

## [NM-211] 3 Staged Spinal Fusion in 15 Year Old Girl with Freeman- Sheldon Syndrome: To Trach or Not to Trach?

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### Introduction:

Freeman-Sheldon Syndrome (FSS) is a rare congenital myopathy and dysplasia characterized by difficult intubation. Patients have a characteristic “whistling face” as well as microstomia and micrognathia. Often they have limb deformities which may result in flexion contractures. We report here a case where a teenage girl with FSS presented to our hospital for a 3-staged scoliosis repair.

### Case Presentation:

A 15 year- old Spanish speaking girl with Freeman-Sheldon Syndrome presented to our hospital for a 3 staged scoliosis repair. The patient had been previously cared for at an outside hospital with six serial back braces and was referred to our institution for her staged scoliosis repair. The patient had 100 degree levo-scoliosis and 116 degree kyphosis with gross chest deformities. Her pulmonary function tests demonstrated greater than 30% predicted FEV1. Her airway exam revealed Mallampati Class 1, 1.5 cm mouth opening, 20 degree neck extension, 30 degree neck flexion, and thyromental distance of 1 cm. Pre-operative planning between the ICU team, spine surgeon, otolaryngology, and anesthesiology took place to discuss the option of placing a tracheostomy for her surgeries, given the potential for difficult airway, poor pulmonary reserve, and prolonged hospital stay secondary to the 3 staged surgical approach. After much thought, we chose to proceed with fiberoptic intubation, with otolaryngology on standby for potential emergency tracheostomy. On morning of 1st surgery, halo placement, the anesthesia team proceeded with an asleep inhalational induction, easy bag mask ventilation, and successfully intubated the patient with cricoid pressure and glidescope, grade 2 view. The patient was in halo traction of 17 pounds for 2 weeks. When she returned for her 2nd surgery, anterior release, IV induction was followed by inability to bag-mask and numerous attempts by multiple anesthesiologists with glidescope and fiberoptic were extremely difficult. The patient was eventually intubated via oral fiberoptic technique. She did not tolerate ventilation, and required vasopressors for hypotension. The case was aborted and the patient was transferred intubated to the PICU. She was diagnosed with post-obstructive pulmonary edema and recovered in the PICU for several days. At this point, the team decided to bring patient back for elective tracheostomy, prior to her next surgery. After the tracheostomy stoma was allowed to mature, the patient returned for her anterior release procedure, which proceeded smoothly. She is scheduled to return to OR for posterior spinal fusion this week.

### Discussion:

Patients with known FSS and difficult airway coupled with poor respiratory function may be good candidates for elective tracheostomy prior to a multiple staged procedure. Although tracheostomy has inherent associated morbidity and mortality, this must be weighed against morbidity and mortality in the patient with competing challenges.

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