1. Introduction and Scope

1.1 This Guideline has been written to support Physiotherapists within the University Hospitals of Leicester (UHL) treating patients admitted to UHL with a diagnosis of Sickle Cell Disease (SCD)

1.2 This guideline applies to all Physiotherapy staff that care for patients with sickle cell disease. These staff have a responsibility to follow this guideline.

1.3 It particularly applies to those Physiotherapists working within the Haematology and any Surgical clinical areas and within the Respiratory Physiotherapy weekday or weekend service.

2. Recommendations, Standards and Procedural Statements

The aim of rehabilitation for patients with sickle cell disease is to improve quality of life, facilitate functional independence and enable safe, effective and appropriate discharge.

All patients admitted with vaso-occlusive crisis should be offered a respiratory assessment by a physiotherapist during their admission, especially those who present with chest, back pain, lower respiratory tract infection (LRTI) or hypoxia.

2.1 Key points to consider:

- Referral should be made to Physiotherapy within 24 hours of admission and if required to Occupational Therapy within 24 hours of admission.
- Pre-operative and post-operative physiotherapy assessments should be offered routinely to sickle cell patients undergoing surgery with a high risk of pulmonary complications. Those with pulmonary risk factors for more general surgery should also be assessed. Prophylactic treatment and advice should be offered as indicated by assessment.
- Rehabilitation should be patient-centred with short term, realistic goals which focus on functional outcomes in order to achieve the best quality of life for each individual patient.

2.2 Initial Therapy treatment on admission:

1. In order to improve compliance, it should be ensured all patients should have adequate analgesia prior to assessment and treatment.
2. Incentive spirometry should be offered to all patients, regardless of clinical findings, unless there is a contraindication to this.
3. Patients use the spirometer every two hours, during waking hours, when in crisis, and patients should continue until the crisis subsides.
4. Incentive spirometry may be supplemented with chest clearance techniques as indicated
5. Physiotherapists should continue to review the patient regularly until it is ensured they are effective with the incentive spirometer and independent in its

use. Prophylactic use in every-day life should be encouraged and the patient provided with an advice leaflet where possible.

6. Alternative treatment to increase lung volumes should be used with those patients for whom incentive spirometry is contraindicated or who are unable to establish an effective technique
7. Gentle mobilisation should begin once pain is under control. Careful monitoring of observations pre and post exercise as well as patient response should be recorded due to an increased likelihood of abnormal exercise response in SCD patients. Ambulatory Oxygen should be used for those patients who have a SaO2 of 95% or less on air.
8. Physiotherapy manual chest techniques are usually contraindicated due to high potential for fracture, high likelihood of increased pain and clotting disorders.
9. Care should be taken in the use of suction due to a high likelihood of clotting disorders and the relative risks clinically reasoned. The advice of the medical team should be sought if in doubt
10. Oxygen therapy should be given to those patients whose SaO2 falls below 95%, this should be maintained during incentive spirometry with nasal cannula, unless otherwise indicated.
11. Systemic hydration should be encouraged unless otherwise indicated in the medical management plan
12. Bronchodilators are not given routinely but should be considered following assessment if indicated.
13. There might be a potential risk from atypical bacteria posed to the treating therapist. If this is the case, the patient will require both standard or enhanced infection prevention precautions, and aseptic techniques for all equipment
14. Intermittent Positive Pressure Breathing (IPPB) may be used as long as pain control has been addressed prior to treatment, normal precautions and contraindications apply.

Physiotherapists working within Haematology, Orthopaedics, General Surgery and Pelvic Health teams should all be aware of the above guidelines because of the increased risk of Acute Coronary Syndromes with general anaesthesia

2.3 On Call Physiotherapy

Standard criteria for on call chest physiotherapy apply and it is inappropriate to call out the physiotherapist in the absence of any of the problems outlined in the on call guidance e.g. solely for the provision of an incentive spirometer

If the patient is admitted to a haematology ward out of hours (16:30 – 08:30) the nursing staff there can provide the patient with an incentive spirometer and instruct the patient on it use. The ward physiotherapist will then review at the earliest opportunity.

If the patient is not on a haematology ward the patient should be referred to the local/ ward Physiotherapy team at the earliest available opportunity within working hours, in the absence of any other on call needs.

