Bilateral Mandibular Distraction Osteogenesis in the Neonate with Pierre Robin Sequence and Airway Obstruction: A Primary Option

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Abstract

Children with craniofacial abnormalities associated with retromicrognathia and glossoptosis often have compromised upper airway flow. In severe cases, emergency intubation is necessary immediately after birth, and tracheostomy is advocated to manage the airway in the neonatal period and to allow for feeding. Early intervention with bilateral mandibular osteogenesis avoids the need for tracheostomy, along with its complications, and it targets the primary etiologic factor of the problem—the anomalous anatomy of the mandible. We report two neonates with severe Pierre Robin sequence managed with bilateral mandibular distraction osteogenesis on day 9 and day 11 of life. The surgical techniques and distraction and consolidation periods were similar apart from the distraction devices used. The procedures were successful with early extubation (day 5 and day 7), oral feeding tolerance (day 11 and day 13) and hospital discharge (day 19 and day 18). Total mandibular distraction was 19 mm and 23.45 mm, respectively. No major complications were reported. Medium to long-term results were good. Bilateral mandibular distraction osteogenesis in the neonate is a safe and accurate procedure and is the primary option in cases of selected severe Pierre Robin sequence.

Keywords

- distraction osteogenesis
- micrognathia
- Pierre Robin
- mandibular distraction

Children with micrognathia often have compromised upper airway flow. This situation has high potential for morbidity and mortality and poses a difficult problem to solve.¹ The clinical signs and symptoms of obstruction are multiple and include direct and immediate signs like decreased oxygen saturation, carbon dioxide retention, positional inspiratory obstruction, episodic apneas or bradycardias, or even complete airway obstruction leading to respiratory arrest and death.² There is also a group of systemic less-defined signs that are of parallel importance in the morbidity of this clinical entity, such as poor

feeding, failure to thrive, insufficient weight gain due to malnutrition, and frequent pulmonary morbidity with long-term effects like pulmonary hypertension and cor pulmonale.³ The majority of children who are born with a symptomatic combination of microretrognathia are diagnosed with Pierre Robin sequence. Other craniofacial disorders that can produce a similar clinical scenario are the Treacher Collins syndrome, Crouzon syndrome, Nager syndrome, 22q deletion syndrome, and bilateral hemifacial microsomia.⁴ Pierre Robin sequence is characterized by the triad micrognathia, glossoptosis, and

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airway obstruction as originally described by Pierre Robin, a French stomatologist, in 1923. It is estimated to occur in 1 in 8500 to 20,000 births.⁵ Other findings that can be present are cleft palate, feeding difficulties, and failure to thrive.⁶ A sequence differentiates from a syndrome as it is considered that its multiple anomalies are caused secondarily by one of the anomalies present; in a syndrome, there is a cluster of anomalies with no apparent relationship between them.⁵ The primary malformation in the Pierre Robin sequence is thought to be mandibular micrognathia, which causes glossoptosis (upward and backward position of the tongue in the oropharynx) with subsequent airway obstruction and cleft palate formation.¹ The degree of respiratory difficulty depends on the severity of the micrognathia and glossoptosis. The majority of children with Pierre Robin sequence have a mild to moderate degree of airway obstruction and can be successfully treated with conservative treatments,^{1,4,7-9} such as continuous prone positioning, continuous nasal positive airway, and nasopharyngeal intubation. On the other hand, moderate to severe obstructive airway symptoms do not respond to these measures and demand more invasive solutions. Traditionally, the most commonly used procedures for this refractory cases have been glossopexy or tongue-lip adhesion (TLA) and tracheostomy.⁴ Less popular treatment modalities have been described, such as hyomandibulopexy and subperiosteal release of the floor of mouth musculature, but have not proven consistent results yet.³ Tracheostomy, the gold standard treatment, has a high rate of immediate and long-term complications and long hospital and neonate intensive care unit stays.⁴ In recent years, bilateral mandibular distraction osteogenesis (DO) has shown up as an alternative for the most severe cases of Pierre Robin sequence with good and consistent results in older children. It has the advantage of addressing the cause of the problem as a truly curative treatment, as it increases the length of the mandible, adding projection to the mandible arch and space to the superior airway. We describe the use of bilateral mandibular distraction in two neonates with severe Pierre Robin sequence requiring very early endotracheal intubation for airway control.

