Target audience	Clinical staff in acute and emergency settings	

The intranet version of this document is the only version that is maintained. Any printed copies should therefore be viewed as "uncontrolled", as they may not contain the latest updates and amendments.

Document Control

Section 1 – Document Information	
Title	Acute and Emergency Management of Sickle Cell Disease in Adults
Directorate	Medical
Brief Description of amendments	
N	
Does the document follow the Trust agreed format?	Yes
Are all mandatory headings complete?	Yes
Does the document outline clearly the monitoring compliance and performance management?	Yes
Equality Analysis completed?	Yes

Section 2 – Consultation Information			
Consultation Completed	☐ Trust wide ☑ Local ☐ Specific staff group		
Consultation start date	23/03/2023	**Consultation end date**	06/04/2023

Section 3 – Version Control		
Version	**Date Approved**	**Brief Summary of Changes**
1	11/05/2023	New document

Section 4 – Approval *– To be completed by Document Control*			
Document Approved	☐ Approved ☐ Approved with minor amendments		
Assurance provided by Author & Chair	☐ Minutes of Meeting ☐ Email with Chairs approval		
Date approved	Click here to enter a date.	**Review date**	Click here to enter a date.

Section 5 – Withdrawal *– To be completed by Document Control*	
Reason for withdrawal	☐ No longer required ☐ Superseded
Assurance provided by Author & Chair	☐ Minutes of Meeting ☐ Email with Chairs approval
Date Withdrawn:	Click here to enter a date.

Contents

1. Scope

To support the clinical management of adult patients with sickle cell disease presenting with complications of their disease to acute and emergency services in the Trust.

2. Introduction

The All Party Parliamentary Group (APPG) Report on Sickle Cell Disease, 'No One's Listening', was published in November 2021 and highlights the avoidable deaths and failure in care for sickle patients in secondary care facilities. Part of the action plan in response to that was to ask that all trusts have a plan in place to ensure compliance with the NICE clinical guideline around the delivery of pain relief within 30 minutes for sickle cell patients; the regional Specialist Haemoglobinopathy Team at Liverpool University Hospitals Foundation Trust (LUHFT) has put together guidance that can be used in hospitals such as ours in the rare event of a sickle cell patient presents in crisis. This guideline is based on that.

3. Statement of Intent

This document aims to standardise the management of sickle cell patients presenting to emergency services in the Trust and to highlight the need for urgent treatment to avoid unnecessary morbidity and mortality.

4. Definitions

Definition	Meaning
Sickle Cell Disease	An inherited condition wherein red clood cells are less flexible than usual leading to complications such as infection, growth retardation, intermittant painful crises, stroke and early mortality
Sickle cell crisis	A situation where inflexible red blood cells obstruct small blood vessels causing tissue hypoxia and consequent severe pain and a risk of organ damage.
Acute chest syndrome	A situation where the blood vessels of the lung are obstructed leading to difficulty in oxygenating the blood and a high risk of death.

5. Duties, Accountabilities and Responsibilities

5.1 Chief Executive

The Chief Executive has overall responsibility for the strategic and operational management of the Trust.

5.2 Medical Director

The Medical Director is the accountable director for this guideline.

5.3 Consultants in acute and emergency care settings

Consultants working in acute and emergency care settings in the Trust should be aware of this guideline and are responsible for ensuring that sickle cell complications are managed in accordance with it. They are responsible for ensuring liaison with the regional haemoglobinopathy service is prioritised.

6. Management of Acute Complications of Sickle Cell Disease

There are some key principles which need to be followed in managing such patients:

