Myotonic Dystrophy FOUNDATION

Practical Suggestions for the Anesthetic Management of a Myotonic Dystrophy Patient

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Overview

Risks of anesthesia in myotonic dystrophy are most significant in the post-anesthesia period, but can be controlled by appropriate management.

During anesthesia, risks stem from the multisystemic features of myotonic dystrophy.

- DM does not increase risk of true malignant hyperthermia reaction beyond that of the general population, though erratic responses warrant avoidance of succinylcholine
- Cardiac rhythm and conduction defects need to be closely monitored.
- Ventilatory failure and poor airway protection require vigilant monitoring and support.
- Gastrointestinal dysmotility requires careful monitoring.

Possible post-anesthesia complications reflect heightened sensitivity and prolonged interaction of sedatives and analgesics with various aspects of myotonic dystrophy.

- Resultant clinical effects of sedation and analgesia include:
    a. Reduced level of consciousness
    b. Impaired ventilatory function
    c. Heightened pharyngeal dysfunction and aspiration
    d. Increased gastrointestinal dysmotility
- The risks can be successfully mitigated by careful and sustained monitoring

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Key management issues

Anesthetic management of patients with myotonic dystrophy (dystrophia myotonica or DM) can be challenging. With this in mind, all procedures requiring anesthesia should be seriously considered by patients, their families, and their physicians to determine whether they are truly necessary. Details about the potential problems merit discussion, especially in patients with myotonic dystrophy type 1 (DM1) as they relate to the form of disease (congenital; childhood; adult onset) and type of anesthesia required (local; regional; spinal; general; or other combination anesthesia protocols).

The following are some of the key management issues for the anesthesiologist to consider when caring for a patient with DM:

1. **General:** Myotonic dystrophy was identified because of its unique effects on skeletal muscle, but was subsequently shown to result in direct effects on most organs, including the CNS, eyes, heart, endocrine, GI and pulmonary systems. Two genetic forms of myotonic dystrophy have been identified: DM1 (Steinert disease) and DM2 (PROMM, proximal myotonic myopathy). Although DM1 patients can present at any age, those with DM2 present in adulthood, and generally have less severe symptomatology than DM1 patients (26, 29). In the congenital and childhood forms of DM1, patients often have narrow facies, a high-arched palate and limited ability to open their mouths fully (24, 30), all potentially complicating intubation. Discussion of intubation procedures along with the issues related to safe pre-anesthesia and post-anesthesia management in a hospital setting with care providers experienced in the treatment of patients with DM1 is needed. Special considerations may also be needed in planning anesthesia for pregnancy/delivery of mothers with DM1 (31) and for procedures involved with in vitro fertilization (32).

2. **Muscle:** Myotonia is muscle contraction with abnormal, prolonged relaxation (3). Exacerbants of myotonia include medications, potassium, hypothermia, shivering, and any mechanical or electrical stimulus (2, 3, 4). Patients with DM also exhibit muscle weakness of varied severity that is secondary to muscle atrophy and degeneration.

3. **Medications:** DM1 patients are exquisitely sensitive to the respiratory depressant effects of analgesics and anesthetics (3). When possible, alternatives to opioids and regional techniques with local anesthetics should be utilized. If opioids are administered (systemic or neuraxial), then continuous pulse oximetry and possibly ICU-level care should be considered given the high risk for respiratory depression and aspiration. Succinylcholine's effects on muscle are unpredictable in DM1 patients and may result in hyperkalemia.

4. **Airway:** Weakness of pharyngeal muscles and a delayed gastric emptying time predispose DM patients to aspiration (3, 21).

5. **Respiratory system:** 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). DM1 patients are especially at risk for post-operative atelectasis and pneumonia as well as respiratory insufficiency requiring prolonged ventilation. A decreased vital capacity pre-operatively serves as an important warning that pulmonary and intensive care unit specialists will need to be part of the team mentioned below to evaluate the respiratory system and plan appropriate post-operative care.

6. **Cardiac system:** DM patients can have cardiac abnormalities that may lead to sudden death secondary to cardiac conduction delays and arrhythmias (3, 6). Due to the prevalence and progressive nature of conduction defects, all DM patients should have an ECG checked annually. Before administration of anesthesia, an ECG should be repeated and any internal cardiac rhythm devices should be interrogated. When indicated by history and physical exam, a pre-operative Holter monitor examination or echocardiogram may be warranted.

