

GSTFT Clinical Practice Guideline

**Management of Children with Sickle Cell
Disease needing an anaesthetic**

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General Anaesthesia in Patients with Sickle Cell Disease

Sickle disease is a genetic disorder due to a single mutation resulting in the substitution of amino acid valine for glutamic acid in position six of the β -chain of the haemoglobin molecule. This change is responsible for the alteration in the state of the haemoglobin with increased viscosity and the development of the 'sickle' shape in deoxygenated states. The inheritance of a homozygous state Hb SS (Sickle cell anaemia) or double heterozygous (SC or SD, SE, SO or S β -thal) predisposed the patient to risk from hypoxic states leading to the risk of increased paracrystal formation, and the well known risk of vaso-occlusion, the prototype pathology.

Sickle cell complications such as bone necrosis, acute chest syndrome, renal impairment and gastro-intestinal ischaemia are thought to be precipitated by inappropriate general anaesthesia. It is therefore vital to provide the optimal setting for children prior to anaesthesia. The risk associated with general anaesthesia is controversial and needs further studies; however it is to maintain adequate oxygenation, hydration and pain control before, during and after surgery. The specialist Paediatrician with interest in haematology should be informed in advance of the admission. If a child has been transfused previously an extended antibody screen should be requested prior to transfusion. In the presence of positive antibody screen arrange appropriately cross-matched blood prior to surgery. The Hb level must be known before any surgery.

Children with sickle cell anaemia are not considered suitable for day case surgery in order to avoid the risk of severe crises due to poor fluid balance or delayed drug reaction:

- IVF should commence soon after going nil by mouth and continue until the child is drinking freely. The rate of fluid should be 100-150% of maintenance except there is risk of acute chest syndrome or otherwise indicated.
- Oxygen saturation should be maintained above 94% and encourage 2-hourly incentive spirometry for older children when they are fully awake.
- Pain: ensure adequate pain control and consider using iv / po morphine post anaesthesia

The risk of general anaesthesia is shown to be improved by appropriate patient selection and the use of blood products. We have divided these children into three different groups for management prior to surgery. The following are recommendations and should be discussed and agreed with all parties: the parents/child, Paediatrician/Haematologist and Anaesthetist.

Group 1

Where the surgical intervention is presumed to be minor and child does not have special risk factors, there may be no need for additional blood transfusion prior to surgery. The category of simple surgical procedures even though not exclusive may be considered of minimal risk of peri-operative complications e.g. insertion of grommets, surface surgery such as herniotomy or orchidopexy and minor dental surgery. The patient's steady state haemoglobin is considered adequate as long as the child is in a stable clinical condition. The Hb is usually about 6-8gm/dl

Group 2

Child with no special risk factors having intermediate risk surgery such as tonsillectomy or adenoidectomy (but moderate obstructive sleep apnoea), intra-abdominal surgery such as cholecystectomy: These may pose standard risk of complications and therefore it is suggested that:

Top-up transfusion to Hb 9-10gm/dl. Total Hb should not exceed 10 g/dl and it should happen at least 48 hours before the planned procedure. Remember always to request for antibody screen

Group 3

Those children who have a history of acute chest syndrome (ACS), stroke, or any sickle cell related organ damage or due any high-risk surgery such as thoracotomy: and **surgery involving posterior eye chamber**

Plan exchange transfusion or sequential top-up transfusions to achieve HbS level less than 30%. Exchange transfusion should occur at least 24 hours before surgery, and will need liaison with the paediatric medical staff, as well as organisation of a PICU bed and consideration of central line insertion under GA to facilitate the procedure.

Emergency surgery

Children requiring emergency surgery should have full blood count to determine the Hb and be cross matched at least for minor antigens and all cases must be discussed with the Paediatric Haematologist/ Paediatrician.

Children undergoing MRI brain scan under general anaesthesia for suspected cerebro-vascular disease

Transfuse to Hb 9-10 g/dl (total Hb not to exceed 10 g/dl except in cases when the S ratio had been reduced with chronic transfusion programme).

Use of surgical tourniquets

Tourniquets for surgery may be used with care in children with sickle cell trait or children on chronic transfusion programme (after careful exsanguination of the limb). However, the safety of tourniquets in non-transfused sickle cell disease patients is yet to be established and should be considered carefully on an individual basis.

Post-operative management

Children should be nursed in surgical HDU post-operatively. Pulse Oximetry should be recorded regularly and supplementary oxygen administered where necessary to ensure SpO₂ > 94%.

Avoid dehydration, and maintenance hydration at 100-150% of normal fluid requirement.

Pain control: It is necessary to consider using IV morphine and where necessary PCA / NCA (patient or nurse control anaesthesia). The standard analgesia should be given regularly and not as PRN, in order to get on top the pain.

References

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