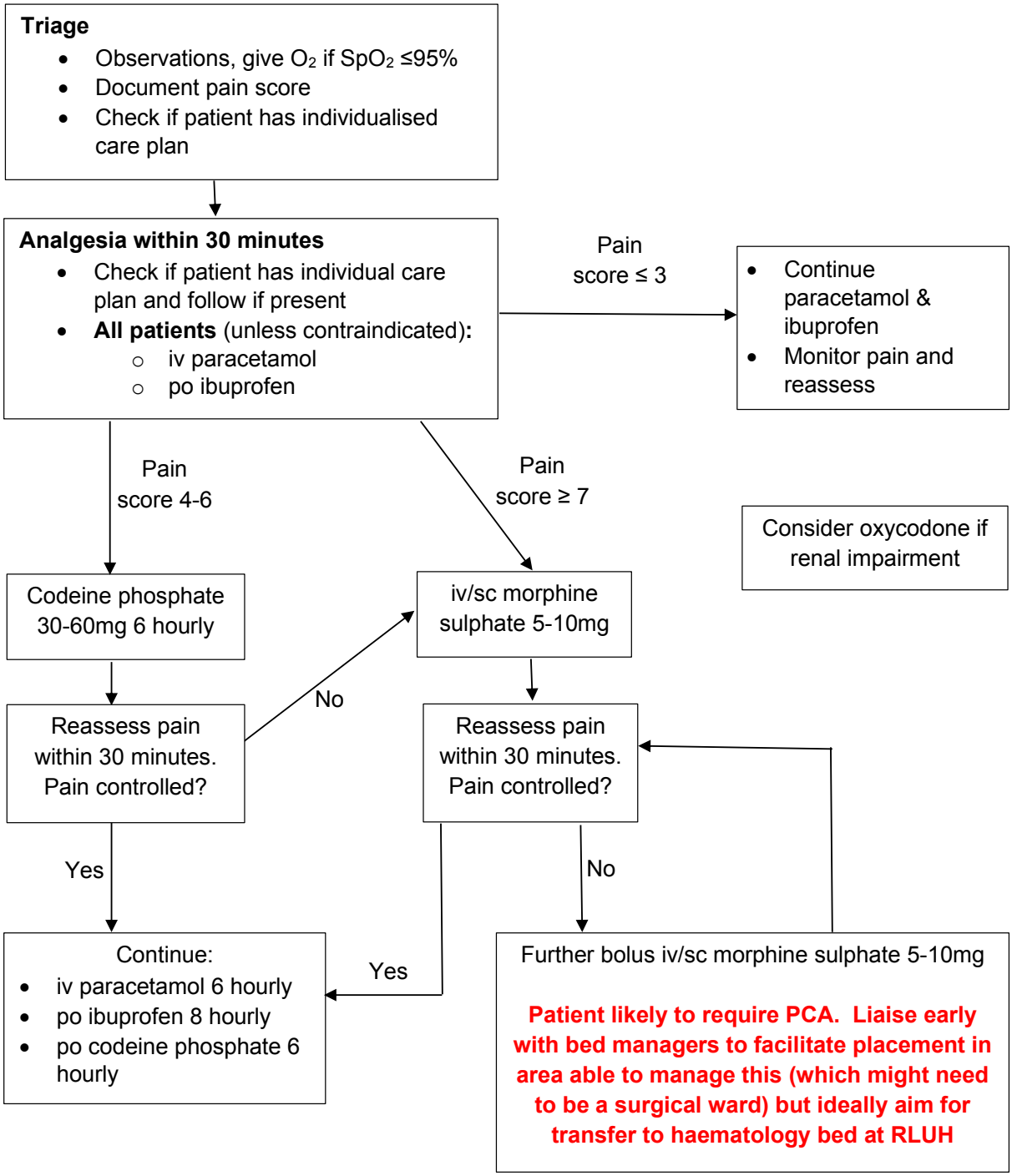
- Inform the Haemoglobinopathy Team at LUHFT of **all** presentations of patients with sickle cell disease via the on-call haematology registrar through LUHFT switchboard (0151 706 2000). The local haematologists will also need to be aware of the admission.
- Bear in mind that seemingly unrelated presentations may be related to sickle cell disease or may have important implications.
- Do not transfuse outside emergency situations without discussion with haematology.
- Patients with sickle cell disease are experts in their condition and must be listened to.
- Individualised care plans may be available; ask the patient if they have one (it may be paper or electronic) and check patient records. The Haemoglobinopathy Team or on-call haematologist at LUHFT may be able to provide a copy.
- Surgery is a period of significant risk for patients with sickle cell disease and should never happen without discussion with the team at LUHFT first.

If admission is required, consider early transfer to the Royal Liverpool Hospital site of LUHFT to facilitate input from the specialist team and admission to the specialist unit.

