

Neonatal Mandibular Distraction Osteogenesis: A Technique for Treating Neonates with Micrognathia and Risk of Respiratory Obstruction

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ABSTRACT

Small lower jaw may be a reason of upper respiratory obstruction in neonates. Neonatal upper airway obstruction is a life threatening condition often requiring urgent interference. Typically, the treating neonatologist first stabilizes the patient's airway via intubation, a nasal airway or proper patient positioning.

Although the tracheostomy is a life-saving procedure in these circumstances, it is associated with numerous complications.

Distraction osteogenesis (DOG) offers an alternative to a tracheostomy for newborns with Pierre Robin sequence, Stickler syndrome, Treacher Collins syndrome, Nager syndrome, and other craniofacial deformities.

The study included 9 patients had micrognathia and upper respiratory obstruction who were managed at Mattel's children hospital at UCLA over the period from 2006 to 2010, 4 males and 5 females, all were full term babies except for one who was born at 33 weeks. All patients had distraction done in the first two months of life.

The diagnosis of airway obstruction was based on oxygen desaturations and duskiness and apnea on feeding. Tongue base collapse was confirmed as the primary site of obstruction by flexible nasolaryngoscopy and direct laryngobronchoscopy and other airway lesions were excluded.

In summary, bilateral distraction osteogenesis is an effective technique of elongating the micrognathic mandibles. Additional studies are indicated to study the long-term outcomes in different diagnostic subgroups of micrognathia with respiratory obstruction.

INTRODUCTION

Small lower jaw may be a reason of upper respiratory obstruction in neonates. Neonatal upper airway obstruction is a life threatening condition often requiring urgent interference. Typically, the treating neonatologist first stabilizes the patient's airway via intubation, a nasal airway or proper

patient positioning. When necessary, the definitive management of upper airway obstruction in these neonates has traditionally been to place a tracheostomy. Although the tracheostomy is a life-saving procedure in these circumstances, it is associated with numerous complications [1]. These include pneumonia, tracheitis, laryngomalacia, subglottic stenosis, bleeding from granulation tissue and death. In addition, long term problems such as delayed speech, suboptimal growth, and developmental delay are common. Removal of tracheostomies placed or upper airway obstruction in the newborn often requires multiple procedures and may not be accomplished until 5-11 years of age [2].

Given these drawbacks, other modalities have been sought to manage neonatal upper airway obstruction. For newborns with Pierre Robin sequence, Stikler syndrome, Treacher Collins syndrome, Nager syndrome, and other craniofacial deformities, distraction osteogenesis (DOG) offers an alternative to a tracheostomy [3]. Distraction osteogenesis is a surgical technique involving the gradual lengthening of bones and has become an accepted procedure in the treatment of craniofacial anomalies in the growing patient [4]. Several series have documented lower jaw lengthening over a one-week period for newborns with micrognathia [3,5]. This successfully ameliorates upper airway obstruction by advancing the tongue and epiglottis [6].

The natural history of mandibular growth in syndromes with micrognathia is not clear. With isolated Pierre Robin sequence (not a syndrome), children can reach normal mandibular projection by six months of age. Many authors think this "catch-up" growth is a result of stimulation of the

mandible by continuous tongue thrusting and neurological development of oral motor skills. However, a subset of these children does not outgrow their micrognathia. The only situation where catch-up growth would be expected is when the mandible is intrinsically normal but positionally constricted in utero. Infants with Stickler syndrome have been reported to show minimal mandibular growth. They have intrinsic mandibular hypoplasia with a concave depression in the body of the mandible (antegonial notching) and a shortened ramus as a primary feature. In syndromes such as Treacher-Collins and Nager syndromes, micrognathia and retrognathia are always persistent and do not self-resolve over time [7].

We conducted this study to evaluate the outcomes of distraction osteogenesis in neonates who were born with micrognathia and glossoptosis and respiratory obstruction whether they have isolated Pierre Robin sequence or syndromic Pierre Robin sequence.

PATIENTS AND METHODS

This study is a combined retrospective and prospective, it includes 9 patients with micrognathia and upper respiratory obstruction who were managed at Mattel's children hospital at UCLA over the period from 2006 to 2010. The group of patients studies did not include any patient who already had a tracheostomy placed at an outside hospital or those patients with micrognathia who had no report of problems with airway obstruction and patients older than two months of age (exclusion criteria). Admission to the NICU was part of the criteria; so many patients with Pierre Robin sequence evaluated as an outpatient at the craniofacial clinic (some treated with tracheostomy or distraction osteogenesis) were not included in the study. Thus, the patients requiring admission to the NICU or PICU represented a more severe form of upper airway obstruction.

