Mandibular Distraction Osteogenesis to Relieve Pierre Robin Severe Airway Obstruction in Neonates: Indication and Operation

Weimin Shen, MD, Cui Jie, MD, Jianbing Chen, MD, Jijun Zou, MD, and Yi Ji, MD

Abstract: Airway management in neonates with Pierre Robin sequence is often challenging. Advancement of the tongue base using mandibular distraction in neonates can avoid tracheotomy and is increasingly propagated. The surgical indication is very important. We establish indication. The result of the preoperative peripheral oxygen saturation of about 40% in the prone position is considered as indication. The distance from postpharyngeal wall to lingual root of more than 5 mm is considered as contraindication; however, distance of less than 2 mm is considered needing performing mandibular distraction in preoperative lateral cephalometric radiograph. A three-dimensional computed tomographic demonstrating severity of retrognathia was generated preoperatively in the computed tomography room. The osteotomies can be performed by extraoral approaches. Nowadays, for precise bone cutting, Surgybone (Silfradent Srl, Italy) devices have been introduced in maxillofacial surgery, which we used for extraoral-approach osteotomies in 6 patients presented in this study. Distraction was performed after bilateral oblique osteotomy and removed after 1-month fixation in the second stage. A follow-up examination after removal of the distractors was performed. Normal appearance and no apnea were observed at 6 months' follow-up.

Key Words: Pierre Robin sequence, mandibular distraction, neonates

Patients
This study reviews 6 consecutive neonates diagnosed with PRS and life-threatening upper airway obstruction from January 1, 2007, through December 30, 2008. A multispecialty neonatal obstructive airway team was formed at the Children's Hospital of Nanjing; the team included a neonatal intensive care unit neonatologist, a pediatric anesthesiologist, a pediatric otolaryngologist, and a pediatric plastic surgeon to assess these patients. The study included 5 neonatal patients either seen primarily at the Nanjing Children's Hospital and 1 neonatal patient referred from other institutions for airway management. Traditional management failed in all these patients, including prone positioning and nasopharyngeal airway intubation. Six neonatal patients have the clinical manifestations of mandibular micrognathia, glossoptosis, and cleft palate formation (Fig. 1). Patient data are listed for 6 patients in Table 1.

Preoperative Management
All management and outcomes for the respiratory distress were recorded for each neonate. Perinatal feeding history and presence of any other congenital anomalies were also recorded. In the management, top priority was assigned to stabilization of the airway. Airway stabilization proceeded from prone positioning with chest roll, to nasopharyngeal airway, to tracheal intubation (either nasal or oral, depending on ease or difficulty). Adequate nutrition was achieved via nasogastric gavage feeding. If the airway could not be stabilized by positioning or insertion of a soft trumpet via the nose into the nasopharynx, surgical management (mandibular DO and the tongue-lip adhesion) was considered. The decision to perform surgery was based on the patient's clinical status, perceived need for long-term respiratory support, result of preoperative peripheral oxygen saturation of approximately 40% in the prone position, failure of nonoperative management, and the surgeon's clinical judgment. A three-dimensional computed tomographic (CT) scan of the head (Fig. 1) was obtained before distraction for surgical planning to define the mandibular anatomy and, specifically, to identify the inferior alveolar nerve foramina on the lingual cortices of the mandibular rami and distal tooth buds in the mandibular bodies, and lateral cephalogram in prone position was obtained. The distance was measured from postpharyngeal wall to lingual root. The distance from postpharyngeal wall to lingual root was listed for 6 patients (Table 2). Decision of mandibular distraction was made through clinical symptom and the distance from postpharyngeal wall to lingual root. The distance from postpharyngeal wall to lingual with neonatal tracheostomy, distraction osteogenesis (DO) is an alternative treatment. In DO, bone is gradually lengthened under tension across a surgically created osteotomy. We describe bilateral mandibular DO for 6 cases with PRS and severe airway obstruction.

