

Practical Recommendations for Surgery and Anaesthesia in Myotonic Dystrophy.

Important general points to remember are:

- 1.) complications are not proportional to the severity of disease; they often arise in mildly affected patients.
- 2.) even though the operation may be uneventful, serious, even fatal problems may occur in the post-operative period; especially when opiates have been given as analgesia.

Pre-operative.

- 1.)Cardiological assessment- ECG essential, 24 hour tape if any indication of arrhythmia from ECG or history.
- 2.)Respiratory assessment - FEV1 and FVC both lying and standing.
Chest X-ray, noting elevation of diaphragm(s) or areas of atelectasis.
Arterial blood gases.
- 3.)Premedication -avoidance of opiates, caution with benzodiazepines.

Peri-operative.

- 1.) Tendency to temporomandibular dislocation - care needed in manipulating jaw.
- 2.) Induction- preferably gaseous;avoid hypnotic agents with slow metabolism such as thiopentone. Adverse reactions have also been reported with propofol. Lower doses are likely to be required.
- 3.) Relaxation- avoid suxamethonium.
short acting non-depolarising muscle relaxants are best used and may be needed in smaller doses; recovery from these may be prolonged.
- 4.) Neuromuscular monitoring is helpful, if available.
- 5.) ECG monitoring because of increased tendency to dysrhythmia.
- 6.) Reversal- neostigmine may produce ACh induced depolarisation blockade.

Post-operative (first 24 hours).

- 1.) Ensure respiration is fully re-established.
- 2.) Cardiac monitoring.*
- 3.) Respiratory monitoring - pulse oximetry, supplemented by arterial blood gas analysis.*
- 4.) Use of a high dependency bed is preferable.*
- 5.) Early chest physiotherapy- these patients are especially prone to post-operative chest infections and atelectasis.
- 6.) Minimal use of opiates for analgesia, instead explore other methods of analgesia, eg.local anaesthetic blocks or non-steroidal anti-inflammatory agents.

Summary -most problems occur when the disorder is unrecognised or when problems are not anticipated; it is worth considering whether regional anaesthesia is a viable alternative or even if the surgical procedure is really necessary.

* The extent to which these precautions are taken will depend on the length and nature of the procedure.