Materials and Methods

Two newborns with Pierre Robin sequence were referred to our department with severe respiratory distress requiring endotracheal intubation on the day 3 and day 1 of life as candidates for tracheostomy (►Figs. 1A, 1B, and 2A). A multidisciplinary team approach—encompassing neonatal intensive care specialists, pediatricians, anesthesiologists, pediatric cardiologists, pediatric surgeons, pulmonologists, and plastic surgeons—was held and a thorough clinical examination was done to determine other possible congenital anomalies or associated syndromes. Case 1 was diagnosed with Weissenbacher-Zweymuller syndrome (association of fetal chondrodysplasia and Pierre Robin sequence), and in case 2, a minor auricular communication was detected through echocardiography and managed conservatively. Both neonates presented incomplete cleft palates. A careful evaluation of the upper and lower airways is mandatory and it should include direct laryngoscopy and flexible fiberoptic

bronchoscopy to evaluate the role of tongue position in the respiratory obstruction and rule out other possible causes such as subglottic stenosis or tracheomalacia. In both neonates, the tongue was positioned superiorly and posteriorly causing pharyngeal obstruction (►Fig. 2B). Also, pulmonary computed tomographic (CT) scans were done to provide information about the lower airways. No other abnormalities were detected in these exams. Neonate 2 was unable to feed orally and was fed via an orogastric tube. Maxillofacial evaluation included clinical examination with measurement of the maxillomandibular discrepancy and imaging techniques to perform cephalometric analysis. In case 1, anteroposterior and lateral craniofacial X-rays were done, and in case 2, we chose to perform 3-D CT, which we think gives much more valuable information (►Fig. 2C). The maxillomandibular disharmony measured at the anterior midline alveolar ridge was 10 mm in case 1 and 12.5 mm in case 2. We proposed bilateral mandibular DO to address the micrognathia/tongue position avoiding tracheostomy. The collected data enabled detailed surgical planning encompassing evaluation of bone availability and quality for screw placement, tooth buds, roots and inferior alveolar nerve localization, occlusion plane and maxillomandibular discrepancy. We planned the vector of distraction, the distractor type and placement (bone location and surgical approach), the mandibular osteotomy line, the location of the activation pin, and we estimated the necessary lengthening. The surgical procedures were performed on day 9 (case 1) and day 11 (case 2) of life. We used single-vector distractors implanted through a submandibular approach in both cases to obtain the best bone exposure. In case 1, a 2.0-mm mandible distractor (Synthes, GmbH, Solothurn, Switzerland®) was used (►Figs. 1D, 1E, and 1F); in case 2, a more adequate 1.0-mm mesh footplate distractor (Synthes, GmbH, Solothurn, Switzerland®) was used (►Figs. 2D, 2E, and 2G). Osteotomy was done in the mandible angle in a plane perpendicular to the occlusion plane (►Fig. 1C). Distraction started without delay in the first postoperative day at a rhythm of 2 mm/d until day 5 and then 1.5 mm/d until day 12 in case 1 and at a rhythm of 1.8 mm/d for 13 days in case 2 (►Fig. 2E). The differences in the distraction rhythm were due to the distractors used. Prophylactic antibiotic therapy with amoxicillin/clavulanate was used during the distraction period. Bacitracin ointment was applied daily prevent pin tract site infection.

Results

A total of 19 mm and 23.4 mm of distraction was obtained in cases 1 and 2, respectively. Obstructive airway symptoms diminished rapidly as the distraction started, and extubation was possible on day 5 in case 1 and day 7 for case 2. At this time, the initially superoretro-positioned tongue could already be seen in its normal horizontal position in the oral cavity (►Fig. 1D). Oral feeding tolerance was obtained on day 13 (case 2) allowing gastric tube removal. Patients were discharged home from the intensive care unit on the postoperative day 19 (case 1) and postoperative day 17 (case 2). There were no complications in the postoperative period.

Figure 1 Case 1. (A and B) Child diagnosed with Weissenbacher-Zweymüller syndrome (association of fetal chondrodysplasia and Pierre Robin sequence) who suffered from severe respiratory distress requiring orotracheal intubation on day 3 of life, which was managed with mandibular bilateral distraction osteogenesis. Note the small, upturned nose with a flat bridge and the retromicrognathia typically seen in this syndrome. (C) Intraoperative view showing the mandibular osteotomy at the angle via a submandibular approach. (D) Postoperative day 5: the initially superoretro-positioned tongue could already be seen in its normal horizontal position in the oral cavity. (E) Postoperative day 7: child with no signs of respiratory distress after extubation and with a noticeable advancement of the mandible in comparison with the preoperative situation. (F) Radiographic image at the end of the distraction period shows an adequate upper airway passage. (G, H, and I) At 6 years old showing a good result with slight retrognathia but with good mandibular symmetry.

