Scottish Muscle Network

NHS SCOTLAND

Peri-operative management of people with Myotonic Dystrophy.

Myotonic dystrophy (or Dystrophia myotonica – DM1) is an inherited condition characterised by progressive muscle weakness and myotonia. Patients develop significant respiratory, cardiac, GI and endocrine problems, which may make surgery and anaesthesia difficult. Patients with DM1 should be considered high risk even if mildly affected. With appropriate planning surgery should be uneventful and return to premorbid function possible. DM1 should not be a contraindication to surgery itself.

General considerations.

Respiratory System –Patients may have respiratory muscle weakness and may use NIV. Patients also have a reduced response to hypoxia (low oxygen) and hypercarbia (high CO2). Patients may have a poor cough and may need extra measures to clear secretions. Obstructive sleep apnoea is common in DM1.

Cardiac system – Cardiac involvement is common and can occur in patients with minimal signs. The commonest complications are conducting system disease and arrhythmias. There may be an increased risk of cardiomyopathy. An ECG **must** be performed.

Gastrointestinal system – laryngeal or pharyngeal weakness as well as delayed gastric motility predispose to aspiration of gastric contents. Post op ileus is common. Constipation is common.

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.

Other – Endocrine problems such as diabetes are common. There may be neuropsychological, psychosocial or learning difficulties which may require extra support in the anaesthetic and recovery room.

Anaesthetic agents – Suxamethonium should be avoided, profound masseter spasm preventing intubation has been reported. Regional techniques are safe and should be used where possible. Patients are at increased risk of respiratory and cardiac depression effects of anaesthetic agents. There is no increased risk of malignant hyperpyrexia compared with the general population.

Surgery – diathermy may induce muscle contraction. This cannot be overcome with neuromuscular blockade and may make surgical access difficult.

Pre-op assessment.

Respiratory – Ask about problems with sleep, cough, frequent chest infections, exercise tolerance, use of NIV and orthopnoea. Measure baseline SpO_2. If <94% consider ABG. Consider measuring FEV_1, FVC, Peak cough flow and nocturnal $TcCO_2$ 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. Care should be taken before attributing syncopal episodes to postural hypotension unless there is a clear history and significant postural drop in blood pressure.

Any abnormal ECG should be referred for formal cardiology assessment.

Consider the need for intraoperative pacing

GI – Ask about problems with dysphagia, reflux and constipation

Type of surgery – Upper abdominal surgery has greatest risk. Day surgery and sedation can be complex due to atypical response to sedatives and anaesthetic agents. Consider HDU / ICU. Consider extra support at induction and recovery.

Anaesthesia - Discuss and promote the use of regional techniques as well as TIVA techniques

Other - If patient is using NIV make sure that they bring ventilator on day of surgery.

Induction and maintenance of anaesthesia.

Aspiration is a risk. Consider the use of sodium citrate, ranitidine and metoclopramide.

A modified RSI with cricoid pressure should be considered. 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 a 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. Maintaining normal end tidal CO_2 is preferable.

Recovery from anaesthesia.

Neostigmine should be considered but used in small doses as may cause myotonia. 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.

Post operative period.

Be aware of respiratory depressant effects of small doses of opioids

Respiratory failure can occur slowly and insidiously in the post op period and close monitoring and awareness of signs and symptoms is crucial. Nursing staff must be alert to drowsiness, confusion, inability to lie flat, inability to clear secretions.

Consider early referral for monitoring and consideration of NIV

SpO_2 should be measured regularly in post op period, consider $TcCO_2$ or ABG if concerned.

Contacts.

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This guideline is not intended to be construed or to serve as a stand[1]ard of care. Standards of care are determined on the basis of 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.

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