NHS SCOTLAND

SPAH
SCOTTISH PAEDIATRIC AND ADULT HAEMOGLOBINOPATHIES NETWORK

Scottish Paediatric & Adult Haemoglobinopathy Network

Physiotherapy guideline for the acute management of adult and paediatric patients with Sickle Cell Disease

1. Purpose of this document
To provide guidelines for physiotherapy staff involved with the acute management of paediatric and adult patients with Sickle Cell Disease.

2. Who should use this document
Physiotherapy staff working with patients with Sickle Cell Disease; including weekend and Emergency Duty staff.

3. Further reference
Reference list included in the guideline.

4. Published by
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For background information see:
"SPAH - Introduction to Sickle Cell Disease for Physiotherapists" (NSD610-017.29)

Acute Physiotherapy Management

Referral Criteria

Patients with SCD can be admitted due to a variety of different reasons as outlined. Patients should be referred for chest physiotherapy (CPT) (see appendix 1) immediately on admission if they;

- Have Acute Chest Syndrome.
- Have had previous Acute Chest Syndrome.
- Require I.V opiate analgesia.
- Are admitted for surgery (e.g splenectomy, abdominal surgery)
- Have back, chest, rib or abdominal pain.
- Have chest x-ray changes.
- Have clinical signs of infection.
- Have decreased mobility[17].

If patients meet any of the criteria listed above they should be referred immediately to the physiotherapy team and, if out of hours, the on call physiotherapist should be contacted.

Chest Physiotherapy (CPT)

Early mobilisation of the patient should be encouraged and supported by the physiotherapist.

The physiotherapist is responsible for starting the patient on incentive spirometry (IS) or an age appropriate form of CPT (i.e. bubble PEP, blowing games, deep breathing exercises). It is essential the physiotherapist discuss the importance of CPT with the patient, parents and patients named nurse ensuring they have appendix 1 and 2. Nursing staff will provide hourly supervision and document on a SCD incentive spirometry sheet (see appendix 3). The physiotherapist will review the patient daily and more often if required.

Acute Musculoskeletal Physiotherapy

Patients with SCD admitted with a VOC often present with very severe pain in a joint or bone. Signs and symptoms include; pain, swelling, reduced range of movement (ROM) and fever. Physiotherapists should liaise closely with the MDT prior to their assessment and provide feedback to the MDT. Pain control is extremely important and requires good communication between the physiotherapist and medical staff to ensure physiotherapy can be carried out comfortably.

The aim of inpatient musculoskeletal physiotherapy is to prevent muscle shortening, optimize joint ROM, maintain muscle strength and where possible return to pre-crisis function.

Differential diagnosis: osteomyelitis, septic arthritis, AVN, DVT and pathological fractures including vertebral arch collapse.

Contraindications: Never use cryotherapy (ice) for children with a VOC as this will result in vasoconstriction and may result in further vaso-occlusion. Mobility and weight bearing may be restricted if DVT, AVN, fractures or septic arthritis is suspected.

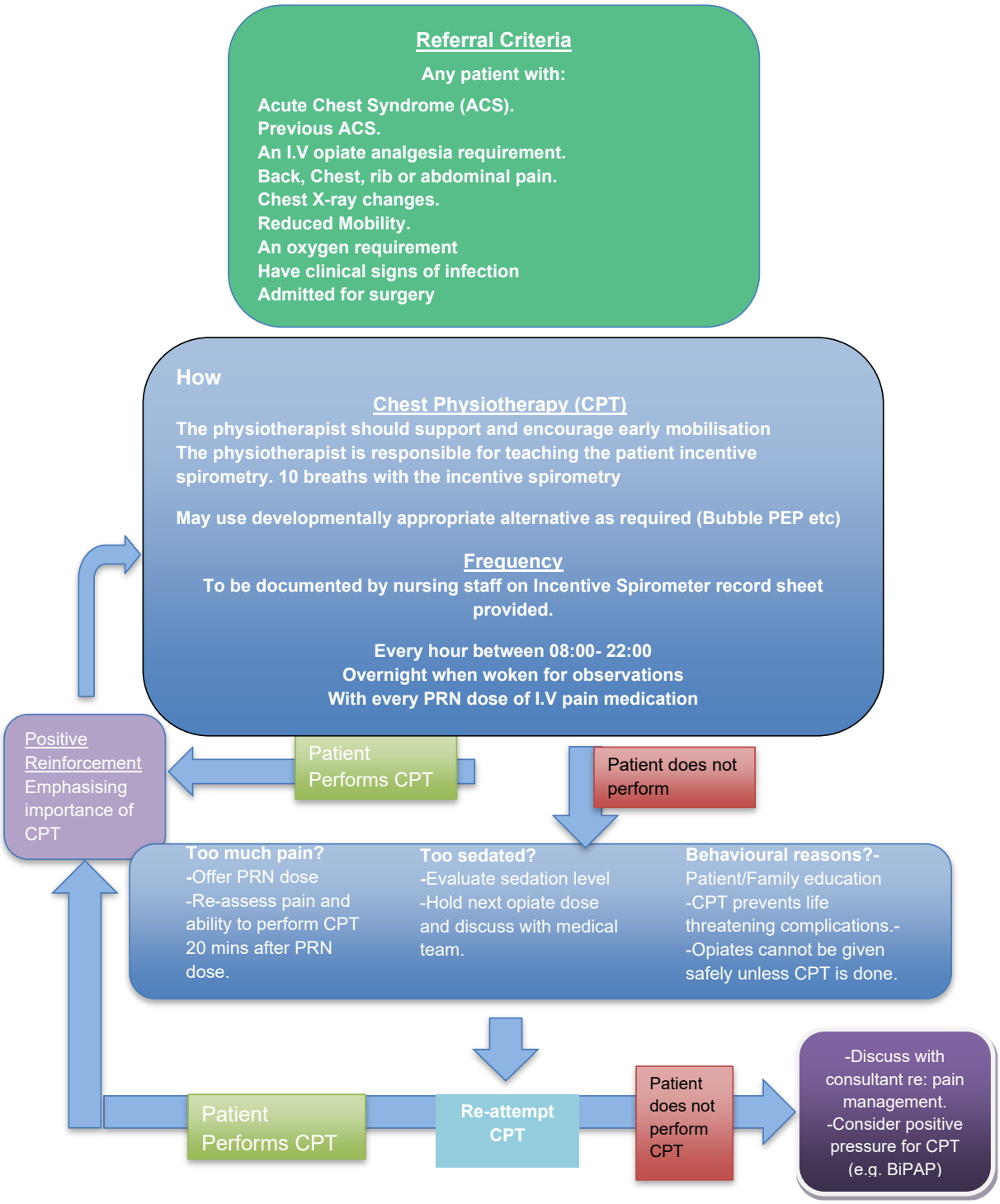
Mobility: Patients should be encouraged to mobilise as soon as possible unless contraindicated. There are usually no restrictions on weight bearing. Mobility aids can be issued but a normal gait pattern should be encouraged.

