Management of Obstructive Sleep Apnea in a Developmentally Delayed Pediatric Patient with Aggressive Behavior and Pierre Robin Sequence

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This is a case of a 15-year-old nonverbal, developmentally delayed boy with history of Fragile X syndrome (FXS), autistic spectrum disorder, Pierre Robin sequence (PRS), subglottic stenosis, and ongoing obstructive sleep apnea (OSA), who required aggressive surgical intervention. OSA related symptoms included loud snoring, apneas, awakenings with gasping breaths, enuresis, and daytime sleepiness. The patient also had a history of behavioral problems including aggressive behavior, which led to his family and his health care providers experiencing challenges caring for him. For example, the patient's behavior made examining him very difficult and prevented ENT exam. Likewise, OSA was difficult to manage due to the patient's inability to cooperate with treatment. The OSA treatment course for this patient is briefly described below.

At age 2 months, the patient began experiencing symptoms of OSA, and tracheostomy was performed. The patient self-decannulated at age 6 years, then subsequently was able to function without a tracheostomy until age 8 years. A repeat tracheostomy was performed at age 8 years, but the family had to struggle with the patient to change his tracheostomy tube, often spending up to 45 minutes each time a tube change was needed. At age 10 years, polysomnography (PSG) was performed in our lab with the tracheostomy capped with a

Passy-Muir valve. This showed mild OSA, including apnea hypopnea index (AHI) of 5.4 and oxygen desaturation nadir of 91%, and at the parents' request, the patient was decannulated. Adenoidectomy and inferior turbinate reduction were performed at the same time to minimize upper airway flow limitation and maximize likelihood of successful decannulation. Tonsils were graded 1+ and felt to be noncontributory to his OSA. Shortly after these surgeries, the patient was noted to have increased sleepiness and worsened aggressive behavior. A repeat diagnostic PSG in our lab two months after decannulation showed severe sleep apnea. At that time, the AHI was 22/h and oxygen nadir was 76%.

QUESTIONS

- 1: What are the treatment options for this pediatric patient with cognitive impairment, aggressive behavior, and severe obstructive sleep apnea?
- 2: If mandibular maxillary advancement were to be performed, what perioperative limitations have to be considered to ensure optimal postoperative outcome in this patient with aggressive behavior and previous tracheostomy?

ANSWERS

- 1: Treatment considerations include a trial of continuous positive airway pressure or oxygen, repeat tracheotomy, or other surgeries, such as mandibular maxillary advancement, distraction osteogenesis or rapid mandibular expansion. Tonsillectomy is an option, but this patient did not have tonsillar hypertrophy.
- 2: The patient's inability to follow dietary restrictions, significant injury from lip biting due to lower lip and chin paresthesia, and anticipated need for prolonged ventilator support have to be considered.

DISCUSSION

The patient in this case was predisposed to OSA due to craniofacial abnormalities associated with PRS, including mandibular hypoplasia and glossoptosis (posterior placement of the tongue).^{1,2} Additionally, this patient had a narrowed subglottic space. Features of FXS (a long and narrow face) may have been contributory as well. Furthermore, FXS related cognitive impairments including autism, impairment in communication, and impairment in social interaction led to the patient being unable to cooperate with previously tried treatment as described above.³

Upon discussing further treatment options with the patient's family, they felt he would not tolerate a trial of continuous positive airway pressure (CPAP) or oxygen, due to his inability to tolerate any type of mask or nasal cannula on his face. Likewise, since the patient could not tolerate an oral device, both distraction osteogenesis and rapid mandibular expansion were not attempted. Repeat tracheostomy was decided against given the patient's risk for self-decannulation, not to mention the risk for introducing foreign objects into the tracheostomy tube.

Our oral and maxillofacial surgeons, who have experience treating children with special needs, were consulted for mandibular maxillary advancement (MMA). Since aggressive behavior prevented physical exam, the patient was taken to the operating room (OR) for examination and dental impressions required for the MMA.