6.1 Acute Painful Crisis

Acue painful sickle cell crisis presents with acute severe pain, usually in the limbs or back, due to blockage of small blood vessels causing tissue ischaemia. Most patients recognise the onset of a crisis early and prompt treatment is vital.

- Acute Painful Crisis is a medical emergency
- **Inform LUFHT Haematology Team at presentation**
- Patients must be triaged and assessed as a priority
- Patients must receive pain relief within 30 minutes with the aim to be pain controlled within 60 minutes
- If available, refer to the patients individualized care plan
- Consider analgesia already taken prior to presentation and chronic analgesia

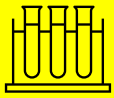

Triage
Observations, give O_2 if SpO_2 ≤95%
Document pain score
Check if patient has individualised care plan
Analgesia within 30 minutes
Check if patient has individual care plan and follow if present
All patients (unless contraindicated):
iv paracetamol
po ibuprofen
Pain score ≤ 3
Continue paracetamol & ibuprofen
Monitor pain and reassess
Pain score 4-6
Pain score ≥ 7
Consider oxycodone if renal impairment
Codeine phosphate 30-60mg 6 hourly
iv/sc morphine sulphate 5-10mg
Reassess pain within 30 minutes. Pain controlled?
No
Reassess pain within 30 minutes. Pain controlled?
Yes
No
Continue:
iv paracetamol 6 hourly
po ibuprofen 8 hourly
po codeine phosphate 6 hourly
Yes
Further bolus iv/sc morphine sulphate 5-10mg
Patient likely to require PCA. Liaise early with bed managers to facilitate placement in area able to manage this (which might need to be a surgical ward) but ideally aim for transfer to haematology bed at RLUH

6.1.1 Further Management of Painful Crisis

All Patients

- IV Fluids
- Oxygen if SpO_2 ≤95%
- Antibiotics if suspected infection – follow sepsis pathway
- Laxatives if constipated or prescribed strong opiates
- PRN Ondansetron for opiate induced itch/nausea
- VTE risk assessment and thromboprophylaxis
- Incentive Spirometry (refer to physiotherapy, in interim encourage 8 deep breaths per hour when awake)

Investigations



- FBC, Reticulocyte Count, Haemoglobin Profile
- UE, LFT, LDH, Lactate
- Group and Save
- Blood/Urine/Sputum cultures if suspicion of infection
- Chest X-ray if chest symptoms or hypoxia
- ABG if SpO_2 ≤94% or significantly less than baseline

Red Flags for Acute Chest Syndrome

- Respiratory Signs/Symptoms
- Chest Pain
- Fever
- SpO_2 ≤95% or increasing oxygen requirements

These should prompt urgent clinical reassessment and discussion with Haematology team

Pregnant Patients

- Inform Obstetric Team of presentation
- Follow normal pain pathway or individualized care plan but **OMIT NSAIDs** unless discussed with Haemoglobinopathy Team/Obstetrics

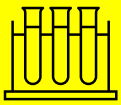
6.2 Acute Chest Syndrome

Acute chest syndrome (ACS) is defined as a new radiodensity on chest imaging accompanied by fever and/or respiratory symptoms. It is an acute complication of sickle cell disease that is potentially fatal and requires immediate intervention. It is closely related to an acute painful crisis but failure to recognise it can lead to deterioration.

Signs/Symptoms

- Hypoxia
- Chest pain
- Signs of consolidation
- Fever
- Tachycardia
- Tachypnoea
- Shortness of breath
- CXR changes (may lag behind clinical features)

Essential Investigations



- FBC, reticulocyte count
- UE/LFT/CRP
- Group & save
- CXR
- Blood cultures
- Sputum cultures
- Atypical pneumonia serology
- Viral respiratory swabs

Management

- Inform LUHFT Haematology urgently
- iv fluids (caution re. overload)
- iv antibiotics if features of infection – follow sepsis pathway; antibiotics as per community acquired pneumonia initially
- Analgesia (see Acute Painful Crisis pathway)
- Chest physio & incentive spirometry
- Critical care review early if concerns re. deterioration
- Ongoing management will be guided by LUHFT Haematology

