Awake Intubation in an 8-Year-Old Female with McCune-Albright Syndrome

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Abstract

Abstract: An 8-year-old with McCune-Albright Syndrome presented for resection of a very large fibrous dysplasia mass of the face with significant distortion of the airway anatomy. She had significant obstructive sleep apnea with daytime somnolence and oxygen desaturations while on room air preoperatively. We were able to successfully manage her airway by providing intravenous sedation, topical anesthesia of the airway and oral fiberoptic intubation in close collaboration with our Otorhinolaryngology colleagues.

Patient History

Past Medical History:
- 50.5 kg 8 year-old female with McCune-Albright Syndrome
- Large recurrent fibrous dysplasia with airway compression and severe OSA

Airway History and Exam:
- Somnolent, SaO2 85% preop (on room air)
- Dyspnea with stridor when supine, very large tonsils
- Anticipated difficult to impossible mask ventilation

Airway Management

- Awake IV start and glycopyrrolate IV premedication
- Otorhinolaryngology at bedside with tracheostomy kit in the room
- Dexmedetomidine infusion (2 mcg/kg/hr), Ketamine boluses 10-20 mg prn
- Topical anesthesia: 2 sprays 14% Benzocaine, 5 ml aerosolized 2% lidocaine
- Fiberoptic bronchoscope loaded with 5.0 cuffed endotracheal tube
- Four failed attempts to view the glottis (see figure 4)
- Topical anesthesia supplemented: 4 ml aerosolized 4% lidocaine
- Four more attempts before successful passage of scope into the trachea
- Endotracheal intubation confirmed, then general anesthesia induced

Discussion

- Anticipated impossible mask ventilation if sedation excessive, particularly if supine
- In order to limit sedation and increase comfort we sought to use the maximum safe dose of topical anesthetic\(^1\,\,2\,\,3\,\,4\)
- Incomplete data on maximum topical doses
- Greater peak serum levels if child <2 yo
- Dexmedetomidine was a valuable component of our sedation regimen (very little respiratory depression and lack of sialogogue effects)

Conclusions

- Preparation and a multidisciplinary approach to the difficult pediatric airway increases safety
- Maximum topical anesthetic dosing is likely equivalent to the maximum intravenous dosing recommendations\(^1\,\,2\,\,3\,\,4\)

References


Consent: Written informed consent was obtained from the patient’s Mother for use of the patient’s medical records, including images, for academic and educational purposes.