Specialists, including a plastic surgeon, otolaryngologist, anesthesiologist, geneticist, pulmonologist, and gastroenterologist, were consulted. Patients were excluded for mandibular distraction osteogenesis (DOG) and offered tracheostomy if they had: 1) central apnea, 2) severe gastrointestinal reflux, or 3) other airway lesions. The remaining patients were considered appropriate candidates for mandibular distraction, and were offered the option of having mandibular DOG or tracheostomy. All parents chose mandibular DOG over tracheostomy.

The information obtained for each patient included the following: Demographic data; type of distraction device used, preoperative and postoperative respiratory status, feeding status and weight gain, sleep problems, duration of active distraction and consolidation, and length of postoperative follow-up. Postoperative follow-up included clinical evaluation, lateral cephalograms and endoscopic visualization of the upper airway.

Osteotomy and device placement:

Under general anesthesia, a combined intraoral and extraoral Risdon approaches were used (Fig. 1).

The inverted L osteotomy is generally preferred over the vertical ramal osteotomy because the advancement of the coronoid process with a vertical ramal osteotomy may cause impingement into the zygoma (Fig. 2). Furthermore, the temporalis muscle remains with the proximal segment and does not counter the distraction with relapsing forces (Fig. 3).

After completing osteotomy, the microdistractor (KLS-Martin, Jacksonville, FL) was fixed with its rod placed at an oblique vector to gain ramal height and body length (Fig. 4).

Postoperatively, nasotracheal intubation and sedation was maintained in the neonatal intensive care unit for the duration of the distraction. Once distraction was completed, patients were brought to the operating room for removal of the turning arms of the distractors and extubation. Airway patency was confirmed by bronchoscopy at the time of extubation. Infants typically started oral feeds the next day and discharged home shortly afterwards.

Radiographic analysis:

Mandibular lengthening and ossification were confirmed by a series of lateral skull films taken preoperatively, immediately after microdistractor placement, at completion of distraction, after microdistractor removal, and at six-months follow-up. Landmarks of analysis included sella (S), nasion (N), orbitale (O), anterior nasal spine (ANS), upper incisor edge (UIE), lower incisor edge (LIE), B point (B), pogonion (Pg), gonion (Go), menton (M), and articulare (Ar). CT scans were also performed on the patients preoperatively and after microdistractor removal.

Outcomes analysis:

All patients were assessed for feeding and breathing conditions, long-term morbidity, growth

and speech. Morbidity and mortality were documented by a thorough search of each patient's medical chart and electronic record with careful review of inpatient records, emergency department visits, routine pediatric care, and all follow-ups with pediatricians, surgeons and craniofacial team meetings. The specific growth parameters examined at final follow-up were height and weight percentiles (determined by standard sex-specific growth charts).

Evaluations were performed every 6 months at craniofacial clinic by pediatricians, geneticists, speech pathologists, plastic surgeons, audiologists, pediatric dentists, orthodontists, and social workers. Age at cleft palate repair was also noted for those patients with cleft deformities in addition to upper airway obstruction.

RESULTS

The study included 9 patients, 4 males and 5 females, all were full term babies except for one who was born at 33 weeks. All patients had distraction done in the first two months of life.

The diagnosis of airway obstruction was based on oxygen desaturations and duskiness and apnea on feeding. Tongue base collapse was confirmed as the primary site of obstruction by flexible nasolaryngoscopy and direct laryngobronchoscopy and other airway lesions were excluded.

All patients had isolated cleft palate. Cleft palate repair was performed at mean age 12 months. After closure of the palate, all patients continued to feed and breathe well (Figs. 5,6,7).

Table (1): Status of pre- and post-operative feeding and growth.

Name Group A	Preoperative feeding status	Postoperative feeding	Preoperative growth	Postoperative growth and development
CR	Gavage feeding	Normal oral feeding	Inadequate weight gain	Normal
GR	Gavage feeding	Normal oral feeding	Inadequate weight gain	Normal
DL	Gavage feeding	Normal oral feeding	Inadequate weight gain	Normal
DB	Gavage feeding	Normal oral feeding	Within normal growth percentiles	Normal
KP	Gavage feeding	Normal oral feeding	No weight gain since birth	Normal
AM	Bottle feeding with Haberman feeder	Normal oral feeding	No weight gain, failure to thrive	Normal
GM	Limited feeding with Haberman feeder	Normal oral feeding	Inadequate weight gain	Normal
JE	Gavage feeding	Normal oral feeding		Normal
VA	Limited feeding with Haberman feeder	Normal oral feeding	Inadequate weight gain	Normal
JL	GT feeding	Normal oral feeding	Inadequate weight gain	Normal

Table (2): Preoperative and postoperative respiratory conditions of the patients.

