Anesthetic Considerations

Myotonic dystrophy presents many concerns to the pediatric anesthesiologist. Myotonia, defined as persistent contraction with delayed relaxation of muscles, is the hallmark of the disease. Patients usually present with hypotonia at birth, then have increased tone and strength and eventually progress to muscle wasting. Avoidance of medications that may stimulate contractions as well as prevention of shivering is imperative.

Cardiovascular symptoms are a major concern with conduction abnormalities present in 90% of patients. Heart block and delayed conduction is common and sudden death has been associated with development of third degree block. Cardiomyopathy with fibrosis may occur and there is an increased risk of mitral valve prolapse. Aspiration risk is high with myotonic dystrophy patients due to bulbar weakness (CN IX, X, XII) along with impaired cough from involvement of the diaphragm and intercostal muscles. They often have dysphagia, reduced peristalsis and may need to be fed via gastric tube. Respiratory depression is due to weakness of the diaphragm and intercostal muscles along with increased sensitivity to many anesthetic medications, opioids in particular. Patients may have hypventilation from hypotonia as well as from central or obstructive sleep apnea.

Our case is of a 14 year old, 62 kg male with type 1 myotonic dystrophy and developmental delay, presenting for cardiac catheterization, EP study and atrial flutter ablation after a prior cardiac arrest upon induction of general anesthesia. The patient was non-verbal, but mother indicated that he had symptoms of gastroesophageal reflux.

In the prior case, he had presented for TEE and cardioversion after a history of atrial flutter for 3 months. His baseline heart rate was 130-140 BPM despite initiation of atenolol. Symptoms of fatigue persisted despite increased beta blocker dose. After sevoflurane inhalation induction and IV placement, he was given succinylcholine and propofol, and was successfully intubated. However, hemodynamic decompensation, bradycardia and arrest occurred. Chest compressions and epinephrine were insufficient for resuscitation and ECMO support was required.

After six days, ECMO was discontinued. Two weeks later, LV function and ejection fraction normalized, and respiratory function was adequate on room air. He was then scheduled for cardiac catheterization, EP study, and ablation.

With knowledge of his prior decompensation, we chose to use monitors in addition to standard ASA monitors to focus on his cardiac function. These included: cerebral oximetry, limited transthoracic echocardiography (TTE) before and during induction, and continuous palpation of pulse. A previously inserted double lumen PICC line provided opportunity for CVP monitoring and inotropic support (norepinephrine at 0.02-0.04 mcg/kg/min) throughout the case. We chose an intravenous induction with 1.5 mg of etomidate, midazolam and 50 mg ketamine. Limited TTE was repeated at each step to assure maintained heart function. Cisatracurium was then administered and the patient was intubated. Post intubation TTE showed that contractility was maintained. Anesthesia was maintained using isoflurane, nitrous oxide and remifentanil infusion. After completion of the ablation, he was extubated and taken to the pediatric intensive care unit for postoperative monitoring. He had an uneventful postoperative course and was discharged two days later.

Avoid:
- Depolarizing muscle relaxants, anticholinesterase inhibitors
- Increased sensitivity to anesthetic medications and risk of respiratory depression

Use:
- Propofol, volatile anesthetics
- Direct infiltration of muscle with LA can attenuate contractions

Consider:
- Awareness and avoidance of drug interactions as patients may be on phenytoin, procainamide, quinines to attenuate contractions
- Prevent shivering or manipulations that may cause contractions