The Impact of Mandibular Distraction and Tongue to Lip Adhesion on Feeding in Infants with Isolated Pierre Robin Sequence: A Systematic Review
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Introduction
Pierre Robin Sequence (PRS) is a congenital disorder characterized by retrognathia/micrognathia, glossoptosis, and respiratory distress and may result in a cleft palate. These craniofacial abnormalities cause significant difficulties with feeding and breathing during infancy and childhood. Though surgical intervention is a potential solution for this population, it can only be performed after a certain level of physical development has occurred, often referred to as the Rule of 10s. Children with PRS have difficulty getting the proper nutrition to optimize growth, especially in combination with breathing difficulties, which can result in failure to thrive. It is clinically important to determine the best practices for managing patients affected by craniofacial anomalies and swallowing disorders, including children with PRS who have a complex array of symptoms that impact feeding.

Mandibular Distraction Osteogenesis
Mandibular Distraction Osteogenesis (MDO) is an intervention technique in which the mandible is gradually lengthened after an initial ostomy. The expansion is slow and steady to allow time for new bone tissue to grow. While expanding the mandible, mucosa, blood vessels, nerves and muscles are also elongated. The process takes about 8 weeks overall.

Methods
- PubMed and CINAHL databases were searched using the search string, “Pierre Robin AND (feeding OR dysphagia OR swallowing) AND (intervention OR treatment OR strategy OR therapy)”.
- Once duplications were removed, a total of 120 articles remained.
- The researchers each independently reviewed 85 titles and abstracts, 50 of which both researchers assessed using the following inclusion and exclusion criteria: Studies had to be on human infants up to 23 months of age, articles must be in English, articles must address feeding, articles which only looked at respiration were excluded.
- Articles were not restricted based on date of publication, except up to the date of the search. Articles which looked at novel techniques or single case studies or case reports were not included.
- The additional restriction of looking only at mandibular distraction osteogenesis (MDO) and/or tongue-lip adhesion (TLA) was later added due to the heterogeneity of results, which made comparisons difficult.
- The researchers had an agreement rate of 94% for which articles would move on to full text reviews and which should be included.
- 25 articles met the inclusion criteria based on this initial review, and after full text reviews and quality appraisals, 16 articles remained.
- Those that remained were determined to be of good quality using LEGEND appraisal checklists and addressed whether participants had PRS with other associated anomalies or syndromes or if they had isolated PRS.

Results
MDO and TLA are generally used to target respiratory distress and low levels of oxygen saturation, but are also very effective at treating feeding and swallowing abnormalities. MDO has been successful at increasing weight gain and oral feeding and at eliminating abnormal tongue movements, penetration into the laryngeal vestibule, stasis of residual material in the laryngeal recess, barium aspiration, gastroesophageal reflux disorder (GERD), and abnormally long pharyngeal phases of swallowing (longer than 1 second). According to some studies, it allows for a quicker transition to oral feeding than TLA, while others assert that oral feeding can begin immediately after the tubes are removed post TLA procedures. While MDO is generally thought to be a last resort when more conservative methods, including TLA, fail, it is far more successful at providing long term relief of feeding disorders in patients with PRS. However, patients with isolated PRS (absence of any concomitant abnormalities or syndromes) tend to have great success when managed via TLA and do not require MDO or other secondary surgeries for recurrent airway or feeding disorders. TLA requires a shorter recover period than MDO. This is important to consider since TLA has been shown to shorten hospital stays, simplify nursing, and make home care less demanding, but the benefits should be weighed against the inability of TLA to completely address anatomic deficiency, its potential long-term effects on the feeding process by restricting tongue movement, and the fact that it often requires secondary procedures where MDO does not.

Conclusion
Some have asserted that MDO does not immediately relieve problems like TLA can and that MDO performed at a young age can cause postoperative feeding difficulties and subsequent decline of growth rate. However, it has a lower rate of respiratory distress recurrence after discharge than TLA, better quality of life, and has been shown to provide better feeding outcomes than TLA in many cases. While the least invasive procedures should be considered first, especially in cases of isolated PRS, the higher immediate and long-term success rates of MDO in both isolated PRS and in patients with PRS with comorbid syndromes and anomalies warrants consideration and further study.

Our results are based on multiple research studies with rather small sample groups. A majority of these studies were conducted using fewer than 50 participants, due to medical complexity of PRS and the relative rarity of studies, which looked at patients with isolated PRS not occurring in conjunction with another syndrome and were conducted largely in hospitals with easy access to participants. These small, non-random studies may have skewed research findings in the direction of success for distractors, as they are most commonly used.

Tongue-Lip Adhesion
Tongue-Lip Adhesion (TLA) is a surgical intervention technique in which the tongue is surgically anchored anteriorly to the lower lip. As the base of the tongue is pulled forward, the oropharyngeal airspace is opened, effectively reducing airway obstruction. This intervention technique does not affect speech production and is a much less invasive procedure than a tracheostomy.

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