Sickle Cell Disease:

Guideline for the use of Incentive Spirometry in Adults admitted with a Sickle Cell Crisis at risk of Acute Chest Syndrome

Profile	
Version:	*V1.0*
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Executive/Divisional sponsor:	*Dr Lisa Pickering, Divisional Chair*
Applies to:	*All staff involved in the care of patients with Sickle Cell*
Date issued:	*February 2019*
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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:
Consultant for adult haemoglobinopathy service

Please set out the legislation, guidance and best practice you consulted for this review:
• 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 • Guideline on the management of acute chest syndrome in sickle cell disease. Howard J et al. Br J Haematol. 2015 May;169(4):492-505

Please identify the key people you involved in reviewing this policy why, and when:

Summarise the key changes you have made and why:
n/a

Executive Summary

Acute chest syndrome (ACS) is one of the leading causes of mortality in patients with sickle cell disease (SCD) with 50% of all patients having at least one episode of ACS during their lifetime. Incentive spirometry has proven benefit in preventing ACS in patients with chest pain and should be considered in all patients with SCD.

This guideline contains instructions for the use of incentive spirometry to prevent ACS in patients with SCD.

1. Introduction

Acute chest syndrome is a life-threatening complication of sickle cell disease. The presentation may include the following signs and symptoms; fever, cough, wheeze, shortness of breath, increased work of breathing or hypoxia.

Patients with sickle cell disease admitted with vaso-occlusive crises are at risk of developing acute chest syndrome, particularly when suffering from back, chest and abdominal pain. Incentive spirometry in addition to pharmacological pain management has been observed to prevent atelectasis and reduce the incidence of acute chest syndrome[1,3,5]. **For management of acute chest syndrome see guideline on Management of Acute Complications in Adults patients with Sickle Cell Disease.**

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 Indications for use of incentive spirometry:

All patients with sickle cell disease who fulfil one or more of the following criteria:

1. Acute chest, back or abdominal pain
2. Chest infection
3. Radiological chest x-ray changes indicating atelectasis and/or infection
4. Increasing oxygen requirements to maintain target SpO2
5. Prolonged immobility

Contraindications:

1. Acute asthma/bronchospasm
2. Undrained pneumothorax

6.2 Initiation of incentive spirometry:

- Appropriate patient selection as per indications and contraindications above
- Any staff member competent in the use of incentive spirometers is able to implement the programme (see incentive spirometry programme below).
- Document in the patient's medical notes and ensure ward nursing team are aware.
- It is not necessary to refer to physiotherapy for initiation of incentive spirometry. However, a physiotherapy referral is indicated if there are concerns about retained secretions/atelectasis that does not improve post commencement of the incentive spirometry programme.
- If pain is limiting compliance, please ensure review of pain relief.

6.3 Incentive spirometry programme:

1. Identify target lung volume for treatment as per table 1, table 2 and table 3.

Please note if using Cliniflo device, progress gradually to target flow rate to achieve desired volume (Flow rate of 100 is easiest, 600 is hardest).

2. Ensure that the patient is sitting in an upright position
3. Set the programme: 10 maximal inspirations.

Split the 10 breaths into smaller sets of 2-3 breaths at a time to prevent hyperventilation.

Advise patient to use the incentive spirometer as instructed every hour from 08.00 to 22.00 while awake.

Please note this is a target guide and may need to be individualised for each patient.

4. If patient is requiring oxygen therapy, this should be continued via nasal cannula during breaths or via oxygen tubing attached to device (Cliniflo only, not possible with Spiroball device).

If on facemask oxygen FiO2 ≥ 40% consider whether it is appropriate/beneficial/safe to remove the face mask to use the incentive spirometer and consider use of nasal cannulae to facilitate.

5. Ensure that advice for cleaning the mouthpiece and information sheet are provided to the patient

6.4 Target volumes for treatment:

Guidance as referenced in Hudson RCI user guide for men

Age	Height 1.47m	1.52 m	1.57 m	1.62 m	1.68 m	1.73 m	1.78 m	1.83 m	1.88 m	1.93 m	1.98 m
20	2000	2200	2400	2600	2800	3000	3200	3400	3600	3800	4000
25	1950	2150	2350	2550	2750	2950	3150	3350	3550	3750	3950
30	1900	2100	2300	2500	2700	2900	3100	3300	3500	3700	3900
35	1800	2000	2200	2400	2600	2800	3000	3200	3400	3600	3800
40	1750	1950	2150	2350	2550	2750	2950	3150	3350	3550	3750
45	1700	1900	2100	2300	2500	2700	2900	3100	3300	3500	3700
50	1650	1850	2050	2250	2450	2650	2850	3050	3250	3450	3650
55	1550	1750	1950	2150	2350	2550	2750	2950	3150	3350	3550
60	1500	1700	1900	2100	2300	2500	2700	2900	3100	3300	3500
65	1400	1600	1800	2000	2200	2400	2600	2800	3000	3200	3400
70	1350	1550	1750	1950	2150	2350	2550	2750	2950	3150	3350
75	1300	1500	1700	1900	2100	2300	2500	2700	2900	3100	3300
80	1250	1450	1650	1850	2050	2250	2450	2650	2850	3050	3250

