Scottish Muscle Network

Anaesthetic guidance for patients with Myotonic Dystrophy

Peri-operative management of people with Myotonic Dystrophy

Myotonic dystrophy (Dystrophia Myotonica, DM1) is an inherited condition which can affect the central nervous system, cardiac, respiratory, gastrointestinal, endocrine and muscular systems in ways that can increase the risk of complications before, during and after surgery.

Patients with DM1 should be considered high risk during sedation and anaesthesia even if mildly affected. DM1 should not be a contraindication to surgery itself, with appropriate planning surgery should be uneventful.

General considerations

System	Considerations
Respiratory	Be aware of possible respiratory muscle weakness. Patients may use Non Invasive Ventilation (NIV) due to obstructive sleep apnoea. A poor cough may require additional measures to clear secretions. Patients may have a reduced response to hypoxia (low oxygen) and hypercarbia (high CO2).
Cardiac	Cardiac involvement is common and can occur in patients who are mildly affected. The commonest complications are conduction system disease and arrhythmias. There may be an increased risk of Cardiomyopathy.
Gastrointestinal	Laryngeal or pharyngeal weakness as well as delayed gastric motility predispose to aspiration of gastric contents. Post-operative ileus is common. Constipation is common.
Other	Endocrine problems such as diabetes and hypothyroidism are common. There may be neuropsychological, psychosocial or learning difficulties which may require extra support in the anaesthetic and recovery room.

Specific considerations

HDU/ ICU availability	Pre-operative discussion with Critical Care should be sought if there are any doubts over anaesthesia risks and definitely if the patient uses NIV. It is always better to have an HDU/ ICU bed booked and cancel if it is not required than the other way round.
Induction/ Recovery	A main theatre anaesthetic facility is preferential over a day surgery facility which would offer extra support at induction and recovery.
Anaesthetic agents	Suxamethonium should not be used; profound masseter spasm preventing intubation has been reported. Discuss and promote the use of regional techniques as well as TIVA techniques Patients are at an increased risk of respiratory and cardiac depression due to the effects of anaesthetic agents. There is no increased risk of malignant hyperpyrexia compared with the general population.
Type of Surgery	Diathermy may induce muscle contraction. This cannot be overcome with neuromuscular blockade and may make surgical access difficult. Upper abdominal surgery has the greatest risk. Day surgery and sedation can be complex due to atypical response to sedatives and anaesthetic agents.
Pregnancy	Increased risk of premature and prolonged labour. Increased risk of uterine atony and post-partum haemorrhage. The use of tocolytics and magnesium sulphate may exacerbate muscle weakness. Babies of affected mothers are at high risk of congenital DM1, characterised by hypotonia, feeding difficulties and respiratory weakness.

Pre-operative assessment

Respiratory:

Ask about problems with:

- Sleep including signs of daytime sleepiness
- Cough
- Frequency of chest infections
- Exercise tolerance
- Signs of Orthopnoea
- Use of NIV (If patient is using NIV make sure that they bring their ventilator on the day of surgery.)

Measure baseline:

- SpO_2. If <94% consider ABG.

Also consider measuring FEV_1, FVC, Peak cough flow and nocturnal TcCO2 study. Many will already be under the care of a respiratory or ventilation team.

Cardiac:

An ECG should always be performed, however a normal ECG does not exclude paroxysmal arrhythmia or conduction block. Any patient with an abnormal ECG should be referred for formal cardiology assessment. Also consider the need for intraoperative pacing. Question whether in addition to an ECG an echocardiogram would provide useful pre-operative information.

Ask about problems with:

- Syncope (Care should be taken before attributing syncopal episodes to postural hypotension unless there is a clear history and significant postural drop in blood pressure)
- Blackouts
- Dizziness
- Palpitations

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Gastrointestinal:

Ask about problems with:

- Dysphagia
- Reflux
- Constipation

Induction and maintenance of anaesthesia

- Avoid pre-medications (e.g. sedatives/ opioids) if possible.
- Cardiac and SpO2 monitoring is essential.
- Consider arterial line.
- Aspiration is a risk. Consider the use of sodium citrate, a PPI and metoclopramide. Consider a modified RSI with cricoid pressure.
- Minimal doses of induction agent should be used.
- Propofol induced pain has caused myotonia.
- Do NOT use suxamethonium.
- If muscle relaxants are required, then small doses of non- depolarising agents should be used.
- Use either an endotracheal tube or a supraglottic device with a gastric port e.g. iGEL, LMA supreme, ProSeal.

- Controlled ventilation through ETT or supraglottic airway is likely to be required to maintain normal end tidal CO_2.
- Keep temperature well controlled as shivering can lead to myotonia

Recovery from anaesthesia

- Neostigmine should be avoided
- Sugammadex is safe and effective for the reversal of rocuronium.
- Allow more time for recovery and ensure that there is full recovery of consciousness and muscle strength before removing ETT / LMA.
- If patient uses NIV extubation onto NIV is appropriate.
- If possible, avoid the use of nerve stimulators which may induce myotonia

Post operative period

- Be aware of respiratory depressant effects of small doses of opioids.
- Respiratory failure can occur slowly and insidiously in the post-operative period and close monitoring and awareness of signs and symptoms is crucial. Nursing staff must be alert to drowsiness, confusion, inability to lie flat and inability to clear secretions.
- Consider early referral for monitoring and consideration of NIV.
- SpO_2 should be measured regularly in post op period, consider an ABG if concerned.
- Patients may be prone to ileus – consider NG tube and early mobilisation.
- Patients may be prone to constipation consider prophylactic aperients.

Contacts

Please contact Dr Malcolm Sim for further advice if required, Consultant in Anaesthesia and Intensive care. Malcolm.Sim@ggc.scot.nhs.uk/ 0141 201 1100

NOTE: This guideline is not intended to be construed or to serve as a standard of care. Standards of care are determined based on 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.