2.4 Options for Discharge to consider with patients and carers:

- Home with no additional support or equipment required
- Home with equipment
- Home with package of care with or wthout equipment
- Consider in-patient rehabilitation in light of patient's diagnosis and condition
- Consider available community therapy services including intensive community therapy on discharge
- Nursing Home/ Residential Home placement

3. Education and Training

3.1 All Physiotherapists working with this patient group will undertake education and training on the treatment of patients with sickle cell disease

3.2 All Physiotherapists will follow the assessment and competence process outlined below for proof of training and competence

Procedure / Process for the assessment of Physiotherapist competence	
No.	**Action**
1	Physiotherapist must read UHL Physiotherapy guideline for treatment of patients with Sickle Cell Disease, and reflect and demonstrate learning
2	Physiotherapist must read: UHL Guideline for Adults with sickle cell disease, acute presentation Trust Ref: C15/2012
3	**Has received 1:1 or group training on, and achieved competence in:** • Identification of the signs and symptoms of sickle cell disease, their implications and red flags. • The contra-indications and precautions needed when treating patients with sickle cell disease • Use of an incentive spirometer for patients with sickle cell disease • Use and provision of Oxygen Therapy • The ability to recognise when basic respiratory treatment is not effective and the patient might require the use of Intermittent Positive Pressure Breathing (IPPB)
4	Can identify signs and symptoms of sickle cell disease and their implications, including any "Red Flags"
5	Can demonstrate understanding of contraindications and precautions with sickle cell disease

3.3 Physiotherapists to be assessed for competence following initial training, the outcome recorded in Appendix 1 at the end of this document.

4. Monitoring and Audit Criteria

What will be measured to monitor compliance	How will compliance be measured	Lead	Frequency	Reporting arrangements
All appropriate staff receive initial sickle cell disease training	During clinical placement within the area.	Line manager	By the end of the clinical placement	Therapy mandatory training database
All appropriate staff review their sickle cell disease compliance/ competence	At appraisal	Line manager	annually	Therapy mandatory training database
Any adverse incident notification submitted, if Physiotherapist is unable to treat patients due to lack of competence, or treatment given to the patient is incorrect	DATIX system	Therapy Clinical Team Leader	As incidents arise	DATIX reports within Therapy service

5. Supporting Documents and Key References

UHL Guideline for Adults with sickle cell disease: acute presentation **Trust Ref: C15/2012**

6. Key Words

"Red Flags" - physical signs or symptoms that alert the treating Therapist to potentially serious pathology

IPPB Intermittent Positive Pressure Breathing: Intermittent Positive Pressure Breathing is a technique used to increase the inspired tidal volume during spontaneous breathing. As the patient breathes in, the IPPB is triggered to deliver inspiratory pressure support to enable a deeper breath. Through the utilisation of collateral channels, recruitment of atelectatic lung is improved and secretions are mobilised.
See separate Equipment Competency for IPPB, therapy z-drive
Z:\Equipment Competencies\Equipment Competencies\Adult and Paediatric Respiratory Competencies\IPPB\IPPB competency 2020.doc

Incentive spirometer The incentive spirometer is used as an exerciser for inspiration exercises. Sustained Maximal Inspiration (SMI) is a conscious inhalation breathing exercise which corresponds to sighing and yawning at a physiological level.
This device:

- encourages a larger breath on inspiration, to help improve lung volumes and force air, via collateral channels, behind secretions. This helps to mobilise secretions up towards central airways to enable easier expectoration by coughing.
- prevents atelectasis and promotes speedier recovery for surgical patients (thoracotomies or laparotomies).
- provides visual and positive feedback when patients inhale at a pre-determined flow-rate or volume and sustain the inflation for a minimum of 3 seconds.

DEVELOPMENT AND APPROVAL RECORD FOR THIS DOCUMENT			
Author / Lead Officer:	Jane Bowler	**Job Title:** Therapy Clinical Team Leader	
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REVIEW RECORD			
Date	**Issue Number**	**Reviewed By**	**Description Of Changes (If Any)**
14/03/2022	V2	Ryan Newton (Macmillan Clinical Specialist Physiotherapist) & Rebecca Loomes (Oncology & Neurology Therapy Team Leader)	Guideline reviewed. No changes to be made.

DISTRIBUTION RECORD:			
Date	**Name**	**Dept**	**Received**

Appendix 1

Physiotherapists to be assessed for competence following initial training.

Record of competence of Physiotherapist.

Action	Demonstrated / discussed	Assessed by	Date of competence
Read UHL Guideline for treatment of adults with Sickle Cell Disease, and the Guideline for the Physiotherapy treatment of patients with Sickle Cell Disease. They should reflect upon and demonstrate learning			
Can identify signs and symptoms of Sickle Cell Disease and their implications, including “Red Flags”			
Can demonstrate understanding of contraindications and precautions with sickle cell disease			
Can demonstrate competence in use of an incentive spirometer			
Can demonstrate competence in provision of and use of Oxygen therapy			
Can demonstrate competence in identification for the need for IBBP			

Physiotherapists should keep this record within their Continuous Professional Development (CPD) file

Self-assessment by reading this guideline and demonstration of the identified treatment techniques every 12 months subsequently. This will be checked by their line manager at appraisal.