Name Group A	Preoperative respiratory status	Postoperative respiratory status
CR	Desaturations in supine and sideways, NICU+O2	Stable on room air, normal feeding and sleepin gwithout desaturations
GR	Frequent desaturations, intubated	Stable on room air, normal feeding and sleepin gwithout desaturations
DL	Desaturations mainly with feedings, NICU+O2	Stable on room air, normal feeding and sleepin gwithout desaturations
DB	Frequent desaturations, intubated	Stable on room air, normal feeding and sleepin gwithout desaturations
KP	Desaturations in supine and sideqays, NICU+O2	Stable on room air, normal feeding and sleepin gwithout desaturations
AM	Desaturation with feeding, NICU, occasoional O2	Stable on room air, normal feeding and sleepin gwithout desaturations
GM	Desaturations in supine position, home management	Stable on room air, normal feeding and sleepin gwithout desaturations
JE	Desaturations in supine, sideways and feeding, NICU+O2	Stable on room air, normal feeding and sleepin gwithout desaturations
VA	Desaturations in supine and sideways, NICU+O2	Stable on room air, normal feeding and sleepin gwithout desaturations

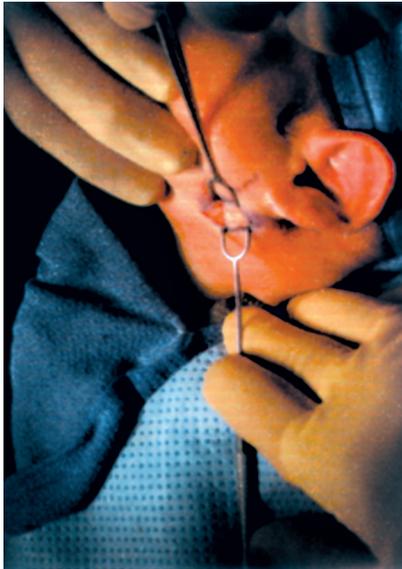


Fig. (1): Risdon incision.



Fig. (2): Marking inverted L osteotomy with bovie.



Fig. (3): Inverted L osteotomy.



Fig. (4): The distractor in place in the zero position. In this case intraoral incision was avoided but Risdon incision is longer and a minimal incision behind the ear lobe was used to bring out the turning arms.



(A)



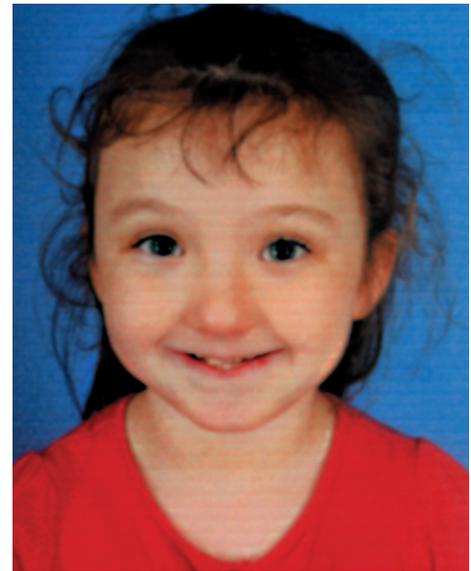
(B)

Fig. (5-A,B): (Left) Right lateral preoperative view. (Right) Right lateral postoperative view of patient with Pierre Robin syndrome.

Fig. (6-A,B): Frontal pre-operative and late postoperative (3 years) views of a patient with micrognathia and upper airway obstruction, iPRS, treated with distraction osteogenesis.



(A)

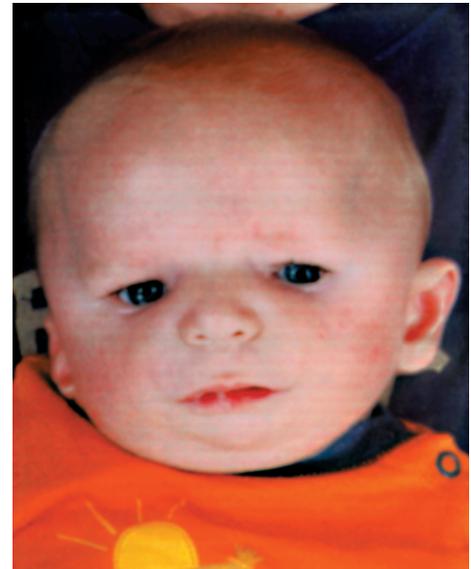


(B)

Fig. (7-A,B,C,D): (Above, left) Frontal preoperative view of a newborn with Treacher Collins syndrome treated with distraction osteogenesis. (Above, right) Frontal postoperative view 2 months after the distraction procedure. (Below, left) Right lateral preoperative view (Below, right) Right lateral postoperative view.



