Dysphagia
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CLINICAL CONUNDRUM

The Role of Dysphagia Assessment in the Identification of Upper Airway Obstruction in Infants

Laura Brooks[1] · Sean Evans[1,2] · Kristan Alfonso[1,2] · Shelby Lawrence[1] · Steven Goudy[1,2]

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Clinical Conundrum

Our patient is a full-term male diagnosed at birth with a soft palate cleft. He was discharged home after routine monitoring on room air, consuming formula via a specialty feeding system. At 6 days of age, he presented to our hospital with cyanosis and unresponsiveness associated with an emesis event after feeding. Physical examination demonstrated mild inspiratory stridor, but his bedside flexible fiberoptic laryngoscopy (FFL) revealed a normal nasopharynx (other than known soft palate cleft), normal oropharynx, patent supraglottis, and patent glottis. Due to audible inspiratory stridor both at rest and during oral feeds, a videofluoroscopic swallow study (VFSS) was performed, revealing only retrograde flow of the thin liquid into the nasopharynx consistent with a cleft palate and several shallow laryngeal penetration events. He was discharged from the hospital, but readmitted at 26 days of age for increase in stridor, apnea, and cyanosis for 2 days. A repeat FFL again revealed a normal upper airway. Speech Language Pathology (SLP) was consulted for a clinical feeding/swallowing assessment. Bedside assessment revealed inspiratory stridor, breath holding, and gasping during the oral feeding trial, with him consuming < 5 ml. The patient's inspiratory stridor and breath holding increased when he was held in near upright and reclined positions, with notable improvement when positioned in sidelying and prone concerning for upper airway obstruction. He was diagnosed with reflux and positioned in prone throughout the majority of the day and night with Intensive Care Unit monitoring. The patient's stridor and increased work of breathing continued, with additional retractions at baseline despite reflux management and positional supports. The patient's lateral images of his previous VFSS were re-reviewed with attention given to the tongue base and mandible and noted to be atypical when compared to similar aged infants by the SLP team (Figs. 1 and 2). What is the cause of his symptoms both at rest and during feeding?

Diagnosis

The VFFS images revealed that the patient's mandible was positioned more posteriorly relative to the maxilla, and the pharyngeal space between the tongue base and posterior pharyngeal wall (retrolingual space) was reduced (Figs. 3, 4 and 5). The Otolaryngology team confirmed the findings concerning for micro-retrognathia and glossoptosis as a cause for upper airway obstruction. Polysomnography was obtained, revealing severe obstructive sleep apnea (apnea–hypopnea index > 10 events an hour without central events). Direct Laryngoscopy and Bronchoscopy (DLB) demonstrated severe tongue base collapse with compression/obstruction of the supraglottic airway that improved with jaw thrust (Fig. 6). The patient was noted to have concomitant laryngomalacia, thus a supraglottoplasty was also performed. Despite the supraglottoplasty, the patient continued to demonstrate stridor, breath holding, and increased work of breathing requiring prone positioning during majority of day and night. Computed tomography scan was obtained, results revealing "micrognathic mandible." The diagnosis of Robin sequence (RS) was made, and soon after, genetic testing confirmed Stickler syndrome. Attempted non-surgical interventions (prone positioning, nasal trumpet) were unsuccessful at improving breathing and resolving oxygen desaturations, and the patient subsequently underwent bilateral mandibular distraction osteogenesis (MDO), resulting in significant clinical improvement of stridor and work of breathing and increased retrolingual space on repeat DLB (Figs. 7 and 8).

✉ Laura Brooks
laura.brooks@choa.org

[1] Children's Healthcare of Atlanta, 1405 Clifton Road NE, Atlanta, GA 30322, USA

[2] Department of Otolaryngology-Head and Neck Surgery, Emory University School of Medicine, Atlanta, GA 30322, USA

