

# PRACTICAL SUGGESTIONS FOR THE ANESTHETIC MANAGEMENT OF A MYOTONIC DYSTROPHY PATIENT

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## FOREWORD:

The anesthetic management of patients with myotonic dystrophy (dystrophia myotonica, DM) can be challenging. "Complications are not proportional to the severity of the disease; they often arise in mildly affected patients" (15). Indeed, there are multiple reports within the medical literature that detail poor outcomes related to the following complications: loss of airway secondary to medication-induced respiratory depression; aspiration of stomach contents; sudden death that is usually secondary to cardiac conduction delays and dysrhythmias. One must consider if, in light of these complications, "regional anesthesia is a viable alternative or if the surgical procedure is really necessary" (15).

The following points about myotonic dystrophy in this foreword can help a vigilant anesthesiologist avoid complications and provide safe anesthesia care to DM patients presenting for surgery:

- 1. General:** "Myotonia" is described as muscle contraction (voluntary or otherwise) with abnormal, prolonged relaxation (3). Triggers for myotonia include certain medications, potassium, hypothermia, shivering, or any mechanical or electrical stimulus (2, 3, 4). Patients also exhibit profound skeletal muscle weakness secondary to muscle degeneration.
- 2. Medications:** DM patients are exquisitely sensitive to the respiratory depressant effects of anesthetic medications (3). Be sure to have appropriate airway and monitoring equipment available when using these medications, and prepare for the likelihood of postoperative mechanical ventilation until strict extubation criteria are met. In addition, postoperative pain control should be managed with NSAIDs, regional techniques using local anesthetics, and acetaminophen when possible. If opioids are employed (systemic or neuraxial), then ICU care and continuous pulse oximetry must be considered given the high risk for respiratory depression and aspiration.

3. **Airway:** Rapid sequence induction with cricoid pressure is recommended. Weakness of the pharyngeal muscles and a delayed gastric emptying time predispose DM patients to aspiration (3, 21). Also, succinylcholine effects are unpredictable in DM patients: one case report describes jaw rigidity and impossible intubation after succinylcholine administration (19); prolonged laryngospasm and cyanosis has been reported in myotonia congenita, but could theoretically also occur in DM (20). Avoid succinylcholine when possible.
  4. **Respiratory System:** The effects of myotonic dystrophy on the respiratory system are profound and common (1, 24). Respiratory muscle weakness predisposes DM patients to restrictive lung disease with concurrent dyspnea and ineffective cough (3). Moreover, arterial hypoxemia and a diminished ventilatory response to hypoxia and hypercapnia are frequent associations (3). Accordingly, these factors place DM patients at an increased risk for pneumonia and other perioperative pulmonary complications (1). Ventilatory weakness contributes to the complex sleep disorders of DM, which frequently results in profound pre-operative sleep deprivation that further complicates post-anesthetic care.
  5. **Cardiac System:** DM patients can have cardiac abnormalities that may lead to sudden death secondary to various cardiac conduction delays or other dysrhythmias (3, 6). Thoroughly evaluate the cardiac system – including echocardiogram, 12-lead EKG, and interrogation of the internal cardiac rhythm device (if present) – before any anesthetic care is given.
  6. **Central Nervous System:** The many CNS effects of DM further complicate perioperative care. For example, behavioral and cognitive problems in the patient and other family members can complicate pre-operative preparation. Hypersomnia is a common and sometimes the primary manifestation of DM that can result from a narcolepsy-like central hypersomnia as well as sleep-related ventilatory insufficiency or obstructive sleep apnea, any of which can lead to profound sleep deprivation in the pre-operative period as well as multiple management difficulties post-anesthesia. Also, DM subjects have heightened CNS sensitivity to sedatives, anxiolytics and analgesics, further impeding ventilatory drive and airway protection. Perioperative casualties often develop several days post-operatively due to aspiration or inadequate monitoring of hypoxia, during the period in which DM patients become increasingly encephalopathic due to sleep deprivation or the unintended effects of medication.
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## **PRE-OPERATIVELY:**