MATERIALS AND METHODS

Indication and Operation
We establish indication. The result of the preoperative peripheral oxygen saturation of about 40% in the prone position is considered as indication. The distance from postpharyngeal wall to lingual root of more than 5 mm is considered as contraindication; however, distance of less than 2 mm is considered needing performing mandibular distraction in preoperative lateral cephalometric radiograph. A three-dimensional computed tomographic demonstrating severity of retrognathia was generated preoperatively in the computed tomography room. The osteotomies can be performed by extraoral approaches. Nowadays, for precise bone cutting, Surgybone (Silfradent Srl, Italy) devices have been introduced in maxillofacial surgery, which we used for extraoral-approach osteotomies in 6 patients presented in this study. Distraction was performed after bilateral oblique osteotomy and removed after 1-month fixation in the second stage. A follow-up examination after removal of the distractors was performed. Normal appearance and no apnea were observed at 6 months' follow-up.

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root of less than 3 mm is considered needing performing mandibular distraction in preoperative lateral cephalometric radiograph.

**Operation**

The operations were performed under general anesthesia using a percutaneous inframandibular border approach. The skin incision is then placed symmetrically approximately 15 to 20 mm inferior to this area.

The skin is injected with 0.5% lidocaine hydrochloride with 1:200,000 epinephrine. Once the incision is made, the subcutaneous fat is then swept in an inferior-to-superior fashion with a periosteal elevator. Dissection should be performed deep to the superficial layer of the deep cervical fascia because the marginal mandibular nerve lies within or just deep to this layer and passes superficial to the facial vasculature. Dissection is continued until the pterygomasseteric sling along the posterior and inferior borders of the mandible is identified. Gentle, blunt dissection is performed until the muscle fibers of the pterygomasseteric sling remain, which minimizes the risk of injury to the marginal mandibular branch. The pterygomasseteric sling is excised with needlepoint cautery along its inferior edge to expose the buccal cortex of the mandible. Subperiosteal dissections are then performed along the buccal and lingual cortices. The osteotomy line is outlined on the bone with a No. 2 pencil. The oblique osteotomy is planned while maintaining adequate bony segments on either side of the osteotomy to allow good footplate fixation (Fig. 3). We perform osteotomy with surgybone devices to achieve clean bony cuts with minimal bone loss or heat generation. A side-biting cutting burr can be easier to control; however, this method sometimes results in bone loss at the osteotomy site. Irrigation of surgybone devices should be used copiously during osteotomy because thermal injury can impair reparative callus formation, thereby inhibiting the distraction process. Unidirectional internal microdistractors (CibeI Inc) with 1-mm raised-head self-tapping, self-drilling screws are used for most distractions in neonates (Fig. 4). The raised-head screws allow easy placement and removal. We have found bone growth over the flat-head screws at the time of distractor removal. Predrilling the mandible with a pilot hole is not required in patients at this age and may result in poor screw fixation. We have used a variety of distractors for mandibular DO in children.

We prefer the oblique osteotomy. The oblique osteotomy begins from the anterior ramal border to the posterior border of the mandibular angle. Next, buccal corticotomy is performed, except at the inferior border, where it is bicortical. The distraction device is then temporarily fitted into the operative site on the basis of a predetermined vector, and its footplates are tailored to accommodate it. A puncture site is made in the postauricular area. A hemostat is then passed through this puncture site into the wound to grasp the activating arm of the distractor device, and the activator is pulled through the puncture site. Once the distractor is secured, the zygoctomoty is converted to an osteotomy with a 2-mm osteotome. The device is then activated, and the mandibular segments are checked to see that they are easily distracted. Failure to move the segments easily signifies incomplete osteotomy, and the site must be inspected, and any remaining bony bridges osteotomized, which is sometimes easier to accomplish with the mandible under tension. Once the segments distract easily, the distractor is closed down, and the segments are brought back in close approximation within a 1-mm gap (Fig. 5). The incision is then packed off, and the contralateral device is placed in a similar fashion. The wound is irrigated

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copiously with antibiotic solution. The pterygomasseteric sling is reconstructed with a polyglactin 4-0 (Vicryl; Ethicon Inc) suture. Restoring the sling is important in functional loading and remodeling of the mandible. The dermis is approximated with a polyglactin 4-0 (Vicryl; Ethicon Inc) suture, and skin is closed with a running subcutaneous 6-0 polypropylene (Prolene; Ethicon Inc) suture. Two weeks after surgery, 1 end of the subcutaneous polypropylene suture is cut and removed (Fig. 6).