Traditionally, MMA relies on a Le Fort 1 and bilateral sagittal split osteotomy (BSSO) that is stabilized with miniplates and microscrews. BSSO is performed via an intraoral approach. Regarding the geometry of the osteotomy, the osteotomy is placed in the sagittal plane. The medial and lateral mandibular cortices are in contact, allowing for smaller (and cheaper) fixation. Postoperatively, esthetics and lip motor function are usually prioritized over lip sensation and diet requirements. Temporary anesthesia in lip and chin after BSSO results from injury of the inferior alveolar nerve and occurs in approximately 12% of patients.⁴ The facial nerve is protected by several soft tissue planes from operative site with the traditional BSSO approach. Also, patients are required to maintain a soft non-chew diet for 6 weeks to prevent hardware failure.

However, it was anticipated that this patient would be unable to comply with the no-chewing diet requirements and that lower lip and chin paresthesia could carry significant morbidity from lip biting. Therefore, instead of standard BSSO, we

Figure 1

Top row: Clinical model photos of mandibular osteotomies and hardware. The photo on the left shows a more commonly used bilateral sagittal split osteotomy of the mandible with traditional set of microscrews and a microplate placed through an intraoral incision. The right photo shows a C osteotomy with a reconstruction plate typically used in trauma and reconstruction. Reconstruction plates are typically placed through a neck incision. The heavier reconstruction hardware was used for this patient as it is able to bear all of the force of the mandible. The smaller plates and screws can break when carrying full loads so one must maintain a soft non-chew diet for 6 weeks after surgery. **Bottom left:** Presurgical photograph of patient showing retrognathic position of the mandible. **Bottom right:** Computer simulated image of anticipated postoperative facial changes.

performed a mandibular advancement with a C osteotomy fixated with reconstruction hardware (**Figure 1**).⁴ The C osteotomy, which was first described in 1968, is rarely performed primarily due to scarring on the neck, risk of marginal mandibular branch of CN VII injury, and the need for larger (and more expensive) hardware.

The geometry of the osteotomy is such that there is little to no contact of the proximal and distal segments of the mandible. Therefore, bone grafting must be done, which requires a second surgical site (anterior iliac crest being most common), with associated morbidity. The osteotomies were performed with computer-fabricated guides derived from virtual planning (Synthes ProPlan, Plymouth, MI) with the best guarantee of preservation of the inferior alveolar nerve. Additionally, the patient was left in maxillomandibular fixation (jaws wired shut) for one week for additional stabilization, then allowed to eat a regular diet 10 days after the procedure.⁴ Repeat tracheostomy and aggressive tracheostomy care were performed to prevent major respiratory complications. He was kept on sedative and paralytic medication until postoperative day (POD) 10 due

to his history of uncooperative behavior and self-decannulation. The patient was weaned from ventilator support on POD 12, and decannulated on POD 17. The perioperative course was otherwise unremarkable.

Four months postoperatively, PSG showed improvement of this patient's OSA and sleep, including AHI 5.6 and oxygen saturation nadir of 92%.

In summary, this patient was not a candidate for noninvasive therapies (CPAP or oxygen) for his severe OSA, and his history of self-decannulation and resistance to having tracheostomy tube changes made the risks of repeat tracheostomy outweigh potential benefit. MMA was successfully performed by taking special measures that included utilizing stronger than normal hardware, temporary maxillomandibular fixation while paralyzed, sedation while the patient was on ventilatory support, and aggressive tracheostomy management.

PEARLS

1. With special perisurgical preparation, MMA is a treatment option in pediatric patients with OSA, cognitive impairment, and aggressive behavior. The surgery should be performed by a team with experience in treating children with special needs.
2. Special considerations must be kept in mind prior to MMA in those with cognitive impairment and aggressive behavior. Sedation may be necessary to examine the patient. Prolonged hospitalization, postoperative maxillomandibular fixation, and sedation with ventilator support may be necessary.
3. In developmentally delayed patients with aggressive behavior, improvement of obstructive sleep apnea may

improve behavior, and as a result, quality of life for the patient and his or her family.

CITATION

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