Scottish Muscle Network

Summary: Peri-operative management of people with Myotonic Dystrophy

Please note full guidance available is here

Pre-operative assessment:
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. Keep temperature well controlled throughout as shivering can lead to myotonia

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.)
If baseline SPO2 is less than 94% consider ABG. Also consider measuring FEV_1, FVC, Peak cough flow and nocturnal TcCO2 study.

Cardiac:
12 lead ECG is essential, if abnormal refer to cardiology. Consider the need for intraoperative pacing and if echocardiogram would provide useful pre-operative.
Ask about problems with Syncope, Blackouts, Dizziness and Palpitations

Gastrointestinal:
Ask about problems with Dysphagia, Reflux and Constipation

Induction and maintenance of anaesthesia

Medications:

- Avoid pre-medications (e.g. sedatives/ opioids) if possible.
- 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.

Monitoring:

- Cardiac and SpO2 monitoring is essential.
- Consider arterial line.

Airway:

- Aspiration is a risk. Consider the use of sodium citrate, a PPI and metoclopramide. Consider a modified RSI with cricoid pressure.
- 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.

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

- 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.
- SpO2 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.

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