Neonatal Mandibular Distraction Osteogenesis

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Abstract

Mandibular distraction has revolutionized the treatment of Robin sequence associated with severe airway obstruction. The distraction technique remains the only intervention that directly corrects mandibular hypoplasia and the retropositioned tongue, providing efficient relief of airway stenosis. Multiple studies have demonstrated the efficacy of distraction in avoiding tracheostomy and decreasing the severity airway obstruction in this patient population. The benefit to avoiding tracheostomy and relieving airway obstruction is superior to that of tongue–lip adhesion. It is, therefore, not surprising that mandibular distraction has become the first-line intervention at many centers for the surgical treatment of Robin sequence. The complication profile associated with mandibular distraction appears low; the most common complication is infection, which can be treated by antibiotics alone. The severity of airway obstruction can be quantified by polysomnogram: This tool has become one of the most widely used objective metrics in the Robin sequence population. Therefore indications for surgery, timing of palatoplasty and long-term assessment of airway function should be performed in conjunction with sleep study analysis. The effects of mandibular lengthening on feeding difficulty in Robin sequence patient remains a topic of controversy. Studies have demonstrated conflicting results: This can be an area of future study. Agreed-upon indications for surgery and definitive protocols of care have yet to be formulized; future research should focus on achieving these goals. Such studies would require agreed-upon terminology for Robin sequence, an increase in comparative and prospective analysis, and the use of quantifiable metrics of clinical results.

Keywords

- ▶ neonatal
- ▶ mandibular distraction osteogenesis
- ▶ obstructive sleep apnea
- ▶ gastroesophageal reflux

Background

Robin sequence is defined as a triad of retrognathia, glossop-tosis, and airway obstruction.¹ Prevalence of this disorder ranges from 8,500 to 20,000 births per year.^{2,3} During fetal development, the hypoplastic mandible displaces the tongue into the palatal space, often preventing normal fusion of the palatal shelves.⁴ A U-shaped cleft involving the secondary and posterior primary palate is seen in 66% to 90% of affected patients.^{2,5,6} Some groups erroneously consider cleft palate to be a diagnostic criterion,^{7,8} despite Robin's original definition of the disease.

Retropositioning of the tongue base causes airway obstruction and increased work of breathing (–Fig. 1). Secondary

comorbidities resulting from airway obstruction include failure to thrive, developmental delay, reflux, feeding difficulties, CO₂ retention, heart failure, brain damage, and sudden death.^{9,10} The morbidity of upper airway obstruction in an infant can be significant. As Randall described,¹¹ the infant “can literally exhaust himself to death unless the obstruction is relieved.” Upper airway obstruction associated with Robin sequence can be significant, with mortality ranging from 1.7% to 65%.^{3,6,12–17}

Robin sequence is recognized as a heterogeneous condition caused by isolated mandibular hypoplasia, or by a multitude of conditions that can produce a similar phenotype. More than 50% of affected patients have an associated syndrome, or genetic or medical anomaly,^{3,18–20} the most

Fig. 1 (A) Lateral view of an infant with significant retrognathia and airway obstruction. Due to the severity of airway obstruction at the tongue base, the patient required mandibular distraction. (B) Sagittal view computed tomography of an infant with Robin sequence. Note the degree of airway obstruction at the tongue base and the cleft of the soft palate.

common of which is Stickler,²⁰ although more than 40 syndromes have been associated with Robin sequence. Due to the widely varied nature of presentation, consensus in diagnoses and management remains elusive, producing significant obstacles to formulating definitive treatment protocols.

Interventions

In cases of mild to moderate airway stenosis, obstruction can be successfully relieved with conservative interventions such as prone positioning, supplemental oxygen, nasopharyngeal tubes, and continuous positive airway pressure (CPAP).^{18,21–23} Patients affected with severe or complex pattern airway obstruction typically require invasive interventions to provide temporary or permanent relief of airway obstruction, including intubation, tracheostomy, tongue–lip adhesion (TLA), and mandibular distraction osteogenesis (MDO).

A definitive and stable airway can be directly achieved through tracheostomy, which was previously the first-line surgical treatment of infants with Robin sequence associated with severe airway obstruction. Unfortunately, several morbidities have been associated with tracheostomy, including increased risk for tracheomalacia, chronic pneumonia, laryngeal stenosis, intellectual and physical impairments, compromised social interactions, and a requirement for complex

nursing care and parental education.^{24–29} In addition, the long-term benefit of tracheostomy is questionable, as tracheostomy is associated with increased cost of medical care due to frequent emergency room visits for treatment of pneumonia.³⁰ Consequently, alternative interventions such as TLA and MDO were developed to avoid tracheostomy in infants with Robin sequence.

