[NM-221] Difficult Airway in a Patient with Nager Syndrome

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Introduction
Nager acrofacial dysostosis is a rare disorder seen in a small subset of the population. It is associated mainly with craniofacial and limb abnormalities. The associated symptoms often include hearing loss, limb abnormalities and dysmorphic cranial features as well as external and middle ear abnormalities, malar hypoplasia, and radial or ulnar abnormalities. They often have speech delays and feeding challenges given their inherent features.
While many of these physical anomalies associated with nager syndrome have a strong impact on a patient's quality of life, there are particular concerns with regards to their airway and anesthetic management given their craniofacial features. Often, patients with Nager syndrome have respiratory compromise from birth requiring intervention. They usually have a hypoplastic mandible and it is often associated with a cleft lip or palate.
Case Description
A 7 year-old boy with Nager syndrome presented for mandibular distraction. The patient's past medical history was significant for respiratory failure and conductive hearing loss. He had a tracheostomy performed at day #5 of life. Our case is unique in that we had the opportunity to contrast his known difficult airway both for the mandibular distraction and 3 months later when he returned to our operating room.
Physical exam revealed downslanting appearance of his eyes and eyelids. He had normal external ears with a hearing aid. He had severe mandibular hypoplasia and a tracheostomy scar. He also had very limited mouth opening with a cleft palate.
Given his unique airway situation, two attending anesthesiologists, as well as a surgical team with a tracheostomy set were present upon induction of anesthesia. Anesthesia was induced using inhaled sevoflurane. During induction, the patient became progressively more difficult to ventilate. Several diffierent airway adjuncts were attempted without success. An Air-Q device was inserted that allowed us to ventilate the patient. Ultimately, this supraglottic device was used as a conduit for fiberoptic intubation with a #5.0 endotracheal tube. The surgery was uneventful. The patient remained intubated and was transported to the pediatric ICU. He returned on POD #2 for extubation in the operating room with the surgical team on standby.
Three months after the initial procedure the patient returned for hardware removal of the bilateral distractors. Again, a surgical team was present in the OR with a tracheostomy set. Anesthesia was induced and the patient was successfully intubated using a video laryngoscope. Surgery was uneventful and the patient was extubated at the end of the case.
Discussion
Although patients with known difficult airways present challenges, our case was particularly interesting as we had the opportunity to compare and contrast the same patient before and after his mandibular manipulation. The potential for complications and adverse events is higher than that of a child with a normal airway. At all times backup plans must be made in the event of difficulty with the airway. A plan was in place for a surgical airway, but was not necessary. With the right personnel and equipment, we were able to proceed with this difficult case in a safe and controlled manner.
