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OTO OBSERVER

MANDIBULAR DISTRACTION OSTEOGENESIS IN NEONATE WITH PIERRE ROBIN SEQUENCE (PRS)

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The patient presented as a 2 month old African American female with micrognathia transferred from outside hospital (OSH) for failure to thrive (FTT) and inability to wean oxygen therapy. She was born full term at OSH and struggled in the neonatal period to take PO so g-tube was placed. Additionally, she was managed with positive pressure ventilation, nasal trumpet and oxygen therapy. She was discharged home at approximately 4 weeks of age and re-presented to an OSH with FTT and respiratory distress. Initially her FTT was thought to be due to socioeconomic factors.

As her evaluation continued a community otolaryngologist was consulted and identified her retrognathia as a source of her symptoms and recommended transfer to the Children's Hospital of Georgia for mandibular distraction osteogenesis (MDO). On arrival, we were met with a very involved and knowledge mother at bedside. She explained in remarkably plain language a complex medical situation: "[She] can't gain weight because she can't breathe."

The patient indeed displayed the hallmark retrognathia, glossoptosis and airway obstruction of PRS and continued to require oxygen support and prone positioning with increase WOB and observed apneas. After reviewing a CT of her facial bones, we confirmed that she was a good candidate for MDO. The presurgical process also involved 3D planning with 3DSystems and Stryker to plan osteotomy sites, appropriate hardware selection and placement. With this technology we were able to obtain 3D printed cutting guide for use during surgery and mandibular models depicting the anticipated 17mm of distraction.

Figure 1: Before surgery (left); 12 days after surgery (right).

Post-operatively, she was admitted to the ICU and remained intubated as planned for 3 days while the initial activation phase began, once there had been a few millimeters of distraction, she was extubated to nasal trumpet and weaned over the course of the next few days as the distraction continued. A total of 12 days of distraction prior to post auricular post removal. The bone was then allowed to consolidate over 10 weeks before hardware removal. She was discharged just 2 weeks after her initial surgery taking all of her nutrition by mouth, on room air without respiratory support and with a very happy mama!

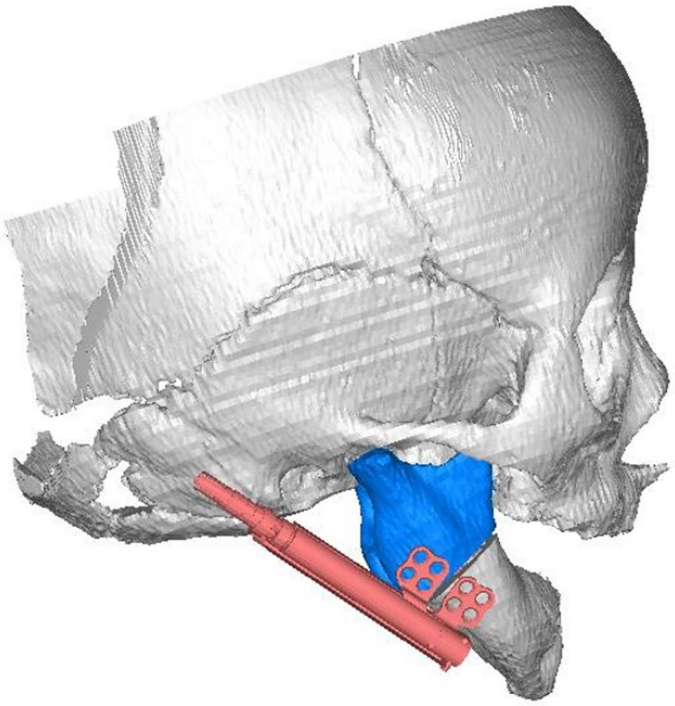

Figure 2: 3D model of mandibular distraction hardware and procedure.
Images above courtesy of 3DSystems
Mandibular Distraction Hardware by Stryker

Discussion

Patients with Pierre Robin Sequence (PRS) have a triad of micro/retrognathia, glossoptosis, and airway obstruction w/ wo cleft palate. Some diagnosis are able to be made in the prenatal period. For infants who have difficulty taking PO, and requiring respiratory support, interventions are necessary. Historically tracheostomy would be performed for airway management and is still required for the most extreme cases. Tongue-lip adhesions have also been used historically however these patients tend to have a higher rate of need for enteral feeding. The use of mandibular distraction osteogenesis (MDO) in many of these patients can reduce the need for nasogastric/gastric tubes and tracheostomy (1). MDO is performed with planned osteotomies application of distraction hardware, an initially activation phase with slow distraction of the osteotomies allowing for new bone growth in the intervening space. This can occur at a rate of 1-1.5 millimeters per day(2).

Conclusion

Mandibular Distraction Osteogenesis is a great option for some neonatal patients with PRS who are experiencing FTT, respiratory failure/distress and oral feeding intolerance in order to avoid tracheostomy and decrease need for enteral feeding.

References:

1. K El Ghoul, C E Calabrese, M J Koudstaal, C M Resnick. 2019. " A Comparison of Airway Interventions and Gastrostomy Tube Placement in Infants with Robin Sequence" *Int, J. Oral Maxillofac. Surg.* 10. 013.
2. J D Sidman, D Sampson, B Templeton. 2009. " Distraction Osteogenesis of the Mandible for Airway Obstruction in Children" *The Laryngoscope*. Vol. 111, Issue 7 p. 1137-1146