[NM-197] Freeman-Sheldon Syndrome and General Anesthesia for Mandibular Osteotomy

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Introduction

Freeman-Sheldon syndrome (FSS) is a rare multiple congenital contracture syndrome with a non-progressive or slowly progressive myopathy affecting the facial, limb, and respiratory muscles. Less than 100 cases have been reported to date. Its pathognomic sign is the “whistling face” due to microstomia and contractures of the facial muscles. Other features include down-slanting palpebral fissures, mandibular hypoplasia, and H- or Y-shaped creases, and contractures of the limbs resulting in club feet, camptodactyly of the hands, and severe scoliosis. Anesthetic concerns include difficult airway, difficult IV placement, risk of post-operative pulmonary complications, and reported increased risk of malignant hyperthermia (MH).

Case Description

BL is a 3 week old, 3.3 kg female with Freeman-Sheldon syndrome presenting for direct laryngoscopy, bronchoscopy, and bilateral mandibular osteotomy. Severe OSA was diagnosed by polysomnography with an AHI of 43.7. The patient was sedated for laryngoscopy with propofol and dexmedetomidine. The ENT surgeon made multiple attempts at laryngoscopy with a Hollinger laryngoscope and flexible and rigid endoscopes, noting an anterior glottic opening and an epiglottis that was difficult to lift. She was intubated on a blind attempt with no visualization of the glottic opening. She was easy to mask ventilate throughout and oxygen saturation remained above 95%. Once intubated with a 3.0 cuffed ETT, she was maintained remifentanil and propofol infusions. She remained intubated electively due to the difficult intubation, severe OSA, and ongoing mandibular distraction.

Discussion

Difficult intubation is a known risk in patients with FSS due to a very small mouth opening, mandibular hypoplasia, limited neck mobility, and muscle contractures. BL was difficult to intubate for all of these reasons. She was intubated once again at 2 months with difficulty by ENT with a Parsons laryngoscope, once with difficulty at 5 months with a grade 3 view by direct laryngoscopy, and then easily at 6 months with a grade 2 view by direct laryngoscopy.

Patients with FSS may be at increased risk of MH, but this has not been proven. In one series, 3 of 19 patients who had general anesthesia developed MH and two more developed unexplained hyperpyrexia (4). There have been two reports of masseter muscle spasm after halothane and succinylcholine, one relieved by dantrolene, the other by termination of halothane (1, 3). There is one case report of muscle rigidity and elevated CK after halothane and succinylcholine (1). It is thought that FSS may be an unconventional myopathy, which would explain a predisposition to MH. Though the risk of MH is unclear, it seems prudent to use a non-triggering anesthetic in these patients. It should be noted that this patient underwent three more anesthetics at our institution, one non-triggering and two with sevoflurane with no evidence of MH.

References

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