1. **Multi-disciplinary medical team:** It is well documented that the medical and surgical management of patients with myotonic dystrophy (DM) can be challenging and fraught with complications (1, 3, 4, 6, 21). For these reasons, coordination of the pre- and post-operative plans for care should be made at least 1-4 weeks in advance using a multi-disciplinary medical team. This team would ideally consist of the surgeon, anesthesiologist, primary care physician, intensivist, and, if indicated, the pulmonologist and cardiologist (see 'consultations' below). Furthermore, the addition of a neuromuscular specialist (i.e. neurologist) with expertise in the pathophysiology and natural course of DM would be highly beneficial.

- 2. Pre-anesthetic evaluation:** In addition to a comprehensive preoperative evaluation completed by the DM patient's primary care physician (PCP), an anesthesiologist should perform a careful and detailed pre-anesthetic assessment 1-4 weeks prior to surgery. Such an evaluation may prevent serious complications and fatalities in DM patients. Their abnormal and often unpredictable responses to common anesthetic medications are well described in the medical literature and were summarized in the foreword (2, 3, 5, 7, 8, 11, 18, 19, 20). Indeed, a thoughtful and comprehensive anesthetic plan is warranted in advance of the day of surgery. The absence of such a preoperative evaluation by the PCP (at minimum) and the anesthesiologist could be considered cause for case cancellation.

The anesthesiologist should devote particular attention to the cardiopulmonary systems during their pre-anesthetic evaluation. It is not uncommon for DM patients to have a history of hypoxia, dyspnea, sleep apnea requiring CPAP, or marked ventilatory muscle weakness necessitating BIPAP. Given the anesthetic implications of these disorders, a measure of their severity is warranted. In addition, further questioning should involve whether or not the DM patient has a history of arrhythmia, heart failure, and/or an internal cardiac rhythm management device. All internal cardiac rhythm devices require interrogation by a cardiac electrophysiologist. A baseline echocardiogram, 12-lead EKG, and a chest radiograph should also be completed preoperatively (see 'consultations' below).

- 3. Consultations:** Based on the PCP's and anesthesiologist's preoperative evaluations and assessments, a thorough cardiopulmonary evaluation by a cardiologist and pulmonologist completed at least 3 weeks prior to elective surgery may be necessary because of the high incidence of morbidity and mortality related to these systems (1, 6). Indeed, all DM patients presenting for surgery should have a preoperative baseline echocardiogram, a 12-lead EKG, and a chest radiograph, each with expert interpretation. Moreover, all internal cardiac rhythm management devices must be interrogated by a cardiac electrophysiologist prior to entering the operating room. Pulmonary function tests (including supine and sitting vital capacities) and preoperative arterial blood gases may also be useful and should be requested at the discretion of the primary or consulting physician(s) before elective surgery.
- 4. Premedications:** DM patients can be exquisitely sensitive to the respiratory depressant effects of commonly used premedications (e.g. opioids and benzodiazepines). Therefore, make sure that appropriate equipment for monitoring and performing urgent intubation are available prior to the administration of premedication, or any other sedative. DM patients also frequently suffer from gastroparesis, predisposing them to episodes of acute pseudo-obstruction, which can be further exacerbated by opioids, further complicating ventilatory function and airway protection.
- 5. Regional anesthesia:** Regional anesthesia including neuraxial techniques have been described in the literature as successful primary anesthetics for DM patients (3, 22). They can help avoid some of the frequent complications associated with general anesthesia in the DM patient. However, there are case reports that describe an "incomplete motor block and shivering sufficient to stimulate myotonic contractures with epidural anesthesia" ([direct quote from 12], 13, 14) in DM patients. After the risks and benefits of regional anesthesia are assessed, techniques should ultimately be employed when applicable.

## **INTRA-OPERATIVELY:**

### **1. Environment:**

- a. Hypothermia and shivering can induce a myotonic contracture (2). Therefore, keep the operating room and table warm so that the patient will be better able to maintain a normal body temperature.
- b. Use warmed IV fluids as well as forced-air blankets during surgery.