7. **Central nervous system:** Hypersomnia is a common and sometimes primary manifestation of DM. It can result from a combination of narcolepsy-like central hypersomnia, sleep-related ventilatory insufficiency, obstructive sleep apnea and periodic leg movement of sleep, any of which can lead to multiple management difficulties post-anesthesia, including prolonged encephalopathy. Potentially profound sleep deprivation can worsen many of the aforementioned multisystemic features of DM.

Pre-operative management issues

1. **Multi-disciplinary medical team:** It is well documented that the medical and surgical management of patients with 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 1-4 weeks in advance of elective procedures using a multi-disciplinary medical team. This team would ideally consist of the neuromuscular specialist, primary care physician, surgeon and anesthesiologist and, if indicated, a pulmonologist and cardiologist. A muscular-impairment rating scale (MIRS) grade determination may be useful as it has been shown to be predictive of perioperative adverse events (25).

2. **Pre-anesthetic evaluation:** In addition to a comprehensive preoperative evaluation completed by the DM patient's neuromuscular specialist, pediatrician/primary care physician (PCP), pulmonary or cardiology consultants (based upon the severity of cardiopulmonary manifestations) along with an anesthesiologist, should perform a careful and detailed pre-anesthetic assessment prior to surgery. The anesthesiologist should devote particular attention to the cardiopulmonary systems. 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. There is also the potential difficulty of airway access due to a narrow face, high-arched palate and limited ability to open the mouth noted previously. Given the anesthetic implications of these symptoms, a measure of their pre-anesthesia severity is warranted. Specific history of dysphagia, dysarthria, dysphonia and aspiration should also be addressed as indication of pharyngeal involvement. Effectiveness of coughing and familiarity with cough assist equipment and BIPAP mask in advance of surgery is very important in assuring successful post-operative care in both children and adults with DM1, especially those with decreased vital capacity as well as difficulty with pharyngeal function; if possible the patient's own BIPAP mask should be available for post-anesthetic use. In addition, further questioning should assess any history of cardiac conduction abnormalities, arrhythmia, heart failure, and presence of an internal cardiac rhythm management device.

3. **Consultations:** Based on the preoperative evaluations and assessments, a preoperative consultation by a cardiologist and pulmonologist may be beneficial. Given the prevalence of conduction defects a preoperative 12-lead ECG is always recommended; an echocardiogram and chest radiograph can be considered, and preoperative cardiac rhythm monitoring may be needed to assess presence of arrhythmia. Moreover, all internal cardiac rhythm management devices should be interrogated (and possibly reprogrammed) prior to entering the operating room. Pre-anesthesia pulmonary function tests are always indicated (ideally including supine and sitting vital capacities) and arterial blood gases may be useful.

4. **Premedications:** DM patients can be exquisitely sensitive to the respiratory depressant effects of commonly used premedication (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.

5. **Regional anesthesia:** Neuraxial anesthetic techniques have been described in the literature as successful primary anesthetics for DM patients (3, 22), as they can help avoid some of the frequent complications associated with general anesthesia. However, there are case reports that describe shivering sufficient to stimulate myotonic contractures with neuraxial anesthesia, as well as incomplete blocks in patients with DM (12, 13, 14).

Intra-operative management issues

1. **Environment:** Hypothermia and shivering can induce a myotonic contracture (2). Therefore, the operating room should be kept warm so that the patient will be better able to maintain a normal body temperature. Warmed IV fluids as well as forced-air blankets should be used during surgery.

2. **Monitoring:**

   a. Employ standard American Society of Anesthesiologists (ASA) monitors including a thermometer (3).

   b. 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 myotonia and be misinterpreted as an indication that neuromuscular blockade has been fully reversed(2).

   e. Invasive cardiac monitoring (TEE, PA catheters, CVP lines) should be reserved for DM patients with significant cardiopulmonary dysfunction. The cardiologist's pre-operative consultation and assessment may help guide the decision of whether to employ these monitors.

3. **Induction:** Etomidate, thiopental, and propofol have all been used safely for induction. Generally, DM patients are more sensitive to these agents and thus require a decreased dose. Adding lidocaine to propofol for induction may reduce the incidence of pain-induced myotonic contractures (27).

   DM patients are at risk for aspiration secondary to their dysphagia and altered gastric motility (21). Therefore, consider administering sodium citrate, an H2-antagonist, and metoclopramide prior to induction. A rapid sequence induction with cricoid pressure should be considered. However, there are reports in the literature of successful use of supraglottic airways and/or mask ventilation alone, especially in children (25).