Tongue–lip adhesion was initially described by Shukowsky,³¹ and has been modified by several others^{21,32,33} to provide a surgical alternative to tracheostomy. Successful outcomes for TLA have been reported to be in the range of 71% to 89%.^{18,22,23,34–36} This established technique is simple to perform, but there have been reports of dehiscence of adhesion, problems with feeding, and limited efficacy.^{35,37,38} TLA is the preferred first-line form of surgical intervention in many centers for patients with Robin sequence who have failed conservative intervention.^{18,21–23,34,36} However, the success of TLA is dependent on anterior and inferior “catch-up growth” of the mandible as the definitive means of airway obstruction relief, a concept that remains debated.^{22,23,35,38,39}

After popularization of the mandibular distraction technique by McCarthy,⁴⁰ MDO was implemented in children with severe mandibular hypoplasia as a method of tracheostomy decannulation.^{37,41,42} The technique was eventually applied to neonates with severe airway obstruction secondary to Pierre Robin sequence in efforts to avoid tracheostomy.⁴³ Mandibular distraction directly corrects the horizontal deficiency of the mandible, moving the tongue forward and relieving the underlying cause of airway obstruction. The effectiveness of mandibular distraction has been demonstrated by several centers^{5,30,38,41–46}; however, critics note concerns of the complexity and morbidity of the procedure, high cost of care, device failure, nerve and tooth injury, and growth disturbance to the mandible.^{18,22,23,34,36} Many surgeons who utilize the TLA technique and attest to the mandible’s ability for catch-up growth have criticized the MDO technique as morbid and overutilized.

Despite the debate regarding the risks and benefits of MDO versus TLA in the treatment of Pierre Robin sequence, evidenced-based analysis comparing these two techniques has been lacking. A multitude of treatment algorithms of care and treatment plan recommendations have been derived,^{23,30,46,47} although the majority of this information is largely based on expert opinion.⁴⁸ Recently, a case-controlled study of infants with isolated Robin sequence demonstrated that MDO is associated with statistically superior outcomes compared with TLA with regards to incidence of tracheostomy, improvements in oxygen saturation, and improvements in apnea/hypopnea index (AHI),⁴⁹ with a complication profile comparable to that of TLA.

Feeding and Growth

The detrimental effects of mandibular hypoplasia on feeding, swallowing, and growth are well described.^{41,50–53} In patients with isolated Robin sequence, increased work of breathing can exhaust the child and prevent adequate caloric intake

during normal feeding. Cleft palate, a common finding in infants with Robin sequence, creates another layer of complexity, as oral suction is not possible and special feeding techniques and bottles are often necessary to ensure necessary alimentation. Furthermore, increased intra-abdominal pressure, a consequence of the increased work of breathing, puts infants at risk for aspiration and gastroesophageal reflux disease (GERD). It is notable that GERD has been found to be associated with failure of distraction by numerous studies.^{5,46,54} In more than 50% of cases, patients with Robin sequence have other associated anomalies, including central nervous system and neurologic abnormalities, syndromic diagnosis, and other disorders that all contribute separate and additive detriment to feeding.^{3,18–20}

In the background of this complex landscape, the effects of mandibular distraction on feeding in infants with Robin sequence have been predictably varied. Two of the most cited studies report apparently conflicting results. At one end of the spectrum, through a combined polysomnogram and esophageal pH analysis, Montesario and colleagues⁴¹ demonstrated that reflux in Robin sequence is temporally correlated with apneic episodes. In their series of 18 patients, aspiration and GERD was successfully treated in all patients following mandibular distraction. At the other end of the spectrum, Spring and colleagues⁵¹ evaluated 10 patients for growth and feeding success after distraction. Postdistraction, seven patients experienced an early decline in growth rate, and three patients experienced feeding disturbance, necessitating nasogastric feeding in two patients and gastric tube feeding in one patient. The difference in outcomes in the two studies may be associated with the difference in patient populations evaluated: In the Montesario study, all patients had isolated Robin sequence, compared with the Spring study, in which 90% of patients also had an underlying syndrome. Contributions of syndromic diagnosis to feeding was subsequently confirmed by Lidsky et al,⁵⁰ who demonstrated that patients with isolated Robin sequence who undergo early mandibular distraction do not require gastrostomy tube feedings in comparison to syndromic patients, 50% of whom require gastrostomy feedings after early mandibular distraction. Future analysis on the effects of mandibular distraction on feeding and growth should separately assess patients with isolated versus nonisolated Robin sequence.

