Appropriateness of Outpatient Anesthesia for a Child with Freeman-Sheldon Syndrome

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Introduction

Freeman Sheldon syndrome (FSS) is a rare multi-system congenital myopathy that presents a unique challenge to the anesthesiologist due to the risk of difficult intubation and intravenous access. The disorder is characterized by significant craniofacial abnormalities, contractures of the distal limbs, and thickened subcutaneous tissue. We review anesthetic management for the application of a Mehta cast to treat congenital scoliosis in a two-year old, ten kilogram female with FSS.

Case Report

The patient initially presented to an ambulatory surgery center for the application of a Mehta cast, however based on the patient's small mouth opening and anticipated airway difficulty, the case was postponed and rescheduled at the inpatient hospital. The patient was brought to the operating room and standard ASA monitors placed. Inhalational induction commenced with sevoflurane with nitrous oxide and oxygen. Mask ventilation was achieved easily with an oral airway. IV access was obtained in the right forearm with a 24 gauge catheter. Endotracheal anesthesia was required secondary to increased abdominal and thoracic pressures during casting. No muscle relaxant was given. Three attempts were made at securing the patient's airway. The first two with direct laryngoscopy utilizing a miller 1 and macintosh 2 blades, revealed a Cormack grade 4 view. The GlideScope® 2 blade was used on the third attempt revealing a Cormack grade 3 view. A styleted 4.0 oral endotracheal tube was then successfully passed through the vocal cords and anesthesia maintained using inhaled sevoflurane. The patient tolerated the procedure and anesthetic without complications. She was extubated in a deep plane of anesthesia and taken to PACU where after four hours of uneventful observation, she was discharged home

Discussion

Cranio-Carpo-Tarsal dystrophy was first described in 1938 as a case series by EA Freeman and JH Sheldon and then became referred to as "Freeman Sheldon syndrome." Despite typically being characterized as sporadic inheritance of the syndrome, there are reports in the literature of familial cases that have proven both autosomal recessive and dominant inheritance. Although no epidemiologic data exists for FSS, there have been less than 100 case reports and even fewer in the anesthetic literature. Patients have a characteristic "whistling face" secondary to microstomia, micrognathia, and pursed lips. These features lead to significant difficulty with intubation and potentially failed intubation. Distal contractures of the limbs along with fibrosis of the subcutaneous tissue lead to challenges obtaining IV access. There have been several case reports indicating the potential for increased susceptibility to malignant hyperthermia in FSS secondary to the congenital myopathy, however this association has not been proven. MHAUS no longer includes FSS as MH susceptible. Inhalation induction was chosen in our case due to anticipated difficulty obtaining IV access and patient comfort. Dantrolene was available in the operating room if necessary but was not utilized. Based upon these characteristics the patient's initial surgery was appropriately delayed at the ambulatory center because of the anticipated difficulty securing the airway. The case later occurred at the inpatient hospital where advanced airway supplies such as pediatric fiberoptic bronchoscopes and a pediatric ENT surgeon were readily available. Based upon the initial successful and uneventful anesthetic management, the patient was subsequently scheduled at the outpatient surgery center for her repeat castings and has had multiple positive outcomes. Despite the potential for difficult airway and IV access, FSS is a complex disorder that can be managed on an outpatient basis if adequate measures are taken in preparation for the anesthetic case.

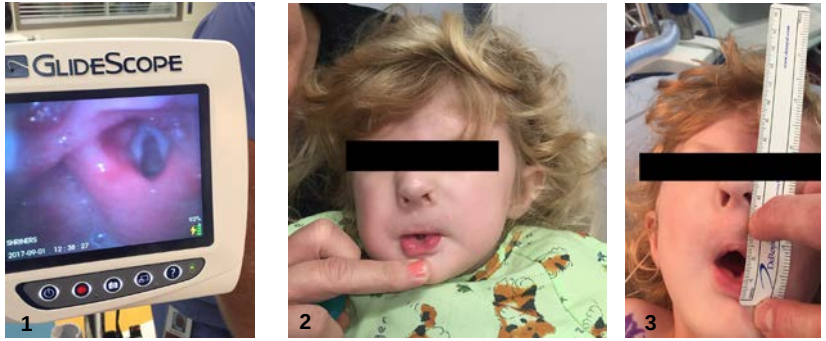

Figure 1: Glidescope visualization of vocal cords on subsequent anesthetics, revealing an improved view; 2: physical exam features in awake patient with the characteristic whistling face and microstomia. 3: physical exam features of asleep patient with approximately 1cm mouth opening.

References

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