Pierre Robin Sequence

Evaluation, Management, Indications for Surgery, and Pitfalls

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KEYWORDS

• Cleft palate • Micrognathia • Glossoptosis • Pediatric sleep apnea
• Mandibular distraction osteogenesis • Tongue-lip adhesion • Pierre Robin Sequence

KEY POINTS

- Pierre Robin Sequence (PRS) is a triad of micrognathia, glossoptosis, and cleft palate, which may occur as isolated findings in an otherwise normal child or be associated with additional syndromic features.
- Children with PRS may exhibit varying degrees of upper airway obstruction.
- Management of upper airway obstruction is best approached conservatively, using nonoperative interventions first.
- The majority of infants are stabilized by nonsurgical measures.
- Children with neurologic deficits at risk for chronic aspiration are better served by traditional surgical interventions such as tracheotomy and gastrostomy-tube placement.
- Mandibular distraction osteogenesis is the only surgical technique that directly addresses the underlying cause of upper airway obstruction in PRS without compromising feeding. It offers results that persist through early childhood.
- Mandibular distraction procedure mandates specialized training.
- Complications of neonatal mandibular distraction osteogenesis:

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Short Term	Long Term
Infection	Tooth loss
Hardware failure	Scarring
Bone resorption and regression	Asymmetry
Nonunion	Corrective orthodontics
Facial nerve injury	Additional surgeries
Open-bite deformity	

Much has been written about airway interventions in children with Pierre Robin Sequence (PRS), with many investigators from a variety of disciplines advocating any number of methods used to manage the problem. This article seeks to review the various methods of treating airway obstruction and feeding difficulty in infants with PRS, and highlights the benefits and limitations of early mandibular distraction osteogenesis, particularly as a way of managing both airway obstruction and feeding difficulty in these children.

PRS was first described in 1923 by Pierre Robin, a French stomatologist, as a diad of micrognathia and glossoptosis.[1] In 1934 he revised the definition to include a triad of micrognathia, glossoptosis, and a U-shaped cleft palate.[2] It is not a syndrome in itself, but rather a sequence in which multiple secondary anomalies are derived from a single anomaly, and affects approximately 1 in 8500 births. The prevailing hypothesis implicates hypoplasia of the mandible (either from a primary growth disturbance or from hyperflexion of the neck) before 9 weeks in utero as the inciting factor. The small jaw positions the tongue posteriorly and superiorly where it lies between the 2 palatal shelves, physically preventing their fusion, which normally occurs between the 8th and 10th weeks of gestation. It is this mechanical disruption of palatal closure and not a primary molecular or genetic factor that leads to the palatal cleft. Many newborns with PRS have upper airway obstruction and exhibit varying degrees of respiratory distress, which may require intervention. In addition, children may struggle with oral feeding, owing to the difficulty in coordinating breathing and swallowing in the context of tongue-base obstruction and a cleft palate.

DIAGNOSIS OF PIERRE ROBIN SEQUENCE

The diagnosis of PRS is typically made at birth. With improvements in prenatal imaging including high-resolution ultrasonography, micrognathia is now being diagnosed as early as the second trimester of pregnancy. In light of this there has been an attempt to standardize mandibular measurements on ultrasonography, including comparison with other cephalometric proportions[3,4] and creation of a mandibular index, which takes into account the anterior/posterior dimension of the mandible as it relates to the biparietal diameter of the fetus.[5] In practice, however, these measurements are rarely used, and micrognathia is often a relatively subjective call by the examining maternal fetal medicine physician. Inspection of the mandible is possible with ultrasonography, but evaluation of the tongue and palate is less reliable. Although the diagnosis of micrognathia can be made in utero, confirmation of the complete triad of PRS before delivery is usually not possible.

Physical Examination

At birth, the micrognathia is the most striking feature. This trait is characterized by a small and retrusive mandible in which the mandibular alveolus is significantly posterior to the maxillary alveolus. Examination of the oral cavity reveals posterior and