ROM: Joint ROM should be assessed and an appropriate active range of motion (AROM) exercise programme provided. A stretching programme that ensures pure joint movements should be initiated if it is indicated.

Strength: Muscle strength should be assessed and if the child has reduced mobility due to pain, then an exercise programme should be provided.

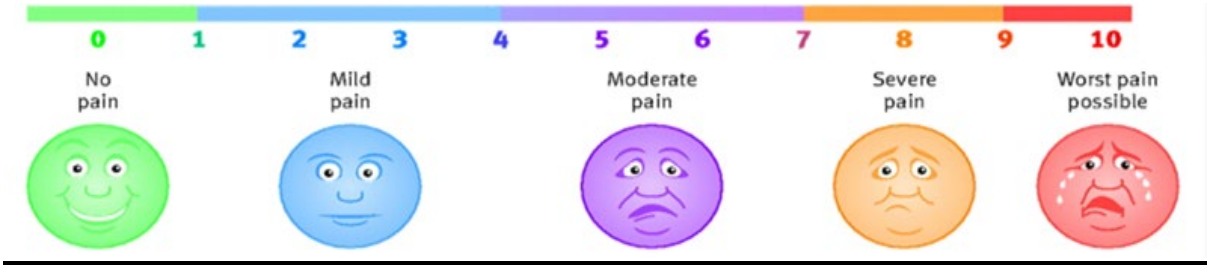
NOTE
This guideline is not intended to be construed or to serve as a standard of care. Standards of care are determined on the basis of all clinical data available for an individual case and are subject to change as scientific knowledge and technology advance and patterns of care evolve. Adherence to guideline recommendations will not ensure a successful outcome in every case, nor should they be construed as including all proper methods of care or excluding other acceptable methods of care aimed at the same results. The ultimate judgement must be made by the appropriate healthcare professional(s) responsible for clinical decisions regarding a particular clinical procedure or treatment plan. This judgement should only be arrived at following discussion of the options with the patient, covering the diagnostic and treatment choices available. It is advised, however, that significant departures from the national guideline or any local guidelines derived from it should be fully documented in the patient's case notes at the time the relevant decision is taken.

Appendix 1. Respiratory Physiotherapy for Acute Pain Admission in Sickle Cell Disease



Appendix 2. Incentive spirometer and pain score information sheet

Pain Score



Incentive spirometer

Instructions:

1. Hold the incentive spirometer (also known as a Triflo) in an upright position, exhale normally and place your lips tightly around the mouthpiece.
2. Inhale slowly and for as long as possible, trying to lift the first and second ball.
3. Keep the balls above the marked line, by breathing in for as long as possible, aiming for 5 seconds.
4. Remove the mouthpiece and exhale normally.
5. Take a small break between each deep breath.
6. Repeat this ten times, every hour.

Appendix 3. Daily incentive Spirometer Record Sheet

Time	**Pain Score**	**Incentive Spirometer** Tick if completed	**Effective** What colour ball was moved? How long did the patient inhale for?