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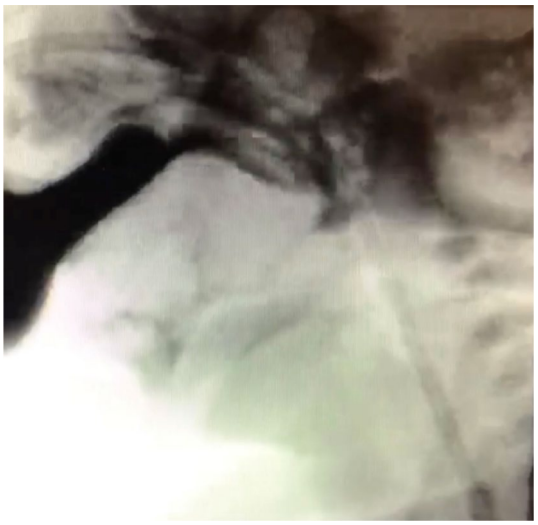

Fig. 1 Lateral VFSS view of our patient

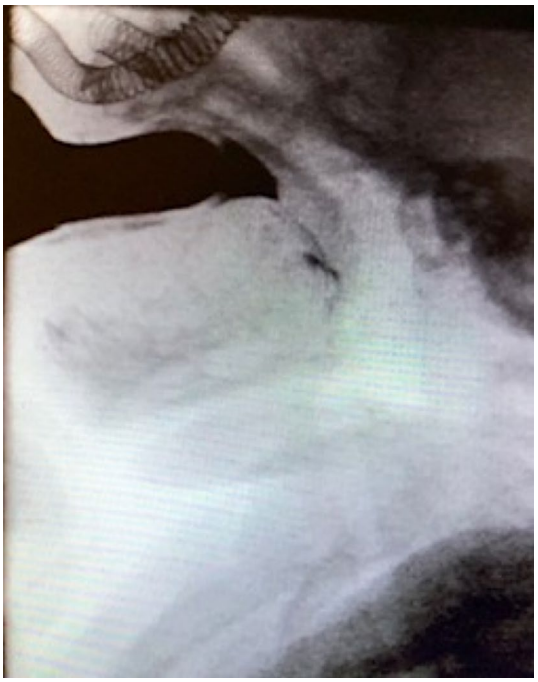

Fig. 2 One-month-old infant for comparison

He was discharged from our hospital sleeping in supine position on minimal oxygen support. Follow-up VFSS revealed aligned mandible and maxilla and improved retrolingual space (Figs. 9 and 10).

This case study highlights the challenges in identification of micrognathia and glossoptosis that can compromise early diagnosis of RS or Syndromic Robin sequence (SRS). There are limitations to the clinical examinations of micro-retrognathia, glossoptosis, and upper airway obstruction. The Jaw Index, for example, is a simple, cost-effective method used

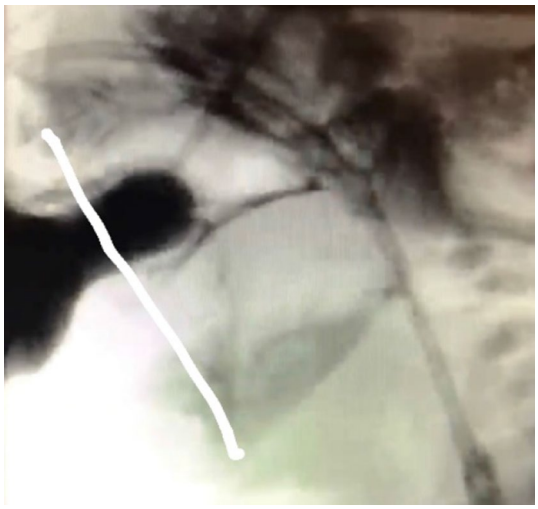

Fig. 3 Lateral VFSS view of our patient with Syndromic Robin sequence (Stickler) mandible

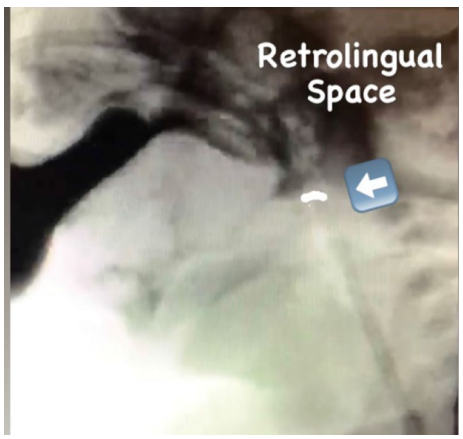

Fig. 4 Lateral VFSS view of our patient with Syndromic Robin sequence (Stickler)-retrolingual space

