Ankyloglossum Superius Syndrome Compromising A Neonatal Airway

Objectives

- Review the current literature on Ankyloglossum Superius Syndrome
- Describe our experience with this syndrome
- Describe the airway implications along with the management of this uncommon congenital anomaly

Study Design and Methods

This is a case report with review of relevant literature.

Introduction and Background

Ankyloglossum Superius Syndrome is a rare entity with few published reports. Our experience begins with a 37-week old newborn born to a mother with limited prenatal care, who presented on day 1 of life with a congenital fusion of the tongue to the hard palate. Physical exam along with fiberoptic endoscopy showed fusion of the floor of the mouth to the anterior hard palate, displacing the tongue into the nasal vestibule. This anomaly likely prevent fusion of the hard palate, resulting in a cleft palate. Furthermore, this malformation resulted in a large anterior oral band, obscuring any view to the posterior oral pharynx. The child was exposed to cocaine in utero by history. Other birth abnormalities included microretrognathia, smooth philtrum, and an ASD and VSD.

The child’s airway was secured through nasotracheal intubation, as the oral airway was significantly limited and the child would not be easily intubated if there was any emergent need. The child was then brought to the operating room for lysis of the previous adhesion. Genetic testing revealed no known genetic variant. The child did well post-operatively, and was subsequently discharged home.

There are few reports of Ankyloglossum Superius in the literature, with no consistent presentation nor commonly identified causation. Seventy ranges from small adhesions to full adhesion and displacement into the nasal cavity, with obstruction of the anterior oral inlet, as was our case. Reports have associated this syndrome as a part of aglossy-adactyly syndrome, Hanhart syndrome, oro-facio-digital syndrome, jejunoo-ileal atresia, or as an isolated entity. There was no reported association with cleft lip or palate. Most children go on to have normal oral function after lysis of the adhesion.

Discussion and Conclusions

Ankyloglossum Superius Syndrome is a rare entity of unknown causes. It has been sporadically described in the literature; however, this congenital defect carries with it the potential for devastating airway obstruction and complications. Here we provide a stepwise approach for evaluating, securing an airway, and treating the defect. Ultimately, nasotracheal intubation along with lysis of the tongue’s fusion to the hard palate provides for a controlled approach to securing and providing an unobstructed oral airway in this unusual congenital anomaly.

References