



Introduction

- Palatoglossal adhesion, also known as ankyloglossia superior, is a rare congenital anomaly
- Superior Ankyloglossia Syndrome is a cluster of congenital malformations including ankyloglossia superior, cleft palate, micro- or retrognathia and limb anomalies
- Superior Ankyloglossia Syndrome presents a unique challenge for airway management
- We present one of fewer than 30 reported cases of Superior Ankyloglossia Syndrome and only the second case in a neonate with extreme prematurity

Case

The patient is an infant born at 25 weeks’ gestation via emergent cesarean section for placental abruption who required positive pressure ventilation for respiratory distress immediately after birth.

Upon placement of a gavage tube for gastric decompression, he was noted to have an anterior attachment of his tongue to his hard palate.



Figure 1. Palatoglossal adhesion

- Other anomalies included cardiac defects, limb abnormalities and cleft palate



Figure 2.



Figure 3

Figures 2 and 3 depict the patient’s upper and lower limb anomalies.

Initial Management

- The patient was transferred from a community hospital in moderate respiratory distress with frequent apneic episodes
- He was taken to the operating room for orotracheal intubation and intubated via a transoral approach with a size 2.5 endotracheal tube over a 2.2 mm neonatal flexible bronchoscope

Further evaluation revealed a thick band of anterior tongue fused with hard palate and a posterior hard palate cleft with otherwise normal laryngeal and tracheal anatomy.

Surgical Considerations

- Bleeding risk in a neonate with low blood volume
- Increased risk of interventricular hemorrhage before 1 week of age
- Body temperature shifts in a very low birthweight neonate

Surgical Management

He underwent release of his anterior tongue at 9 days of age under general anesthesia.

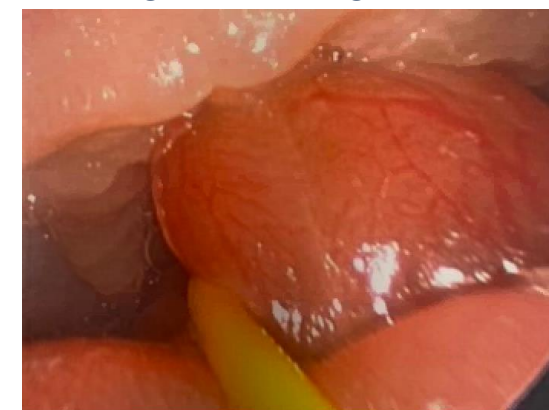


Figure 4. A vessel loop was passed around the tongue to provide traction in conjunction with a cotton tip applicator

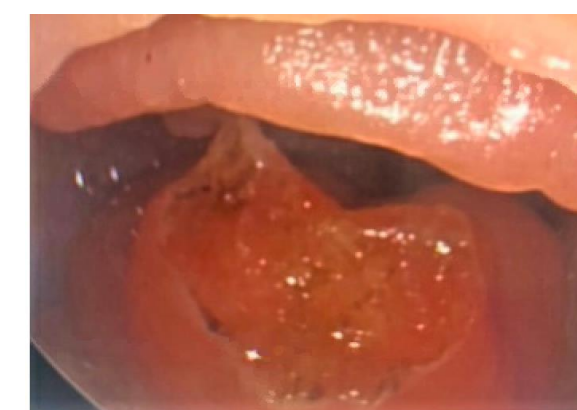


Figure 5. A Colorado tip bovie at 5W was used to incise the tethered tongue tip.

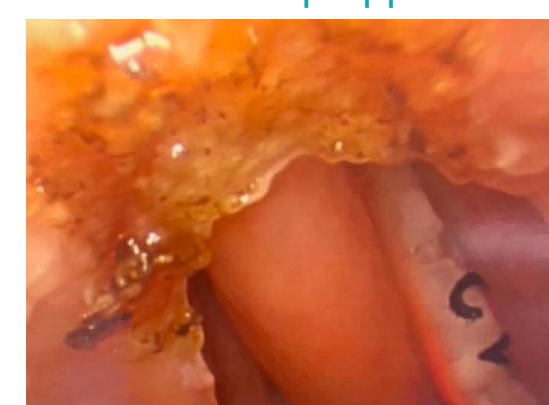


Figure 6. The hard palate defect after release of anterior tongue



Figure 7. Five interrupted 6-0 vicryl sutures were placed to close the raw mucosal edge

Post-op Management

- The patient remained intubated for 3 days post-op due to concern for airway compromise in the setting of post-operative tongue swelling
- A cotton tip applicator was passed intraorally twice a day to prevent re-adhesion of the tongue to the hard palate

The patient was successfully extubated to nasal CPAP on post-operative day 3.

- He underwent whole exome sequencing which revealed a variant of unknown significance in IRF6
- At age 4 months, he continues to require nasal CPAP due to chronic lung disease of prematurity and hiatal hernia causing pulmonary restriction
- He has poor oral intake due to nasal CPAP use as well as hypoglycemia requiring nutrition via naso-jejunal feeds

Discussion

- Literature search identified 27 cases discussing palatoglossal adhesion or Superior Ankyloglossia Syndrome
- Glosso-palatal release procedures were described under both local and general anesthesia
- A variety of potential causative mechanisms have been postulated such as amniotic banding, but no clear underlying pathophysiology or genetic cause has been elucidated
- Superior Ankyloglossia Syndrome presents difficulties in airway management as it can preclude the ability to perform direct laryngoscopy and makes orotracheal intubation difficult
- Despite the associated anomalies that have been reported with this syndrome, laryngeal anomalies are not common
- In a premature neonate, these airway concerns become more critical, as positive pressure ventilation and intubation may be temporarily necessary as lungs develop

Literature Review

Characteristic	Value
Gender	
Male	14 (51.9%)
Female	13 (48.1%)
Gestational Age	37.1 weeks ± 3.4
Age at time of repair	194d ± 658 Median: 17d
Weight at time of repair	3.02 kg ± 2.03 Median: 2.60
Anesthesia type	
Local	8 (29.6%)
MAC	7 (25.9%)
General	10 (37.0%)
Not reported	2 (7.4%)
Type of intubation	
None	15 (55.6%)
Nasotracheal	4 (14.8%)
Orotracheal	3 (11.1%)
Not reported	5 (18.5%)
Pre-op imaging	
None	18 (66.7%)
Fetal Ultrasound	3 (11.1%)
CT	2 (7.4%)
MRI	3 (11.1%)

Characteristic	Value
Maternal Age	25.3y ± 7.8
Delivery Method	
Normal Vaginal	5 (18.5%)
Cesarean	4 (14.8%)
Unknown but reported as uncomplicated	4 (14.8%)
Not reported	14 (51.9%)
Family History of similar anomalies	
Yes	9 (33.3%)
No	12 (44.4%)
Not Reported	6 (22.2%)
Respiratory distress	4 (14.3%)
Pre-op feeding difficulty	23 (82.1%)
Post-op feeding difficulty	2 (7.1%)
Associated syndrome or chromosomal abnormality	4 (14.3%)
Associated anomalies	
Cleft Lip/ Palate	17 (60.7%)
Limb Anomalies	11 (39.3%)
No associated anomalies	14 (24.8%)