Team Evaluation

Patients with Robin sequence are assessed for treatment by a multidisciplinary team including specialists from neonatology, genetics, otolaryngology, developmental pediatrics, pulmonology, and plastic surgery. All patients under consideration for surgical intervention undergo polysomnography evaluation. Patients who demonstrate no central sleep apnea, an AHI of 20 or greater, or who have significant CO₂ retention should be considered for mandibular distraction. We would note that these sleep study parameters are based on clinical experience rather than scientific assessment, as there are no evidence-based protocols on sleep study values

for mandible distraction indication to date. A noncontrast computed tomography (CT) scan is obtained to assess bone stock and morphology of the lower jaw and temporomandibular joint (TMJ) in preparation for surgery (–Fig. 2). A CT assessment should be performed in all Robin sequence patients prior to distraction as ankylosis or absence of the TMJ as well as inadequacy of bone stock may preclude success of mandibular distraction. Likewise, aberrations in mandibular morphology may affect the vector of the planned distraction. To detect presence of any secondary airway anomalies that may preclude the success of distraction, such as tracheomalacia, subglottic stenosis, and tracheal rings, laryngoscopy and bronchoscopy are performed prior to distraction, commonly during the same anesthetic. Currently, our team does not view laryngomalacia as a contraindication to distraction, which will be discussed later in this article.

Author's Preferred Technique

The principles of distraction require vascularized bone of adequate stock as a substrate, bone-to-bone apposition of the free bony segments, rigid fixation, and vascularized soft tissue coverage. My preference in technique is described here; however, adherence to the principles of distraction and bone generative surgery are the most important factors in the successful application of this technique.

An extraoral approach is used for direct exposure to the mandible without risk to oral contamination. A Risdon incision is placed 2 cm inferior to the mandibular border. Sharp dissection is performed to and through the platysma, and a small superior and inferior platysma flap is mobilized to assist in layered closure of the field. Wide subperiosteal exposure of the ramus, angle, distal body, and sigmoid notch is obtained. A coronoidectomy is performed with a sagittal saw or Kerrison punch. A near-complete vertical ramus osteotomy is then performed from the sigmoid notch to the antegonial notch. A full corticotomy is made over the buccal face of the mandible and along the superior and inferior aspects of the lingual surface of the mandible, leaving small bony bridge along the lingual surface of the jaw. The purpose of the near-complete osteotomy is to maintain alignment of the mandible during placement of the distractor devices. The vertical ramus osteotomy avoids injury to the infra-alveolar nerve and the developing tooth buds. Alternatively, an ultrasonic bone cutter may be used to make the described inverted L osteotomy or a mandibular body osteotomy.

In cases in which the mandibular deficiency is in the body and the morphology of the mandible is otherwise normal, a horizontal vector is used. A vertical vector has been advocated by several institutions^{38,46}; however, the deficiency in most cases of Robin sequence is in the horizontal plane. In other disease processes such as Treacher Collins syndrome, the bony deficiency is in the mandibular ramus. In these cases, a vertical vector can be considered. However, a vertical vector risks iatrogenic damage to the temporomandibular joint, either directly by placement of the device and screws too close to the condyle, or indirectly by forcing the vector of distraction into the glenoid fossa. TMJ ankylosis in the infant

Fig. 2 (A) Computed tomography with three-dimensional reconstruction of a patient with Robin sequence showing the typical pattern of horizontal deficiency of the mandibular body. This patient underwent mandibular distraction using a horizontal vector. (B) This patient has Robin sequence and a long mandibular body. The ramus, however, is deficient in vertical height and there is an inferior rotation of the mandibular body such that the ramus and body are virtually parallel. These anatomical differences affect location of the osteotomy and vector of distraction. In this case, a vertical vector was applied to the ramus. (C) This patient with Treacher Collins and Robin sequence is missing their condyle and the majority of their ramus. The coronoid is still present and can be confused with a diminutive condyle. Mandibular distraction was not offered to this patient.

is a morbid complication and a challenging clinical problem. Although incidence of this complication has been reported as high as 10%,⁴⁶ we have been successful in avoiding this complication at our institution through the implementation of a horizontal vector in virtually all Robin sequence patients who require distraction.⁵⁵

For the distraction, a 20-mm, semiburied, uniplane distraction device (Micro Zurich mandibular distractor, KLS Martin, Jacksonville, FL) with a ratchet is used in most cases. Longer devices are sometimes used in cases of extreme micrognathia or in older children. The ratchet system on the device prevents “backing” of the activation by parental error or manipulation by the infant. The device is affixed to the lower border of the mandible and the activation arm exits percutaneously behind the earlobe. To affix the footplates to the mandible, high-profile screws are used, as they are easy to apply and more easily removed compared with 1-mm screws. When using a ratchet system distractor, high-profile screws can impinge on the ratchet during full activation; therefore, the placement and type of screws used should be carefully considered.