Consolidation periods lasted 6 and 12 weeks for cases 1 and 2, respectively, and control X-rays were taken at the end of distraction period and 6 weeks later to evaluate bone consolidation (►Figs. 1F and 2G). Neonate 2 developed two episodes of bronchiolitis during the consolidation period, which caused for the delay in distractor removal. In both neonates there was an apparent difficulty in mouth closure during the consolidation period that gradually faded by postoperative month 3.

Both patients have undergone palatoplasty without complications. The follow-up period for case 1 is 6 years and the child presents some degree of retrognathia (without any lateral deviations), which may render an orthognathic surgical procedure later in adolescence (►Figs. 1G, 1H,

and 1I). She is under orthodontic follow-up and has shown normal tooth eruption. Case 2 has a follow-up of 2 years and an apparently normally growing mandible without signs of recurrence (►Fig. 2I). They both have normal growth patterns. No facial palsy was seen in either child, and case 1 confirms to have normal inferior lip sensation.

Discussion

Micrognathia is an important cause of airway obstruction in the neonate. It is also the primary malformation in Pierre Robin sequence.⁵ The majority of children with Pierre Robin sequence are asymptomatic and can be treated conservatively.^{2,4,8,9} However, in moderate to severe cases of micrognathia, airway

Figure 2 Case 2. (A) Child with Pierre Robin sequence who suffered from severe respiratory distress requiring nasotracheal intubation on day 1 of life and was managed with mandibular bilateral distraction osteogenesis on day 13 of life. (A, B, and C) Note the severe maxillomandibular disharmony with retromicrognathia, evaluated as 12.5 mm in the computed tomographic scan that causes an important upper airway obstruction visible on the radiographic examination. (D) Intraoperative view showing the application of the 1.0-mm mesh footplate distractor (Synthes, GmbH, Solothurn, Switzerland®). (E) Distraction started on postoperative day 1 at a rhythm of 1.8 mm/d. (F) At 3 weeks postoperatively, the child was able to breast-feed without problems. (G and H) Radiographic images at the end of the distraction period and at 8 months postoperatively, respectively, showing the correction of the maxillomandibular disharmony, “opening” of the upper airway passage, and stability of the result. (I) At 2 years old showing a good result with an apparently normally growing mandible and normal development of primary dentition.

obstruction symptoms develop frequently and disturb basic functions like sleeping and/or feeding, thus becoming an important cause of morbidity and mortality.² They can even become constant and require continuous tracheal intubation for breathing. Traditionally, glossopexy/TLA procedures were the first choice in the approach to these patients and if they failed to relieve the symptoms, or in the most severe cases, tracheostomy was advocated.⁴ TLA is a relatively simple procedure and is considered effective^{8,9,10} when obstruction is limited to the tongue base. However, it has early complications in up to 55% of the cases,¹⁰ dehiscence being the most frequent, and longitudinal evaluations reveal that patients require additional procedures for ultimate control of airway, feeding, and orthognathic relationship.¹¹ The fact that TLA is only a soft tissue procedure not addressing the skeletal malformation of micrognathia must contribute to these unsatisfactory results.

Also, controversy exists in the impact of TLA in future tongue function.^{9,11} Tracheostomy in infants has a specific impact, and morbidity and mortality rates are significant. The tube may become displaced or plugged with secretions resulting in death if early recognition and treatment are missed. The mortality rate from the tracheostomy alone independent of the underlying diagnosis is as high as 5%.⁶ Long-standing tracheostomies are associated with tracheomalacia, chronic bronchitis, recurrent granulomas, laryngeal stenosis, and growth retardation.³ Average age of decannulation is 2 to 3 years, and this duration has a significant impact in speech and language development.^{2,3} Another important subject is the complex and specific nursing care surrounding the tracheostomized infant.

