

**Practical Recommendations for Surgery and Anesthesia in
Myotonic Muscular Dystrophy
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Important general points to remember are:

1. Complications are not proportional to the severity of the 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 in any indication of arrhythmia from ECG of history.
2. Respiratory assessment:
 - a. FEV1 and FVC both lying and standing
 - b. Chest x-ray, noting elevation of diaphragm(s) or areas of atelectasis
 - c. Arterial blood gases
3. **Premedication avoidance of opiates, caution with benzodiazepines**

Peri-operative:

1. Tendency for 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:
 - a. Avoid suxamethonium
 - b. Short acting non-depolarising muscle relaxants are best used and may be needed in smaller doses; recovery from these may be prolonged.
 - c. Neuromuscular monitoring is helpful.
 - d. ECG monitoring because of increased tendency to dysrhythmia.
 - e. **Reversal – neostigmine may produce Ach induced depolarization blockade.**

Post-operative (first 24 hours):

1. Ensure respiration is fully re-established.
2. Cardiac monitoring*
3. Respiratory monitoring – pulse oximetry, supplemented by ABG.*
4. Use of 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 anesthetic blocks or non-steroid anti-inflammatory agents.

Summary: Most problems occur when the disorder is unrecognized or when problems are not anticipated; it is worth considering whether regional anesthesia 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.

REMEMBER: Problematic medications for MMD patients:

- Quinine, procainamide, tocainide (especially for those with cardiac problems)
- General anesthesia: Avoid thiopentone, suxamethonium, propofol, neostigmine, and halothane. Use fluranes, non-depolarizing blockers, spinal anesthesia, etc.
- Liquid paraffin
- Neuropletics (including metoclopramide)
- Opiates
- Benzodiazepines