Sickle Cell Disease - Surgery - Full Clinical Guideline

Reference no.:CG-HAEM/2015/001

1. Summary

This is a practical guideline and check list for assessing and treating patients with sickle cell disease requiring surgery under general anaesthetic.

2. Introduction

Sickle cell disease is a hereditary red cell disease. These patients are at increased risk of painful sickle cell crisis. Hence precautions need to be taken to reduce the chance of sickle crisis and other complications peri-operatively. This document provides guidance on the overall management.

3. Aim and Purpose

To offer guidance for all clinical staff assessing and treating adult patients with sickle cell disease (and not sickle cell traits/carriers) for any procedures under general anaesthetic at Royal Derby Hospital.

4. Definitions

SCD – Sickle Cell Disease

5. Surgery under general anaesthetic in sickle cell patients (Adult) – Full clinical guideline

Indications:

Surgery is not infrequently indicated in patients with sickle cell disorders. There is an increased requirement for cholecystectomy (the commonest operation performed in this patient group), splenectomy, hip replacement and ENT procedures. Tourniquets should not be used (ischaemia could lead to sickling). For minor procedures under local anaesthetic, there are no additional precautions to be undertaken. For procedures under general anaesthetic, there is the need for great care and precaution to reduce the chance of sickling and other complications peri-operatively.

It is now a standard practice with pre-op assessments when Sickle cell tests are done for patients of Afro-Caribbean or African origin, if status is not already known.

Planning in advance:

Details of surgery should include the operation date and the surgeon for communication. The timing of admission should be set well in advance, by liaison between surgeons and haematologist. This will enable advice to be given regarding risks, appropriate care and blood transfusion. For patients undergoing splenectomy, pre-operative immunisation is required.

The anaesthetist should be made aware of the patient in advance of the operation date, and given the chance to review the patient at the pre-operative assessment visit.

Key issues of relevance include:

- Baseline lung function
- Cardiac function
- Recent sickle cell crisis history, especially any chest syndromes
- Recent infections especially respiratory

Pre-operative care and care planning as below:

Assessment of requirement for post-op ITU care (eg if prophylactic CPAP and/or incentive spirometry) is indicated. Pre-operative training in respiratory physiotherapy exercise is advisable, especially for abdominal and thoracic surgery.

Pre-operative tests (well in advance)
1. Ensure full sickle genotype is known, including HbS%, HbA%, HbF%.
2. Full blood count
3. Consider screening for G6PD deficiency, if not already undertaken
4. UEs, LFTs.
5. Cross match sample: ensure any blood provided will be fully Rh and Kellcompatible, and HbS negative
6. ECG
7. CXR and lung spirometry
8. Echo: if > 18yrs old and not performed in last 2years
9. SaO2

Transfusion
In elective cases*, pre-operative transfusion should always be discussed with Dr S Hebballi.*
In emergency cases *on-call Consultant Haematologist should be consulted about the need for transfusions prior to surgery.*

Pre-operative transfusion, given either as a top-up transfusion or exchange transfusion (EXT), would be required for some SCD patients depending on the type of surgery, previous history of crisis, baseline Hb level and sickle phenotype.

It must be remembered that red cell units for sickle cell patients will usually have to be ordered in from the National Blood Service, which will increase the time it takes to make the blood available for the transfusion. Therefore in requesting blood for emergency transfusion, it is best to liaise directly with the blood bank staff, informing them that the blood is required for a sickle cell patient in addition to providing relevant details on the crossmatch request form.

After EXT the surgery date must not be changed. Top-up transfusion can be given immediately prior to the planned surgery. Post transfusion Hb should not exceed 100g/l. We need to be mindful that red cell transfusions always carry risks (eg red cell alloantibody

formation, transfusion reactions, transfusion-transmitted infection, hyperhaemolysis syndrome and a possible increase in post-op infections).

Peri-operative care:

Avoidance of dehydration which is likely to precipitate sickle crisis:
Encourage clear oral fluids until nil by mouth(NBM. If period of NBM is >2hrs then administer maintenance iv fluids. Maintenance iv fluids should be continued post-op until patient has adequate oral intake.

Thromboprophylaxis
All sickle cell patients should be considered at high risk of VTE and given prophylaxis unless a definite contra-indication exists. Usually it will be LMW heparin until discharge.

Oxygenation
Monitor SaO2 from the time pre-medication is administered
Maintain SaO2 >95% at all times with O2 if required
Hyper-oxygenate at induction of GA. Monitor SaO2 for at least 24hrs post-op and keep >95%. Liaise closely with physiotherapist and intensivists to minimise chest complications. The critical care team should be consulted early for respiratory support
Encourage use of incentive spirometry post-op in all cases of abdominal and thoracic surgery and in other cases where there is any concern about baseline lung function. If incentive spirometry devices not available, then 10deep breaths (or, in children under 6yrs, blow bubbles) every 2 hrs when awake.
Consider CPAP for 24hrs for major abdominal or thoracic surgery. Consider supplemental O2 on 2nd and 3rd post-op nights for major abdominal or thoracic surgery. Low threshold for use of bronchodilators (eg use of asthma, chest syndrome. Encourage early mobility

Temperature regulation
Chilling can lead to sickling. Ensure theatre and post-op recovery areas are warm. Consider warming iv fluids. Active warming techniques to be used post-op until patient is able to maintain body temperature. DO NOT use ice packs to reduce swelling

Infection management
Infections can precipitate sickling. Prophylactic antibiotics only to be used as per the surgical protocols. Post-splenectomy prophylactic penicillin (or erythromycin if allergic) should be commenced post-op. Maintain very active surveillance for post-op infections (wound, chest, other). Regularly assess iv cannula sites. Early targeted antibiotics if any signs of infection. Physiotherapy if chesty. If temperature >38, obtain cultures and start antibiotics according to local protocols.

Post-operative pain
Monitor pain closely and document on a scale of 1-10. Treat pain according to normal post-op protocols. Monitor respiratory rate carefully if using iv opiates/PCA.

DO NOT use ice packs to reduce swelling. Unusual pains, especially in sites away from operative site and /or in limbs, joints or chest wall/ribs is probably from sickle cell crisis so should be reported to the haematologist immediately. Refer to the Trust's Guideline on Management of an Acute Painful Crisis in Sickle Cell Disease Reference No: CG-T/2013/170 (as a link)

Post Operative care and prior to discharge

Post-operative transfusion should always be discussed with Dr S Hebballi or on-call consultant haematologist. Review by Haematology Doctor and Specialist Nurse for Haemoglobinopathies (available on 07469405987 directly or via Haematology Secretaries on Ext 87973) is mandatory. Review daily FBC, retics, Biochem, CRP. Prescribe usual medication. Inform Haemoglobinopathy nurse specialist of discharge. Check follow up arrangements. Patients with a sickle trait detected for the first time, in the pre-op assessment clinic, can be referred to the Haematology Specialist Nurse for Haemoglobinopathies (for counselling if patient requests)

6. References

TAPS, Transfusion Alternatives Preoperatively in SCD, study (Williamson et al Lancet 2013

Guideline on the management of Acute Chest Syndrome in Sickle Cell Disease , British Committee for Standards in Haematology (BCSH) Guidelines

East Midlands guidelines 'Sickle Cell Disease: Guidelines for peri-operative plans (adults)

7. Documentation Control

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