### **2. Monitoring:**

- a. Employ standard American Society of Anesthesiologists (ASA) monitors including thermometer (3).
- b. Strongly consider attaching an external pacer/defibrillator to the patient. DM patients are at high risk for arrhythmias and sudden death (6).
- c. Consider placing an arterial line in order to verify the adequacy of oxygenation and ventilation via blood gas interpretation, and for continuous blood pressure monitoring.
- d. Monitor neuromuscular blockade with a peripheral nerve stimulator, but do so with caution: the electrical stimulus could induce a myotonia and be misinterpreted as sustained tetany indicative of full reversal of neuromuscular blockade (2).
- e. Invasive cardiac monitoring (TEE, PA catheters, CVP lines) should be reserved for DM patients that have significant cardiopulmonary dysfunction. The cardiologist's pre-operative consultation and assessment may help guide the decision of whether to employ these monitors.

### **3. Induction:**

The superiority of one specific induction agent over another has not been established for elective surgeries. Etomidate, thiopental, and propofol have all been used safely for induction. However, using agents with a short beta half-life seems logical to minimize the possibility of prolonged postoperative mechanical ventilation.

- a. Ensure adequate pre-oxygenation.
- b. DM patients are at risk for aspiration secondary to their dysphagia and altered gastric motility (21). Therefore, consider administering sodium citrate, an H<sub>2</sub>-antagonist, and/or metoclopramide prior to induction. Lastly, a rapid sequence induction with cricoid pressure is warranted.
- c. Rapid Sequence Induction:
  - i. Maintain cricoid pressure
  - ii. A hypnotic agent with a short beta half-life (e.g. propofol) is recommended in light of the exaggerated apneic response characteristic of DM patients. Titrate the hypnotic to effect—a lower dose is likely to be sufficient in a DM patient.

- iii. Avoid succinylcholine. The DM patient's response to succinylcholine is unpredictable and may lead to a difficult or impossible intubation secondary to exaggerated contracture, masseter spasm, and laryngospasm (2, 19, 20). In addition, "because of dystrophic muscle changes, it is possible that in advanced cases succinylcholine might result in an exaggerated hyperkalemic response" (9).
  - iv. Tracheal intubation can be successful in DM patients without a muscle relaxant (9). If a muscle relaxant is needed, then a non-depolarizing agent with a short recovery index should be chosen (e.g. Rocuronium, Cis-atracurium) (7).
  - v. The temporomandibular joint may have a tendency to dislocate in DM patients. Laryngoscopy and jaw manipulation should be done with care (15).
- d. Difficult Airway: Follow the *ASA Difficult Airway Algorithm* (23).

#### 4. Maintenance:

- a. **Volatile agents:** DM patients are no more susceptible to the development of malignant hyperthermia than the rest of the general population (16, 17). Volatile anesthetics are effective for maintenance of anesthesia, but they may exacerbate a patient's cardiomyopathy secondary to their myocardial depressive effects. In addition, desflurane, for example, may be the agent of choice considering its theoretical advantage of faster emergence upon completion of surgery (3).
- b. **Muscle relaxation:** If possible, avoid muscle relaxants altogether and maintain akinesia with deep inhalational/intravenous anesthesia, or have the surgeon infiltrate the skeletal muscle tissue within the surgical field with local anesthetic. When further muscle relaxation is required, use a non-depolarizing agent remembering that DM patients will exhibit an exaggerated response to it. Therefore, initial doses should be reduced while subsequent doses titrated to effect via the peripheral nerve stimulator (2).
- c. **Intravenous agents:** Safe and effective anesthesia using propofol and remifentanyl for total intravenous anesthesia has been described in the medical literature (4, 5).
- d. **Intravenous Fluids:** Consider using crystalloid fluids that do not have any added potassium. DM patients have reduced Na<sup>+</sup>-K<sup>+</sup> pump capacity and may be prone to the development of hyperkalemia (10). There is no apparent contraindication to the use of colloids.