**RESULTS**

This group was composed of 6 neonates with isolated PRS. Six neonates required intubation and ventilator support before operation to maintain ventilation. All patients had intermittent resting oxygen saturation levels of less than 40% before operation. Further deterioration was noted during feeding in some patients. Age at operation was 7 to 21 days (mean, 13.14 days; median, 6.5 days). One neonate (patient 2) had severe growth retardation when initially evaluated and underwent nasogastric feeding support until he weighed 3.5 kg. In 1 case, a tongue-lip adhesion was performed. Mandibular distraction was performed after the failure of tongue-lip adhesion management. Mandibular distractors were placed during operation with the patient under general anesthesia, and a surgical separation was created in the ramus of the mandible. All procedures were completed in less than 2.5 hours, with less than 40 mL of blood loss. The patients were kept intubated, and distraction was initiated on postoperative day 1 at a rate of 1.2 mm/d, then switched to a rate of 1 mm/d after 6 mm of distraction. We normally use a 60-mm distractor and distract either to the full length of the 20 mm or until the mandibular gum line is 0 to 2 mm in front of the maxillary gum line, whichever occurs first. The overcorrection is performed to compensate for the regenerative contraction that can occur. We use a 4-week consolidation period before distractor removal, which is performed through the previous incision with a small posterior extension to account for the length added to the distractor. All cases were not complications. All patients were extubated within 4 to 6 days of distractor activation. A good clinical indicator of successful distraction was correction of the tongue from the initial vertical to a physiologically normal horizontal posture on physical examination. Nasoendoscopy was performed with the patients under sedation to confirm the relief of tongue-base obstruction before the patients were extubated. None of the patients needed any form of supplemental oxygenation beyond 20 days after operation. None of the patients who underwent distraction needed any additional airway support, treatment, or surgery. Nasogastric gavage feeding was started on the first day after surgery. All neonates were feeding 100% orally at 1 month of age (as in patient 6 in Fig. 7).

**DISCUSSION**

Pierre Robin,1 a French stomatologist, first reported the association of micrognathia, glossoptosis, and cleft palate in 1923. By 1974, the triad was known as the Pierre Robin sequence, which indicates that it is a condition that includes a series of anomalies caused by a cascade of events initiated by a single malformation. The primary malformation in PRS is thought to be mandibular micrognathia, which causes glossoptosis and cleft palate formation.4 There are various options for airway management for a patient with PRS. It is reasonable to begin with the most conservative measures, such as placing a nasopharyngeal airway and keeping the child in a prone position. Positive-pressure mask ventilation of the airway is also beneficial. Other options include glossopexy procedures, tongue-lip adhesions, or subperiosteal release of the floor of the mouth combined with glossopexy.5 The literature suggests that the tongue-lip adhesion could successfully relieve airway obstruction that is unresponsive to positioning alone in most patients with PRS.6,7 However, complications include dehiscence, infection, submaxillary duct obstruction, and lip scarring. Denny et al8 indicate that long-term follow-ups show a high incidence of secondary intervention requirements. For the most severe cases, tracheostomy is used to secure a stable airway. Tracheostomy in infancy carries a significant burden. The mortality rate from the tracheostomy alone independent of the underlying diagnosis is as high as 5%.9 However, because of the high complication rate in neonatal tracheostomy,

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**TABLE 2. Distance from Postpharyngeal Wall to Lingual Root in Lateral Cephalogram (Fig. 2)**

