Anesthetic Management of an Infant with Juvenile Myasthenia Gravis
Undergoing a Transsternal Thymectomy

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Introduction
- Myasthenia gravis is an autoimmune disorder where antibodies target the postsynaptic acetylcholine receptors at the motor end-plate.
- Juvenile MG, which occurs before the age of 19 years, is uncommon, accounting for less than 10% of all cases.
- Prepubertal onset is even rarer, and distinctions include:
  - Equal prevalence in males and females
  - Higher prevalence of ocular symptoms
  - Increased incidence of seronegativity
  - Commonly absent thymoma
- Long-term prognosis tends to be better if diagnosis is made in the first five years of life.

Case Background
- 22 month-old 10 kg boy initially diagnosed with ocular myasthenia gravis
- Started on pyridostigmine with some symptomatic improvement
- One month later, presented with acute dysphagia
- Started on prednisone & IVIG for myasthenic crisis
- Scheduled for transsternal thymectomy

Management
- On the day of surgery, physical exam revealed bilateral ptosis and ophthalmoplegia
- No recent dysphagia or respiratory issues
- Peripheral IV access was obtained following a mask induction with sevoflurane
- A train-of-four nerve stimulator was attached before titrating a total of 1.5 mg of vecuronium in 0.5 mg increments
- Easy intubation with direct laryngoscopy
- Fentanyl 25 mcg given before surgical incision
- Transternal thymectomy was performed
- No additional muscle relaxant or opioid given
- Total surgery time was 43 minutes
- Train-of-four showed 4/4 twitches
- Muscle paralysis was reversed and the patient was extubated after fulfilling criteria for extubation
- An additional 5 mcg of fentanyl was given five minutes after extubation before transport to the intensive care unit
- ICU course uneventful
- Patient discharged home the following day
- One-month follow-up appointment with Neurology: near-complete resolution of ocular symptoms and no reports of dysphagia or respiratory issues at home.

Discussion
- Commonly used anesthetic agents have the potential to cause generalized weakness that may require postoperative ventilation in patients with myasthenia gravis.
  These patients are resistant to succinylcholine; anti-acetylcholine receptor antibodies prevent its binding to acetylcholine receptors. Conversely, these patients demonstrate an increased sensitivity to nondepolarizing muscle relaxants because of the reduced number of available acetylcholine receptors. The response to nondepolarizing muscle relaxants becomes less predictable in patients receiving pyridostigmine therapy, which can make the patient more resistant to neuromuscular blockade than expected. When administering a nondepolarizing muscle relaxant, it should be given in small incremental doses and guided by a train-of-four nerve stimulator.
  Underdosing the anticholinesterase reversal agent may result in residual blockade. However, overdosing may lead to a cholinergic crisis, which is also characterized by weakness.
  Therapeutic doses of opioids for analgesia do not affect neuromuscular transmission in myasthenia, but may contribute to central respiratory depression. For this reason, Fentanyl, a short-acting titratable opioid is a good option.

Summary
- The use of sevoflurane and vecuronium provided ideal intubating and surgical conditions for a patient with juvenile myasthenia gravis undergoing a transsternal thymectomy
- Careful dosing, guided by neuromuscular monitoring allowed us to use muscle relaxant and reversal agent, and safely extubate the patient in the OR.

References
- K Keragia. Anesthesia for the Patient with Myasthenia Gravis. In: UpToDate, Post TW (Ed), UpToDate, Waltham, MA.