

Anesthesia Protocol for Myotonia Congenita

This is a protocol written up by the anesthesiologist of one of our forum members:

There are several concerns when anesthetizing patients with myotonia congenita. First off there appears to be an increased risk of developing malignant hyperthermia (MH) which is a very dangerous and often fatal anesthetic complication induced by depolarizing neuromuscular blockers like succinylcholine and by the inhalational anesthetics. The inherited predisposition to developing MH is heterogeneous. There is a defect in Ca^{++} influx into unrestrained muscle contraction. MH is linked to a defect in the muscle sarcolemma ryanodine receptor. This of course is not the same defect as that seen in myotonia congenita. It appears that many causes of intraoperative muscle rigidity in patients with myotonia are misdiagnosed as MH. It is a little unclear if the risk for MH is increased but for now it is best to assume that it is increased and to have the anesthesiologist deliver what is called a non-triggering anesthetic. This can be easily done.

Patients with myotonia are at increased risk of developing masseter muscle spasm of the jaw and this can lead to inability of the anesthesiologist to manage the patient's airway and will often lead to cancellation of the case before it gets started. Masseter muscle spasm is often associated with MH but is also seen in myotonia leading to some confusion in making the proper diagnosis in this emergency situation. Masseter spasm is seen most commonly after succinylcholine administration.

Besides the risk of MH. Patients with both types of Myotonia should never be given the depolarizing muscle relaxants like Succinylcholine. This causes sustained muscle contraction instead of the expected relaxation and can lead to the inability to intubate or ventilate the patient, which could be fatal. Also patients with myotonia who are given Succinylcholine may have a massive release of potassium leading to cardiac arrest. Prolonged surgeries leading to muscle compression or the administration of Succinylcholine may lead to post-operative muscle breakdown in the form of rhabdomyolysis which can also cause acute renal failure and death.

Patients with myotonia also run the risk of having prolonged weakness after surgery which may increase their need for prolonged ventilation and endotracheal intubation. This may be caused by the neuromuscular blocking agents or by the inhalational anesthetics.

Patients with myotonia are also at increased risk of pulmonary aspiration and postop pneumonia.

Regional anesthetic techniques like spinal or epidural anesthesia, should be safe in patients with myotonia. Local anesthetics can also be used.

In summary, before your child has anesthesia, you should talk to the anesthesiologist and explain his disorder and make a plan for a non-triggering anesthetic which will avoid most of the problems listed above and can be readily delivered.

Preparation of the operating room anesthesia machine and preparation of the anesthesiologist are very important in anesthetizing these patients.

This is the protocol that I have written up based on communication with several anesthesiologists who are experts in surgery for patients with myotonia congenita. It was reviewed by the head of obstetric anesthesia for Brigham and Women's Hospital.

Check pseudocholinesterase levels before surgery to determine risk from anesthetics and depolarizing agents - if low use extreme caution

Make sure filter medium and tubes have not been contaminated with inhalation anesthetics; evacuate operating room air thoroughly before patient enters

Warm IV fluids and operating table if possible. Keep warm blankets on patient during the surgery to avoid triggering myotonia from chilling.

Anesthetics/Analgesics: Propofol, Fentanyl and Versed

Paralyzing Agent: Vecuronium (NO succinylcholine)

Local - Bupivacaine, no vasoconstrictors (no Lidocaine); epinephrine will trigger myotonia

IV saline only - no potassium added. Glucose/dextrose slow drip to avoid insulin response

Monitor potassium levels throughout surgery - cardiac arrest from hyperkalemia is a bigger risk than malignant hyperthermia

This is an article from MDA's Quest magazine on anesthesia. Myotonia is listed on the second page:
Coping with Anesthesia, <http://static.mda.org/publications/Quest/q73anesthesia.html>