Guidance as referenced in Hudson RCI user guide for women

Age	Height 1.47m	1.52m	1.57m	1.62m	1.68m	1.73m	1.78m	1.83m	1.88m
20	1900	2100	2300	2500	2700	2900	3100	3300	3500
25	1850	2050	2250	2450	2650	2850	3050	3250	3450
30	1800	2000	2200	2400	2600	2800	3000	3200	3400
35	1750	1950	2150	2350	2550	2750	2950	3150	3350
40	1700	1900	2100	2300	2500	2700	2900	3100	3300
45	1650	1850	2050	2250	2450	2650	2850	3050	3250
50	1600	1800	2000	2200	2400	2600	2800	3000	3200
55	1550	1750	1950	2150	2350	2550	2750	2950	3150
60	1500	1700	1900	2100	2300	2500	2700	2900	3100
65	1450	1650	1850	2050	2250	2450	2650	2850	3050
70	1400	160	1800	2000	2200	2400	2600	2800	3000
75	1350	1550	1750	1950	2150	2350	2550	2750	2950
80	1300	1500	1700	1900	2100	2300	2500	2700	2900

Target Flow to achieve volume (As per DHD CliniFLO device guidance)

Time breath held	3s	4s	5s	6s	7s	8s
Flow rate						
100 ml/s	300 ml	400 ml	500 ml	600 ml	700 ml	800 ml
200 ml/s	600 ml	800 ml	1000 ml	1200 ml	1400 ml	1600 ml
300 ml/s	900 ml	1200 ml	1500 ml	1800 ml	2100 ml	2400 ml
400 ml/s	1200 ml	1600 ml	2000 ml	2400 ml	2800 ml	3200 ml
500 ml/s	1500 ml	2000 ml	2500 ml	3000 ml	3500 ml	4000 ml
600 ml/s	1800 ml	2400 ml	3000 ml	3600 ml	4200 ml	4800 ml

6.5 How to use the incentive spirometer:

1. Place the incentive spirometer mouthpiece in your mouth and seal your lips around the mouthpiece.
2. Take a slow deep/long breath in through your mouth.
3. Make sure that the yellow ball or disc stays on the smiley face throughout the breath in.
4. Try to hold the breath for *desired number of seconds* before breathing out
5. You will be given advice on how many times to repeat and frequency throughout the day.
5. You will show you how to take your Incentive Spirometer apart and put it back together correctly.
6. The mouth piece and tubing should be rinsed in warm, soapy water after each day and left to air dry. Do not place it in the dishwasher, boil it or bleach it.

Documentation:

- Initiation and details of the programme should be documented within the medical notes including date of issue of the device.

6.6 Discontinuation of Programme:

Medical staff, Respiratory / Haematology CNS or physiotherapist will be responsible for deciding when to discontinue the incentive spirometry programme. Patients should fulfil all of the following:

- Chest, abdominal and/or back pain subsided
- Mobilising independently
- No clinical signs of ongoing respiratory infection

OR

- Medical team, Respiratory CNS or Respiratory Physiotherapists consider patient unsuitable to continue for other reasons.

The incentive Spirometer can be taken home and reused if indicated for up to 3 months.

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.*

6. Associated documentation

7. References

Bellet at al (1995) Incentive Spirometry to Prevent Acute Pulmonary Complications in Sickle Cell Diseases. New England Journal of Medicine; 333:699-703

Howard J et al (2015). Guideline on the management of acute chest syndrome in sickle cell disease. Br J Haematol;169(4):492-505

Hsu et al (2005) Positive expiratory pressure device acceptance by hospitalised children with Sickle Cell Disease is comparable to incentive spirometry. Respiratory Care, 50(5):624-7

Kopecky et al (2004) Systemic exposure to morphine and the risk of acute chest syndrome in sickle cell disease. Clinical Pharmacology Therapy. March;75(3):140-6.
NHS Evidence National Library of Guidelines (2006) Sickle cell disease in childhood: standards and guidelines for clinical care

Ong G L (2005) Incentive spirometry for children with sickle cell disorder. Nursing times: Vol: 101, Issue: 42, Page: 55

Sylvester et al (2004) Pulmonary function abnormalities in children with sickle cell disease Thorax 2004 59: 67-70