**FIGURE 2.** Distance from postpharyngeal wall to lingual root in lateral cephalogram before (A) and after (B) operation of the sixth patient.
other innovative surgical procedures have been recently applied. Mandibular distraction is an effective way to treat PRS patients and avoids a tracheostomy in 90% to 95%. Mandibular DO to treat airway obstruction from micrognathia in PRS offers many advantages over other surgical options. After osteotomy, DO is used to induce new bone formation between bony surfaces that are gradually separated. In 1973, Snyder et al reported the first experimental craniofacial application of DO in a canine model. The first clinical report of craniofacial DO was published in 1992 by McCarthy et al. They described gradual mandibular elongation in patients with congenital hypoplasia. There have been recent reports of successful DO for airway obstruction in the management of children with micrognathia. Sidman et al prospectively evaluated 11 children with tongue-base airway obstruction who were treated with mandibular DO. Decannulation or extubation was achieved in all cases. Similarly, Monasterio et al successfully used mandibular DO in 15 pediatric patients with airway obstruction. Denny and Kalantarian applied mandibular DO in 5 neonates, and tracheotomy was avoided in all cases. Mandell et al reported an 88% success rate in 8 children with PRS treated with DO. More recently, Roy and Patel described 8 children with PRS who were successfully treated with mandibular DO using the same internal microdistractor that was used by our team. These studies show that DO is successfully used to achieve mandibular advancement. With this technique, the genioglossus and geniohyoid muscles and the tongue base are brought forward, and the supraglottic airway obstruction is relieved in patients with micrognathia.

In neonates with PRS, several surgical procedures have been described to manage the airway. Especially, tongue-lip adhesion has been advocated to relieve the airway obstruction and is propagated by Danielle and Jeffrey as first-line treatment of severe airway obstruction associated with PRS. In this article, the authors emphasized indication of operation. The surgical indication is very important. The result of the preoperative peripheral oxygen saturation of about 40% in the prone position is considered as the indication. The distance from postpharyngeal wall to lingual root above 5 mm is considered as contraindication; however, less than 2 mm is considered needing performing mandibular distraction in preoperative lateral cephalometric radiograph. Less than 2 mm is considered needing performing tracheostomy. Mandibular DO avoids a tracheostomy. The reasons for the indications of mandibular DO, as we chose, were that the patients could not improve respiration when their oxygen saturation as measured by pulse oximetry was less than 40% in the prone position. At this condition, we must perform the tracheostomy. When the distance from postpharyngeal wall to lingual root was less than 2 mm, we also need to perform the...
tracheal cannula or the tracheostomy as respiration could not be improved for the operation of tongue-lip adhesion. Therefore, it is necessary for mandibular DO. However, DO does have limitations in the treatment of neonatal micrognathia. First, the procedure offers only a gradual improvement in the airway. The distraction usually progresses at a rate of 0.5 to 1.2 mm/d. Thus, the airway must be secured by some other means (endotracheal tube or tracheotomy) and intensively monitored throughout the distraction. Second, the DO device can be unwieldy, and the entire procedure is labor-intensive. Extreme care must be taken to ensure that the device remains securely attached to bone and that the screws are turned appropriately. Patient data must be accurately charted. The nursing staff and parents must diligently care for the incision and post sites. Third, the effect of mandibular DO on dental development is controversial. Injury to premolar tooth buds during the osteotomy may render them nonviable. Additional anatomic concerns include injury to the marginal mandibular branch of the facial nerve and the inferior alveolar nerve and asymmetry of face. If we can ensure correct indications and careful operative procedure, the adverse factors can be avoided. Most preliminary reports show favorable mandibular growth after DO for children with PRS. This allows the child to be successfully extubated or decannulated and typically allows the child to begin a regular oral diet. Patients must fulfill several criteria before mandibular DO is performed. The surgeon must be certain that the patient has adequate mandibular bone stock and the level of anoxia. Distance from postpharyngeal wall to lingual root must be established in lateral cephalogram. A preoperative comprehensive airway endoscopy is essential to rule out any other pathologic finding. A tracheal intubation can be performed; this should provide temporary relief of the airway obstruction. If the patient does not meet these criteria, then tracheotomy should be strongly considered.

REFERENCES
17. Roy S, Patel PK. Mandibular lengthening in micrognathic infants with the internal distraction device. Arch Facial Plast Surg 2006;8:60–64