6.3 Stroke

Stroke

- Discuss at Presentation with LUFHT Haematology Team – management differs to other patients
- Urgent Imaging (CT/CTA)
- Clinical Stabilisation
- Transfusion (exchange) likely to be required – d/w Haematology
- Thrombolysis may be an option but MUST BE DISCUSSED WITH HAEMOGLOBINOPATHY CONSULTANT PRIOR TO PRECEEDING

6.4 Priapism

Priapism

- Fulminant priapism is defined as prolonged attack lasting >3hours
- **Fulminant priapism is a medical emergency requiring treatment within 1 hour**
- **Management**
  - Confirm priapism
  - Encourage to pass urine (may require catheter)
  - po/iv hydration
  - Keep warm (do not apply ice packs)
  - Analgesia
  - Urgent discussion with urology regarding management
  - Exchange transfusion is usually not indicated

6.5 Emergency Surgery

Emergency Surgery

- **Do not deny urgent surgery due to sickle cell disease BUT must be discussed with haematology at the earliest opportunity**
- **Management**
  - Transfusion – discuss with LUHFT Haematology as approach depends on patient/procedure
  - Good oxygenation and SpO_2 monitoring throughout
  - Enhanced care (critical care bed post-op)
  - Hydration
  - Chest physio and incentive spirometry post-op
  - Analgesia
  - VTE prophylaxis
  - Low threshold for antibiotics

7. Training

Training required to fulfil this policy will be provided in accordance with the Trust's Induction Mandatory and Risk Management Training Policy - Training Needs Analysis.

8. Monitoring Compliance

8.1 Key Performance Indicators (KPIs) of the Policy

No	Key Performance Indicators (KPIs) Expected Outcomes
1	Monitoring of adverse events related to sickle cell disease using DATIX system

9. References

No	Reference
1	All Party Parliamentary Group Report on Sickle Cell Disease – "No One's Listening"; November 2021. extension://efaidnbmnnnibpcajpcglclefindmkaj/https://www.sicklecellsociety.org/wp-content/uploads/2021/11/No-Ones-Listening-Final.pdf

10. Equality Analysis Form

The screening assessment must be carried out on all policies, procedures, organisational changes, service changes, cost improvement programmes and transformation projects at the earliest stage in the planning process to ascertain whether a full equality analysis is required. This assessment must be attached to all procedural documents prior to their submission to the appropriate approving body. A separate copy of the assessment must be forwarded to the Patient Inclusion and Experience Lead for monitoring purposes. Cheryl.farmer@sthk.nhs.uk. If this screening assessment indicates that discrimination could potentially be introduced, then seek advice from the Patient Inclusion and Experience Lead. A full equality analysis must be considered on any cost improvement schemes, organisational changes or service changes which could have an impact on patients or staff.

Equality Analysis			
Title of Document/proposal/service/cost improvement plan etc:	Na		
Date of Assessment	02/03/2023	**Name of Person completing assessment /job title:**	Dr Toby Nicholson
Lead Executive Director	Medical Director		Haematology Consultant

	Does the proposal, service or document affect one group more or less favourably than other group(s) on the basis of their:	**Yes / No**	**Justification/evidence and data source**
1	Age	No	Click here to enter text.
2	Disability (including learning disability, physical, sensory or mental impairment)	No	Click here to enter text.
3	Gender reassignment	No	Click here to enter text.
4	Marriage or civil partnership	No	Click here to enter text.
5	Pregnancy or maternity	No	
6	Race	No	Click here to enter text.
7	Religion or belief	No	Click here to enter text.
8	Sex	No	Click here to enter text.
9	Sexual Orientation	No	Click here to enter text.
	Human Rights – are there any issues which might affect a person's human rights?	**Yes / No**	**Justification/evidence and data source**
1	Right to life	No	Click here to enter text.
2	Right to freedom from degrading or humiliating treatment	No	Click here to enter text.
3	Right to privacy or family life	No	Click here to enter text.
4	Any other of the human rights?	No	Click here to enter text.

Lead of Service Review & Approval	
Service Manager completing review & approval	Click here to enter text.
Job Title:	Click here to enter text.