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