NHS SCOTLAND

SPAH
SCOTTISH PAEDIATRIC AND ADULT HAEMOGLOBINOPATHIES NETWORK

Scottish Paediatric & Adult Haemoglobinopathy Network

Paediatric Guideline - Surgery & Anaesthesia

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.

General anaesthesia in patients with sickle cell disease is associated with a significant risk for post-operative complications, especially acute chest syndrome. Surgery should be undertaken with close liaison and good communication between the surgeon, anaesthetist and medical and nursing staff.

PREOPERATIVE MANAGEMENT

Patients should be scheduled early on the operating list to ensure that they avoid prolonged fasting time and are unlikely to be cancelled. Consider IV hydration whilst fasting.

TRANSFUSION

Most patients with sickle cell anaemia (HbSS, Sβthal) are relatively asymptomatic with an Hb around 65g/l**.** This chronic steady state anaemia itself is not an indication for transfusion. The decision to transfuse a patient preoperatively depends on the type of operation and the patient's past sickle related complications. Patients with a history of recurrent chest crises or central nervous system disease or patients undergoing major surgery are at greater risk of developing sickle related problems peri-operatively.

Pre-operative transfusion:

Hb SS undergoing elective LOW* and MEDIUM*-RISK surgery

- 'top up' to a target Hb 100 g/l if Hb < 90 g/l is recommended

Hb SS undergoing elective HIGH-RISK* surgery

- offer exchange transfusion pre procedure (refer to Exchange Transfusion Guideline that is available on the SPAH website)
- Hb SC undergoing MEDIUM*- and HIGH-RISK* surgery
- Pre-operative transfusion (likely exchange) is recommended

The need for transfusion in patients with other genotypes should be assessed case by case, taking into consideration perceived severity of phenotype and complexity of surgery, when a detailed care plan should also be formulated pre-procedure, which should include the transfusion management (i.e.: 'top-up' x exchange).

Patients who need emergency surgery ideally should also be offered 'top up' transfusion if the Hb is low (<90 g/l), provided this does not result in unacceptable delay to the procedure. In cases where Hb is > 90 g/l, surgical risk is LOW* and pre-operative transfusion is expected to cause unacceptable delays, it is reasonable to proceed to surgery and make arrangements to transfuse the patient intra- or post-operatively if needed.

*Risk Stratification for Surgical Procedures (Koshy et al)

1. Low-risk surgeries: eyes, skin, nose, ears, distal extremities, dental, perineal, inguinal area (i.e.: inguinal hernia repair, myringotomy, dilatation, curettage).

2. Medium-risk surgeries: throat, neck, spine, proximal extremities, genitourinary system, intra-abdominal (i.e.: orthopaedic, abdominal, tonsillectomy, C-section, splenectomy, cholecystectomy).

3. High-risk surgeries: intracranial, cardiovascular, intrathoracic (i.e.: craniotomy, heart valve replacement).

INTRAOPERATIVE MANAGEMENT

Care should be taken to avoid factors that may precipitate a sickle crisis. These include hypoxia, cold, dehydration, pain and acidosis. The majority of crises in the perioperative period occur post-operatively.

Optimise oxygenation with anaesthetic agent per standard anaesthetic practice
Avoid hypoxia, (continuous pulse oximetry), hypercarbia, hyperventilation, over-hydration
Avoid or minimise tourniquets, avoid cold packs.

POST-OPERATIVE MANAGEMENT

Consider IV hydration if oral intake delayed post-operatively
Supplemental oxygen to maintain oxygen saturations >95% for 18-24 hours post-operatively
Incentive spirometry
Aggressive pain management
Encourage early ambulation
Consider pharmacological thromboprophylaxis if immobility anticipated for >72h.

References

1. Guidelines on red cell transfusion in sickle cell disease. Part II: indications for transfusion. Davis, BA. BJH, 2017, 176, 192-209.

2. The Transfusion Alternatives Preoperatively in Sickle Cell Disease (TAPS) study: a randomised, controlled, multicentre clinical trial. Howard, J. Lancet, 381, 930-938.

3. Koshy M. Surgery and Anaesthesia in Sickle Cell Disease. Blood, 86 (10), 1995; 3676-3684).