Surgical Considerations in Pierre Robin Sequence

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KEYWORDS

• Pierre Robin sequence • Airway protection • Micrognathic patients • Mandibular Distraction

KEY POINTS

- A portion of Pierre Robin infants will escape surgical intervention for airway protection because of postnatal growth at the posterior ramus and condylar growth sites.
- The use of a Decision Tree Model with appropriate consultants and diagnostics studies is useful to determine the best candidates for mandibular distraction.
- There was a greater that 50% reduction in tracheostomy when mandibular distraction was included in the treatment algorhythm.
- Lower morbidity and health care costs was seen when airway protection was achieved with mandibular distraction as opposed to tracheostomy.

INTRODUCTION

In the early twentieth century, Pierre Robin described a triad of manifestations in neonates that included micrognathia, glossoptosis, and upper airway obstruction.[1] Robin's description is now considered to be a sequence of developmental events without a specific genetic basis. Reports of the incidence of Pierre Robin sequence (PRS) vary widely due to the comparative rarity of the manifestations and differences in diagnostic criteria. Specifically, the presence of cleft palate is a debatable absolute inclusion criterion for PRS among some institutions. The 2 best studies from regional or institutional data in Liverpool and Copenhagen estimate the incidence for PRS between 1 case per 8500 neonates and 1 case per 14,000 neonates.[2,3]

PRS can be seen as an isolated phenomenon or in combination with other malformations. From a compilation of data from the authors institution and other centers, isolated PRS occurs in 58% to 70% of patients.[3–6] In patients who had PRS in combination with syndromes, Stickler, 22q deletion, and Treacher Collins syndrome have been the most prevalent. When PRS occurs in isolation, in utero deformational factors such as intrauterine growth restriction are generally considered to be important.[7]

ANATOMIC CHANGES IN PIERRE ROBIN SEQUENCE

The approach to a patient with PRS begins with a complete understanding of the anatomic basis of the sequence. The mandible is a first pharyngeal arch derivative and formed by the primordium of migrating neural crest cells that appear during the fourth week of gestation.[8] At 6 weeks of gestation, the trigeminal nerve stimulates endochondral osteogenesis from Meckel cartilage, forming the major structures of the mandible. When genetic syndromes or deformational factors disrupt prenatal growth, micrognathia ensues and causes the retropositioning of the tongue. If this process occurs before the eighth week of gestation, closure of the palatal shelves is prevented, thereby resulting in the formation of a U-shaped cleft palate.[9]

Postnatal mandibular growth occurs via the coordination of appositional and endochondral growth at 2 major sites. The increase in mandibular length is largely accomplished by appositional forces at the symphysis, resulting in the formation of bone at the posterior border of the ramus and the anterior border of the body during the first year of life.[10] Subsequently, chondrogenic proliferation at the condyles precedes endochondral

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ossification and increase in vertical ramus height. Although neonates born with PRS may have clinically significant symptoms, a portion of children may escape surgical intervention due to postnatal mandibular catch-up growth. This phenomenon is generally more common to patients with isolated PRS, likely due to the release of interference by deformational intrauterine factors after delivery. Work by Randall and colleagues[11] emphasized that the catch-up effect is limited to only a subset of patients. They conducted a longitudinal evaluation of a cohort of patients with neonatal micrognathia with mild-to-severe respiratory obstruction and discovered 3 patterns of mandibular growth. One group of patients eventually showed an almost normal size and position of the mandible. A second group of patients showed persistent micrognathia throughout childhood. A third group of patients had a similar type of underdevelopment of the mandible but with compensatory mechanisms that allowed a fairly normal anterior relationship. Clinically, Monasterio and Figueroa, in separate studies, observed that some patients who were treated conservatively continued to have respiratory disturbances throughout life.[12,13] Thus, it is clear that specific criteria are necessary to systematically identify patients who need definitive intervention versus those who need expectant management.

GENERAL CONCEPTS FOR TREATMENT

The most critical consequence of PRS is upper airway obstruction in the neonatal period. The severity of airway obstruction is largely defined by the interventions necessary to maintain patency. In the mildest of circumstances, side or prone positioning may be sufficient to prevent respiratory compromise. In patients who cannot maintain airway patency with positioning alone, intubation using a nasopharyngeal tube or an endotracheal tube may be necessary. In neonates who require intubation for survival, a surgical procedure is virtually universal in order to extubate the child. The current armamentarium of airway interventions includes glossopexy, mandibular distraction, or tracheostomy.

The nature of the upper respiratory problems has been defined by several investigators. Using fiber optic endoscopy, Sher sought to delineate the causes for airway obstruction in PRS. With the criteria of micrognathia, cleft palate, and airway obstruction, Sher found that obstruction can occur via

1. Retropositioning of the tongue to compress against the posterior pharyngeal wall
2. Retropositioning of the tongue to compress the soft palate to the posterior pharyngeal wall
3. Lateral pharyngeal wall collapse
4. Generalized pharyngeal wall collapse

Correlating his observational data to outcomes, Sher found that glossopexy relieved airway obstruction symptoms when retropositioning of the tongue resulted in compression of the pharyngeal wall. All 3 latter circumstances could not be relieved by glossopexy alone. Cozzi and Pierro added to the knowledge of pathologic airway obstruction by noting the oropharyngeal collapse brought about inadequate pharyngeal support to resist the force of high inspiratory negative pressures.[14] They reported that in normal breathing the pharynx does not collapse in the face of negative intrathoracic pressure, because a neuromuscular mechanism motored by the genioglossus retains patency. In PRS, retrodisplacement of the mandibular attachment of the genioglossus diminishes the ability to hold the tongue out of the pharyngeal airway. Although glossopexy has the ability to temporarily relieve obstruction due to a retropositioned tongue, the procedure depends on sufficient catch-up growth for reversal. Glossopexy does not address the core anatomic disturbance. Based on this assessment of the medical literature, the preference of the authors is to either offer definitive surgical management or nonsurgical management.

In the event that a neonate demonstrates signs of critical upper airway obstruction, the authors advocates for an algorithm that emphasizes distraction osteogenesis with the caveat that other abnormalities are absent. Over the past 2 decades, mandibular distraction osteogenesis has become an important technique to reduce the proportion of children with PRS who undergo tracheostomy.

The general concept of distraction begins with an initial osteotomy followed by a mechanical linear force from the distraction device that serves to direct the formation of bone. There are 3 phases of the distraction process: latency, activation, and consolidation. Latency is the period immediately following the osteotomy, which is usually 0 to 2 days for the mandible. During the activation phase, distraction occurs at a specific rate and rhythm. A typical rate of distraction is 1 mm/d with a rhythm of turning the devices twice a day (eg, 0.5 mm in the morning and 0.5 mm at night). The length of distraction is determined by the size of the defect or the length necessary to correct a functional problem. Finally, molding the regenerate may also be a consideration during the activation phase to close an open bite, correct form, or improve symmetry while the bone remains soft. Consolidation