at the bedside to quantify retrognathia by measuring "Alveolar overjet × (Maxillary arch/Mandibular arch)." However, Wiechers et al. [1] highlighted that the Jaw Index may not capture the functional or clinical complications that the infant is experiencing given that it assesses retrognathia, which is similar to but not identical to micrognathia. They also underscored the challenges in obtaining an accurate measurement given infant activity that may compromise the results, such as crying. Poets et al. [2] described the challenges to diagnosing glossoptosis as the presentation may vary based on the position, age, and muscle tone of the infant. Infants with stridor and stertor often initially undergo assessment via awake endoscopy. However, glossoptosis tends to be more apparent in a relaxed state, which is unlikely to be present during an endoscopic evaluation

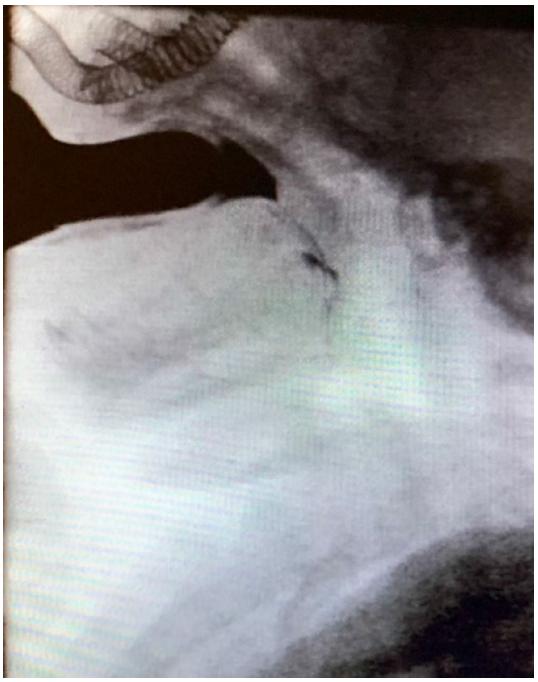

Fig. 5 One-month-old infant without RS or any syndrome for comparison

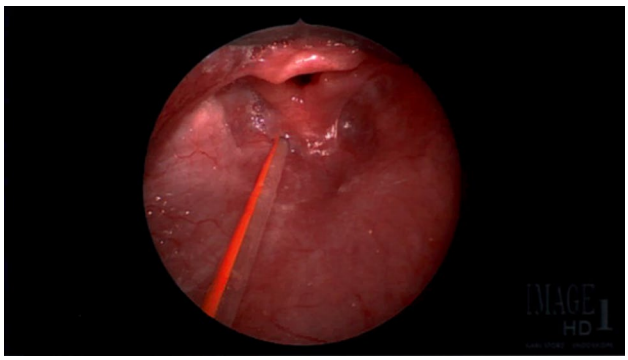

Fig. 6 Initial Direct Laryngoscopy Bronchoscopy (DLB)

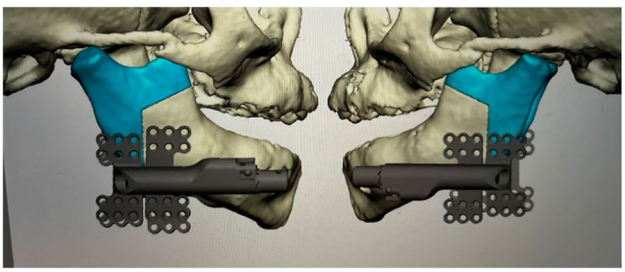

Fig. 7 Surgical planning for mandibular distraction osteogenesis (MDO)