#### 5. Emergence:

- a. **Reversal agents:** Neostigmine has been purported to induce myotonia (18). Therefore, avoid its use and plan for the non-depolarizing muscle relaxant effect to simply wear off.
- b. **Extubation:** Considering the multi-systemic effects of DM (cardiopulmonary pathology, profound peripheral weakness, altered gastric motility, pharyngeal weakness with poor airway protection, increased sensitivity to all anesthetic medications) adhere to strict extubation criteria. These patients may need supportive mechanical ventilation in the PACU and perhaps in the ICU until extubation criteria are met. Additionally, be aware that there is an increased risk of delayed-onset apnea after extubation during the immediate 24 hours after surgery, and even

longer if post-operative opioid analgesics are administered. Close and continuous monitoring of cardiopulmonary function (SpO2 and EKG) is needed during this time period.

- c. **Disposition:** Consider ICU admission if there is an anticipated need for mechanical ventilation, significant opioid analgesia, or other necessary critical care management.

## **POST-OPERATIVELY:**

Admission to the intensive care unit (ICU) for postoperative management should always be considered given the significant complications that may occur as a result DM. At the very least, patients should be monitored postoperatively with continuous pulse oximetry and EKG for a period of 24 hours. Below are specific points that support these recommendations:

### **1. Pain Control:**

- a. First and foremost, consider the use of regional anesthesia, NSAIDS, and acetaminophen (rectal or oral) for control of postoperative pain. If these medications/modalities are contraindicated, then the use of opioids must be administered with caution and vigilant monitoring (see below).
- b. The exquisite sensitivity of DM patients to the respiratory depressant effects of opioids (systemic or neuraxial) can equate to fatal outcomes in the postoperative period. The most common route of opioid administration that places DM patients at high risk for respiratory depression is intravenous, yet there is a case report that details respiratory depression following a small dose of epidural morphine as well (8). Another case report demonstrated adequate analgesia with epidural opioid administration without respiratory depression (11). Ultimately, these patients need to be closely monitored. An ICU is therefore the safest environment in which to administer postoperative opioids, titrating them to effect. Lastly, be aware that opioids can exacerbate one of the common features of DM, gastrointestinal paresis. Depending on the severity, gastroparesis could increase the risk of reflux and aspiration.

### **2. Pulmonary Considerations:**

In a retrospective analysis of 219 DM patients who underwent surgery under general anesthesia, Matheiu et al found that most perioperative complications were related to the pulmonary system (1). In particular, DM patients who were symptomatic, who underwent upper abdominal surgery, or who had severe muscular disability were especially at risk. Therefore, "careful monitoring during the early postoperative period, protection of the upper airways, chest physiotherapy, and incentive spirometry are mandatory" (1).

It can not overstated just how important continuous monitoring is in a DM patient during the postoperative period, especially if ventilatory function is compromised secondary chest or abdominal surgery, pain, or muscle weakness inherent of the disease. Delayed-onset apnea is most likely to develop in the first 24 hours postoperatively, and an exaggeration of any baseline hypersomnia could become apparent with morbid results. An ICU would be most appropriate for the detection and treatment of these complications should they arise.

## SUMMARY:

1. Perform an extensive preoperative evaluation. Organize a multi-disciplinary medical team.
2. Use regional anesthesia when appropriate.
3. Be cautious with premedications (benzodiazepines and opioids).
4. Keep the patient warm.
5. Consider applying defibrillator/pacer pads.
6. On induction, be aware of the high likelihood of aspiration and other airway complications. Avoid succinylcholine when possible.
7. Adhere to strict extubation criteria. Given the effects DM has on the pulmonary system, anticipate the need for supportive mechanical ventilation until extubation criteria are met.
8. Plan for the continuous SpO<sub>2</sub> and EKG monitoring postoperatively.
9. Manage postoperative pain with NSAIDs, regional techniques, and acetaminophen when appropriate. Use opioids with extreme caution.
10. Encourage aggressive pulmonary toileting postoperatively.

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