Once the device is secured to the mandible, the distractor is activated for several millimeters. There should be no resistance on the device. The osteotomy on the lingual surface may be completed by activation of the device alone. Completion of mandibular osteotomy should be confirmed by palpation using an osteotome or a suction cannula. Direct vision will not suffice for confirming that the osteotomy is complete. One can separate the bony fragments several millimeters in a bone that has not been completely transected. What has been previously reported as early consolidation during activation is more likely an incomplete osteotomy. Once a full osteotomy is confirmed, the bony edges are opposed without any intervening soft tissue, and the Risdon incision is closed in layers.

After a latency period of 5 days, activation commences at a rate of 1 mm/d to the maximal allowable length of the distraction device (20 mm). Serial cephalograms are used to monitor progress and confirm symmetrical bilateral advancement. Devices are removed in a second operation after 8 weeks of consolidation. At the time of device removal, integrity of the generated bone is assessed by direct visualization and palpation.

Once mandibular distraction is complete, the patient is followed by a plastic surgeon and pulmonologist to monitor mandibular development and recurrence of obstructive breathing patterns. A follow-up polysomnogram is obtained within 3 months of distractor removal. As sleep study analysis can identify and quantify airway obstruction, it is an important tool to assess airway function in these patients.⁵⁶

Effectiveness

The effectiveness of mandibular distraction has been reported to be 92 and 100%.^{5,38,41,44,46,57,58} Patient characteristics associated with failure of distraction include GERD, need for Nissen fundoplication, and absence of a cleft palate.⁵ The association of failure of distraction with presence of GERD

and need for Nissen fundoplication suggests that these patients have severe upper airway obstruction due to increased respiratory effort.⁴¹ This is associated with increased intrathoracic negative pressure, leading to the use of abdominal muscles that increase intragastric pressure causing immaturity of the esophageal sphincter. Although prior studies have demonstrated that early mandibular distraction can improve feeding and resolve aspiration,⁴¹ there may be significant damage to the respiratory tract caused by persistent neonatal reflux, which can lead to increased work of breathing, hypoxia, and pneumonia. This specific patient population may benefit from early diagnosis and intervention of GERD to improve respiratory outcomes.

Complications

The complication profile associated with mandibular distraction has been previously defined by Davidson et al.⁵⁹ Major complications are defined as events that result in an adverse outcome and that cannot be resolved with invasive therapy, such as TMJ ankylosis. Moderate incidents are those that can result in adverse outcomes and be resolved with invasive therapy. Minor incidents are events that do not lead to adverse outcomes and can be resolved with noninvasive therapy. The average complication rate of mandibular distraction as performed on the infant population for the treatment of Robin sequence is 34%, with the majority of complications described as minor or moderate incidents rather than major incidents (► **Table 1**).⁵⁵ The most common of these complications include surgical site infection (the majority of which are successfully treated with antibiotics alone), neurapraxia, unfavorable scars, dental injury, device failure, and nerve injury. Compared with this procedure performed in non-infants,⁵⁹ this complication rate is favorable, indicating that mandibular distraction may be safer when performed in infants versus older children and adults.

Laryngomalacia

A controversial topic of mandibular distraction in the Robin sequence population relates to contraindications to care. Laryngomalacia has historically been identified as a concurrent diagnosis that would preclude successful distraction.^{37,46,60}

However, critical analysis of the literature demonstrates that this recommendation has been made without any empiric clinical evidence.

Our experiences demonstrate that laryngomalacia, the most common cause of stridor in infancy, is a common concurrent diagnosis (up to 23%) in patients with Robin sequence. In cases in which laryngomalacia and tongue-based airway obstruction are present without any other airway anomaly, mandibular distraction is considered. Patients are assessed by a pediatric otolaryngologist, and supraglottoplasty is performed as needed. To date, 11 patients with Robin sequence and laryngomalacia have been treated at our institution. Ten patients successfully avoided tracheostomy, and one patient with a preintervention tracheostomy was successfully decannulated.

Our experiences raise many unanswered questions regarding the utility of mandibular distraction in this subset of patients with Robin sequence. It is certainly possible that some of these patients may have been successfully treated by supraglottoplasty alone. In these patients, laryngomalacia was often diagnosed the day of surgery for mandibular distraction by laryngoscopy. If traditional protocols were followed, these patients would not have been committed to jaw lengthening, but rather to tracheostomy, which is associated with significant morbidity^{24,25,27–29,61} and an increased economic burden of care.³⁰ Although the efficacy of mandibular distraction in this subset of patients with Robin sequence and laryngomalacia is far from proven, we strongly believe that patients with these concurrent diagnoses should not be routinely committed to tracheostomy; rather, both disease entities should be evaluated and treated for successful alleviation of airway-based obstruction.