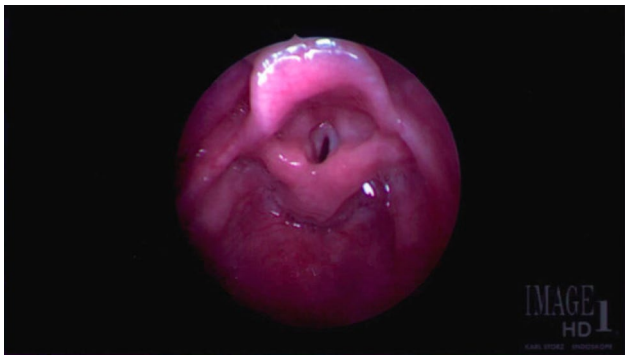

Fig. 8 DLB post-MDO

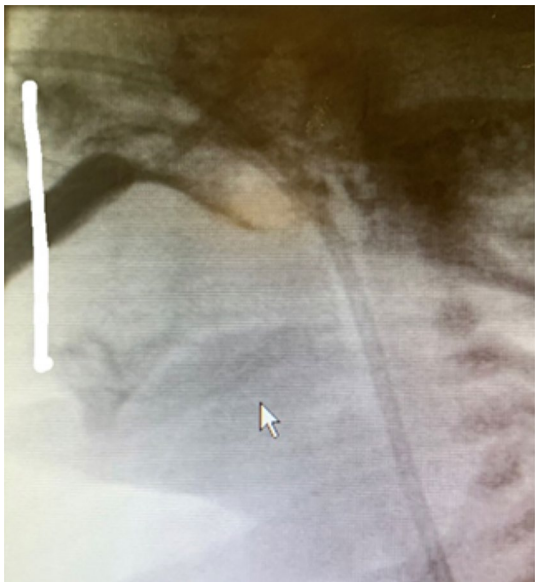

Fig. 9 Aligned Maxilla and Mandible post-MDO

of the upper airway in an awake infant who is crying and agitated. Another challenge in early diagnosis of RS is that the degree of upper airway obstruction is variable in these infants, compromising the ability for the physician to discriminate between what is normal and what is abnormal during examination. The mandible is less developed and more posteriorly displaced than the maxilla at birth for all infants [3], which highlights the potential for overlooking mandibular micrognathia with clinical observation.

Conclusion

Robin sequence is a congenital condition characterized by micrognathia, glossoptosis, and airway obstruction, often with cleft palate. Patients with RS frequently present with dysphagia associated with impaired suck-swallow-breathe

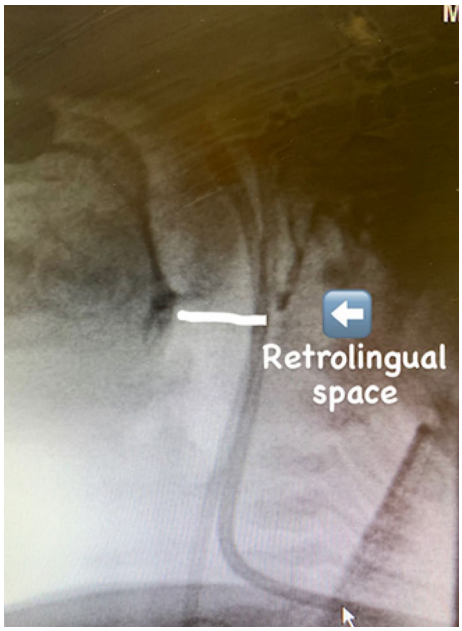

Fig. 10 Increase in retrolingual space post-MDO

coordination and impaired suction strength in those with a cleft palate. Dysphagia can result in aspiration, respiratory compromise, and failure to thrive. SRS increases the risk of morbidity and mortality [4] and can further contribute to swallowing dysfunction. While early identification is critical to mitigate the consequence of upper airway obstruction in RS, there are challenges to the early diagnosis of RS with clinical observation and FFL. A VFSS is frequently obtained in patients with dysphagia and offers the SLP a unique view allowing observation of the mandible position and retrolingual space in the lateral view. This information may be useful/complementary in guiding the treatment team to investigate the possibility of RS as a cause of upper airway obstruction and dysphagia.

Declarations

Conflict of interest Dr. Steven Goudy disclosure: CEO of Dr. Noze Best. Dr. Kristan Alfonso disclosure: Clinical consulting agreement with a Cochlear Implant company. The authors declare that they have no conflict of interest. There are no financial disclosures associated with this paper.

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Laura Brooks M Ed

Sean Evans MD

Kristan Alfonso MD

Shelby Lawrence MA

Steven Goudy MD