

ANAESTHESIA AND SURGERY

This should ideally be done in a hospital with full service, PICU facilities and staff familiar with the care of Duchenne. There are a number of issues in providing anaesthesia for these patients, mainly respiratory but also consideration needs to be given if on steroids for steroid cover during the stress period of surgery.

The exclusive use of a total intravenous anaesthetic (TIVA) technique is strongly recommended, due to the risk of malignant-hyperthermia-like reactions and rhabdomyolysis when patients with duchenne are exposed to inhalational anaesthetic agents. **Depolarizing muscle relaxants like succinylcholine are absolutely contraindicated due to the risk of fatal reactions.**

In an emergency - appropriate referral is required:

• If a child is ambulant with no respiratory compromise - needs referral to an anaesthetist with paediatric experience.

• If a child is non-ambulant and has respiratory involvement (FVC <1.5L or <50%) - needs referral to the nearest hospital with PICU facilities once stabilised.

Remember:

DO NOT use depolorising muscle relaxant

DO cover for 'stress' if required when a child is on steroids.

To prevent the risks particularly post operative failure to extubate, atelectasis and pneumonia, appropriate assessment pre-surgery is required. If the pre-operative respiratory function is sub-threshold then non-invasive ventilation and cough assist techniques will need to be employed post surgery and these techniques need adequate training preoperatively to ensure a smooth course.

Pre-operative training in manual assisted cough techniques necessary:

• Baseline peak cough flow is <270LPM

• Baseline maximum expiratory pressure is<60 cm water (Older teenage and adult values)

Preoperative training in the use of nocturnal non-invasive ventilation is:

- Strongly recommended if baseline FVC <50% predicted
- Necessary if baseline FVC<30% predicted

After careful consideration of the above, patients may be eligible for surgery, albeit with increased risk, if preoperative training has been completed in assisted cough techniques and non-invasive ventilation.