(A)



(B)



(C)



(D)

Complications:

In the present study, two complications occurred in two patients: Cheek abscess requiring incision and drainage and Marginal mandibular nerve weakness.

Outcome assessment:

Bilateral mandibular distraction osteogenesis has resulted in avoidance of tracheostomy, complete elimination of obstructive airway symptoms, and correction of micrognathia in 9 cases. Average patient follow-up was 49 months. Moderate retrognathia was noticed in one patient and a slight asymmetry in another patient.

DISCUSSION

Since McCarthy et al. [8] used DOG for the reconstruction of the deficient mandible in children, the technique has become increasingly accepted as a management option for micrognathia, as evidenced by recent series reported by Denny and Kalantarian [5] and Sidman et al. [9].

However, because the technique is of relatively recent clinical application, little has been written on the results of bilateral mandibular distraction. This study was designed to examine the results of bilateral mandibular distraction in patients with Pierre Robin sequence. In this study, only patients with severe upper airway obstruction facing tracheostomy were involved. End point of distraction was based on relief of the upper airway obstruction and overcorrection was attempted in all cases. With the patient intubated, immediately after distractor placement the patient is able to rest with appropriate airway and nutritional support while the distraction progresses.

In our protocol, distraction starts on the first postoperative day and continues at a rate of 1.8mm per day. The change in latency does not alter the properties of the regenerated bone in mandibular distraction osteogenesis. Indeed, no latent interval may be necessary in craniofacial distraction [10]. The age of the patient plays a role with bone healing in neonates being reported to occur twice as fast as in adults [11]. This difference has been attributed to the larger pool of undifferentiated mesenchymal cells, which are capable of forming osteoblasts at a more rapid rate in skeletally immature neonates [12]. In general, the tendency is to use shorter latency periods and more rapid distraction in very young patients to avoid premature consolidation of the regenerate. Mandibular distraction rate of 2mm/day can be used in those young patients [4].

The age of the patients may also be a factor for shorter consolidation period, as younger patients such as infants have a better healing potential. 6- to 8-week consolidation period is most commonly used in mandibular distraction osteogenesis. A longer consolidation period is beneficial in terms of allowing adequate maturation of the bony callus, but the lengthy consolidation period is associated with morbidity [12]. In this study, the mean consolidation period was 80.2 days.

Bilateral mandibular distraction osteogenesis resulted in avoidance of tracheostomy, complete elimination of obstructive airway symptoms and correction of micrognathia in all patients.

The natural history of mandibular growth in syndromes with micrognathia is not clear. With isolated Pierre Robin sequence (not a syndrome), children can reach normal mandibular projection by six months of age. Many authors think this "catch-up" growth is a result of stimulation of the mandible by continuous tongue thrusting and neurological development of oral motor skills. However, a subset of these children does not outgrow their micrognathia. The only situation where catch-up growth would be expected is if the mandible is intrinsically normal but positionally constricted in utero.

Failure to thrive is common in Pierre Robin sequence (PRS) children because they expend a great deal of energy to breathe against an obstructed airway. Therefore, their energy reserves are depleted for other facets of life such as eating and even growth and development [14]. Difficulty breathing can also lead to incoordination of suckling and swallowing, which contributes to eating problems [15]. Providing the neonate with a functional airway can result in dramatic improvements in feeding. Just as retroposition of the tongue base caused by micrognathia causes airway obstruction, the abnormal structural position also hinders the swallowing mechanism. By repositioning the chin point anteriorly, the tongue base consistently follows and is advanced out of the airway, thus correcting the airway obstruction. As an integral part of this process, the tongue progresses from a vertical orientation to a horizontal position, and the effectiveness of swallowing is greatly improved [5].

In this study, following distraction, all patients returned to an oral diet postoperatively. All patients were running in the average percentiles for their growth and attained good development.

Cleft palate was found in all patients. The mean age at palatoplasty was 12 months. Patients con-

tinued to do well after palatoplasty with no obstructive airway symptoms.

In summary, bilateral distraction osteogenesis is an effective technique of elongating the micrognathic mandibles. Additional studies are indicated to study the long-term outcomes in different diagnostic subgroups of micrognathia with respiratory obstruction. Such studies should provide treatment alternatives to improve long-term results.

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