Anesthetic Management of Patients with Myotonic Dystrophy – Risks & Recommendations

Quick Reference Version

Myotonic dystrophy (DM) is a genetic disorder that affects CNS, cardiac, respiratory, gastrointestinal, endocrine, and muscular systems in ways that increase the risk of anesthesia.

Anesthesia Guidelines for pre-operative, intra-operative and post-operative care of DM patients, summarized below, can be found at at **https://www.myotonic.org/toolkits-publications**

Anesthetic Risks, as detailed in the Guidelines, result from DM effects that include:

- Cardiac conduction defects and potentially fatal arrhythmias
- Ventilatory insufficiency and poor airway protection
- Gastrointestinal dysmotility that frequently results in pseudo-obstruction
- Erratic responses to succinylcholine - though DM does not increase true malignant hyperthermia reactions, this drug should not be used in DM patients
- Prolonged and heightened sensitivity to sedatives and analgesics so that serious complications, including heightened risk of aspiration, are most common in the post-anesthesia period due to drug induced:
    - Reduction in level of consciousness
    - Exaggerated ventilatory weakness
    - Pharyngeal dysfunction with reduced airway protection
    - Gastrointestinal dysmotility and potential pseudo-obstruction

Methods to mitigate risk, detailed in the Guidelines, are summarized below:

- Preoperatively evaluate pulmonary, cardiac and gastrointestinal DM features in addition to its neurological and neuromuscular effects
- Use regional anesthesia when possible, to reduce or eliminate the need for general anesthesia
- Avoid pre-medications (e.g. sedatives and opioids) to the extent possible
- Keep the patient warm
- Consider precautionary application of defibrillator/ pacer pads
- On induction, anticipate aspiration, and avoid the use of succinylcholine
- Adhere to strict extubation criteria. Given DM effects on CNS, GI, ventilatory and pharyngeal function, prepare the patient for prolonged post-anesthesia mechanical ventilation, commonly after having fully regained consciousness
- Prepare the patient for prolonged ventilatory assistance, for example by prior initiation of BiPAP with a mask that is immediately available post-anesthesia
- Plan for continuous SpO2 and ECG monitoring post- anesthesia until the patient fully regains pre-operative status, or longer if analgesics or sedatives are used in the post-anesthesia period
- Manage postoperative pain without narcotics when possible
- Encourage aggressive pulmonary toilet after anesthesia, including by use of a mechanical cough- assistance device if necessary

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