   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, the use of succinylcholine may result in hyperkalemia in cases of severe disease (9). When possible, tracheal intubation should be attempted 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). The temporomandibular joint may have a tendency to dislocate in DM patients, therefore laryngoscopy and jaw manipulation should be done with care (15).

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. Desflurane 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. Therefore, initial doses should be reduced and subsequent doses titrated to effect via a peripheral nerve stimulator (2).

    c. **Intravenous agents**: Safe and effective anesthesia using propofol and remifentanil 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+-K+ 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). However, the risks of residual muscle relaxation in this patient group are significant. The use of muscle relaxant without reversal was shown to be an independent risk factor for an adverse perioperative event (25, 26). Therefore, reversal should be considered when non-depolarizing muscle relaxants are used; Sugammadex has been used uneventfully to reverse muscular blockade in DM patients (26).

    b. **Extubation:** Due to 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 post-operatively for several days or more. If abdominal distention and/or severe post-operative pain are likely, these problems will exacerbate pre-operative weakness of cough and respiratory muscles. Maintaining post-operative ventilation can speed effective recovery and enhance pain management. Additionally, there is an increased risk of delayed-onset apnea and death after extubation during the immediate 24 hours after surgery, and even longer if post-operative anxiolytics, sedatives or opioid analgesics are administered. Close and continuous monitoring of cardiopulmonary function (SpO2 and ECG) is needed during this time period.

Post-operative management issues

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

1. **Pain control:** Consider the use of regional anesthesia, NSAIDS, and acetaminophen for postoperative pain control. If these medications/modalities are contraindicated, then the use of opioids must be administered with caution and vigilant monitoring. The exquisite sensitivity of DM patients to the respiratory depressant effects of opioids (systemic or neuraxial) has resulted in 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. Analgesia can be safely achieved in DM patients with epidural opioid administration (11), but even with a small dose of epidural morphine, respiratory depression can result (8). Furthermore, opioids exacerbate the gastrointestinal paresis intrinsic to DM, and increase the risk of reflux, aspiration, and additional ventilatory impairment from meteroism or pseudo-obstruction.

2. **Pulmonary considerations:** In a retrospective analysis of 219 DM1 patients who underwent surgery under general anesthesia, Matheiu et al. found that most perioperative complications were related to the pulmonary system (1). In particular, DM1 patients who were symptomatic, who were undergoing upper abdominal surgery, or who had severe muscular disability were especially at risk. In these patient groups, post-operative monitoring, aggressive chest physiotherapy, and incentive spirometry are recommended (1). Delayed-onset apnea is most likely to develop in the first 24 hours postoperatively, and exacerbation of baseline hypersomnia could become apparent. An ICU may be most appropriate for the detection and treatment of these complications.

Summary

- Perform an extensive preoperative evaluation with a multi-disciplinary medical team
- Use regional anesthesia when appropriate, either alone or to reduce general anesthesia
- Avoid or be cautious with pre-medications (anxiolytics, sedatives and opioids)
- Keep the patient warm
- Consider prophylactic application of defibrillator/pacer pads
- On induction, be prepared for the high likelihood of aspiration and other airway complications. Avoid succinylcholine
- Adhere to strict extubation criteria. Given the effects DM has on the pulmonary system, anticipate, and fully prepare the patient and family for the need to provide supportive mechanical ventilation until extubation criteria are unequivocally met, often after the patient is fully conscious
- If prolonged non-invasive ventilatory support is anticipated, for example following thoracic or abdominal surgery, start BiPAP support preoperatively and have the patient's BiPAP available for post-procedure use
- Plan for continuous SpO2 and ECG monitoring postoperatively until the patient fully regains pre-operative status
- Manage postoperative pain with NSAIDs, regional techniques, and acetaminophen when appropriate. Use opioids with extreme caution
- Encourage aggressive pulmonary toilet postoperatively, using a mechanical cough-assist device if necessary. Preoperative training with a cough-assist can facilitate its use in the postoperative period

Addendum 1:

List of authors and acknowledgements

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Richard Moxley, MD,

John W. Day, MD, PhD

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Addendum 2:

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The mission of the Myotonic Dystrophy Foundation is to enhance the quality of life of people living with myotonic dystrophy and advance research focused on treatments and a cure.

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663 Thirteenth Street, Suite 100, Oakland, California 94612
415.800.7777 | info@myotonic.org | www.myotonic.org