DO involves gradual bone lengthening across an osteotomy line and was initially described for limb orthopedic surgery

by Codvilla in 1905 and later refined by Ilizarov in the 1940s for lengthening of long bones.¹² Clinical application of this technique to the craniofacial skeleton was first described in the 1990s by McCarthy et al, and had an important contribution by Molina and Ortiz-Monasterio with the development of specific mandibular distraction devices and technique.⁴ DO has gradually been used as a new technique to obtain mandibular lengthening in patients with mandibular hypoplasia and airway obstruction and has proven to be safe, accurate, and capable of good and lasting results.^{1,2,4,6,9,12–16} It is a relatively simple and quick (90 to 120 minutes in our series) surgical procedure and has been used successfully in adults and children.^{2,4,6,7,9,12} It classically involves three distinct time periods: latency, distraction, and consolidation phases. In the specific case of neonate DO, these periods can be accelerated because of the particularly high metabolic rate. So, the latency period after osteotomy can be diminished to 1 to 5 days; distraction rhythm can be up to 2 mm/d, and consolidation periods of 6 to 16 weeks are enough to obtain strong “new bone.”^{2,4,15} Several groups have reported successful results like ours with DO for airway obstruction management in the children with micrognathia.^{2,6,9,12–16} Mandibular DO addresses the primary etiologic factor—micrognathia—as it lengthens the mandible, bringing forward the genioglossus and geniohyoid muscles and the tongue base.¹² These anatomic modifications increase the upper airway volume (mean improvement in cross-sectional area of 67%) and relieve the supraglottic airway obstruction.^{3,6} Cohen et al¹⁶ described 16 children with upper airway obstruction (eight with tracheostomies) who underwent treatment with mandibular DO. They managed to decannulate seven of the eight tracheostomized patients and obtained clinical improvement of sleep apnea signs and symptoms in the remaining children. Denny et al⁴ reported the successful use of mandibular DO to correct airway obstruction in 10 young patients (eight under 30 months of age), with measurable airway increase and clinical improvement obtained in all patients. Two patients with Nager syndrome and tracheostomy were successfully decannulated and became asymptomatic. The same group reported a series of five neonates with Pierre Robin sequence and life-threatening airway obstruction in which early mandibular DO (6 to 26 days of life) avoided the need for tracheostomy, completely eliminated the obstructive symptoms, and corrected the micrognathia.¹⁴ Rachmiel et al³ reported on the 3-D CT scan analysis of 12 children with obstructive airway symptoms treated with bilateral mandibular DO. They showed increased airway volume in all patients along with gradually increased oxygen saturation and disappearance of obstructive sleep apnea symptoms. Long-term results have also been encouraging. In a 12-year experience of DO procedures for mandibular hypoplasia, Molina¹⁷ considered it to be the treatment of choice for the surgical correction of unilateral or bilateral cases with minimal patient discomfort and an exceedingly low complication rate. Denny and Amm⁶ reported a 5-year follow-up of 11 neonates with Pierre Robin sequence in which DO was applied successfully to avoid tracheostomy. Normal sleep studies, normal growth, and normal mandibular shape

were obtained with an undisturbed tooth eruption sequence. Genecov et al¹⁵ reported the use of DO in 67 children with mandibular airway obstruction syndrome. Tracheostomy was prevented in 96% of the patients treated, and decannulation after DO was possible in 92%. They concluded that mandibular DO is an excellent treatment for mandibular airway obstruction in children younger than 6 years who do not respond to conservative measures.

The recent development of smaller and more adequate internal distractors, specifically for use in the neonate, enables its use in these cases. They are more precise, as they are in direct contact with the bone, and device failure is less common than with the use of external distractors (3% versus 10% on external devices).¹⁵ The major inconvenience is that, in neonates, visualization is often quite limited through an intraoral approach, making exposure of the osteotomy site and placement of the device quite difficult. The external submandibular incision gives excellent exposure for pin or screw placement, device placement, and completion of the osteotomy.² It is also true that the submandibular incision is relatively hidden, especially when compared with the scarring that results from the use of external devices.

Nevertheless, limitations and complications exist in this technique, and the surgical risks are not negligible. Facial nerve injury, tooth buds injury, and bleeding are potential risks.² Also, the surgical procedure is delicate and a correct device implantation is pivotal. The most common postoperative complication is pin site infection^{13,15} that responds to oral antibiotics. Other complications reported are bony non-union, premature bone consolidation, and facial nerve paresis during distraction phase that always resolved with stopping or reverting of the lengthening.¹³ Device failure is also significant and more common on external devices (10% versus 3% on internal devices).¹⁵

Conclusion

Based on our experience and on the several reports in literature, we consider that a detailed and extensive workup performed by an experienced multidisciplinary team is essential in the evaluation of a neonate with micrognathia and airway obstruction. In the case of a severe obstruction limited to the tongue base/hypopharyngeal space, bilateral mandibular DO is a safe and accurate procedure that acts directly on the etiologic factor—micrognathia—and successfully avoids the use of a more aggressive and complication-prone procedure as tracheostomy. So, it should be considered the primary option in selected cases of neonates with Pierre Robin sequence.

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