Cleft Palate Repair

Cleft palate reconstruction after mandibular distraction is a topic that has had little attention in the literature. Unfortunately, patients with Robin sequence have a high incidence of obstructive sleep apnea⁶² and airway complications after cleft palate repair (22% to 47%).^{63–66} Many centers advocate delaying cleft palate repair in the Robin sequence population until 18 months of age to limit risk to postoperative airway complications. Our unit follows an airway-assessment

Table 1 Incidence of Complications Associated with Neonatal Mandibular Distraction

Total Incidence: 34%		
Minor (15%)	Moderate (15%)	Major (9%)
SSI (treated with antibiotics)	SSI (requiring surgery)	Dental injury
Facial neurapraxia	Device fracture	Residual anterior open bite
Self-extubation	Widened neck scars	TMJ ankyloses
Hypertrophic scar	Device exposure	Death
Device loosening	Incomplete osteotomy	
	Bilateral odontomas	

Abbreviations: SSI, surgical site infection; TMJ, temporomandibular joint.

protocol prior to cleft palate repair in which polysomnography is used to screen all patients with Robin sequence prior to cleft palate repair. Those infants with an AHI of > 5 have their palatoplasty delayed and other interventions are considered, including adenoidectomy/tonsillectomy or repeat mandibular distraction. Using this protocol, we have decreased our airway complication rate to 7%, a comparable incidence to patients with isolated cleft palate who undergo palatoplasty.

Future Directions

The ideal goal for surgical therapy is toward protocol-driven care in which standardized, validated guidelines are used to drive surgical indications. Future studies in the surgical treatment of Robin sequence should be conceptualized with this endpoint in mind. The following will be important topics to be addressed in future research:

1. **Definitions:** One of the most important steps in treating a complex disease process is defining the pathology. To date, no consensus has been reached with regards to the definition of Robin sequence. Although the triad of the micrognathia, glossoptosis, and airway obstruction was Robin's original description of the sequence, many practitioners substitute airway obstruction with cleft palate in their definition of the disease. More concerning, some practitioners and researchers define Robin sequence as a child with two of the three characteristics of Robin's triad, with or without cleft palate, confounding any efforts to clearly define this complex patient population. Furthermore, there have been attempts by others to further subdivide Robin sequence patients to syndromic/nonsyndromic or isolated/nonisolated. Until agreed-upon definitions are created, accepted, and followed, research in this patient population will be plagued by inconsistency and inaccuracy, limiting progress of care. Robin's original description should be the strict definition of the triad (micrognathia, glossoptosis, and airway obstruction).
2. **Evidence-Based Decisions:** Although many articles have been published on the surgical treatment of Robin sequence, the majority of these studies are in the form of retrospective, noncomparative review, with conclusions largely based on expert opinion.^{22,23,30,46,47} There is an overall lack of clinical comparative data and virtually no prospective trials regarding the surgical treatment of Robin sequence.⁴⁸ A greater emphasis on the production of level I–III clinical studies will benefit the future formulation of evidence-based decisions.
3. **Quantifiable Metrics:** To produce reliable scientific data, a quantifiable metric will be required to compare results between centers. Although avoidance of tracheostomy is an important endpoint, it is a binary metric that is an uncommon occurrence after mandibular distraction. A better metric would be the polysomnogram results, which quantifies the severity of airway obstruction. Although sleep studies are subject to interpretation, it remains the most widely used and reliable tool to measure obstructive breathing patterns in this patient population.⁵⁶ The poly-

somnogram should be a standard measurement device to assess the severity and resolution of airway obstruction.

4. **Outcomes Analysis:** Mandibular distraction has been demonstrated by multiple studies to be an effective means of relieving airway obstruction in appropriately selected infants with Robin sequence. Unanswered questions remain regarding the long-term function of patients after jaw lengthening. Long-term data on jaw and tooth development, growth, neurocognitive outcomes, and need for further surgery is generally lacking and should be addressed in future studies.

Conclusions

Mandibular distraction is a safe and effective intervention for neonates with Robin sequence who experience severe airway obstruction. The construction of validated protocols of evaluation and intervention will be critical for standardizing care and improving patient outcomes. A clear definition of Robin sequence, prospective and comparative scientific research, standard use of polysomnography, and long-term outcomes analysis should be strongly considered in future research.

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