17-year Follow-up after Mandibular Distraction Performed in an Infant with Robin Sequence

Takako Nakao, MD
Misato Katayama, MD
Yasushi Fujimori, MD, PhD
Shuichi Kawai, DDS, PhD
Takashi Nuri, MD, PhD
Koichi Ueda, MD, PhD

Summary: Robin sequence is a congenital anomaly, first described by Pierre Robin as a triad of micrognathia, upper airway obstruction and glossoptosis. The most significant airway management technique is the use of distraction osteogenesis to advance the mandible. On the other hand, late complications such as permanent dentition loss and malformation of the damaged teeth have been reported. Therefore, long-term follow-up after mandibular distraction is important. In our case report, we describe 17 years of follow-up after mandibular distraction performed in a 4-month-old male infant with Robin sequence. The patient was a male with a cleft palate. The tracheotomy and tongue–lip adherence was performed 1 month after birth, because the percutaneous oxygen saturation decreased to 70% during feeding and crying. However, postoperative X-ray examination revealed that he could not obtain sufficient airway after the operation. For the purpose of closing the tracheotomy early, we performed bilateral mandibular distraction at the age of 4 months. The bilateral corticotomy was performed on the mandibular body and an internal traction device was placed. The mandible was bilaterally lengthened by 17.5 mm, and expansion of the airway was confirmed by X-ray. A cleft palate operation was performed at the age of 17 months. At the age of 3 years, the tracheotomy orifice closed and speech/language training and orthodontics were begun. Dental panoramic radiographs showed favorable mandibular growth. No additional operation has been performed. Finally, his orthodontic treatment was completed without any complications at age 17. (Plast Reconstr Surg Glob Open 2020;8:e2651; doi: 10.1097/GOX.0000000000002651; Published online 6 February 2020.)

I n the presentation of Pierre Robin sequence, Pierre Robin described neonates with micrognathia, glossoptosis, and respiratory distress. Robin later added cleft palate deformity as an associated feature. During embryogenesis, the patient with Pierre Robin sequence has a small lower jaw that prevents tongue descent from between the palatal shelves and results in the U-shaped cleft palate, glossoptosis, and mandibular retrognathia.

The most serious manifestation is mechanical obstruction of the upper respiratory tract caused by depression of the tongue. Tracheostomy has been considered for management of upper airway obstruction; however, the morbidity of 25% to 50% and perioperative mortality of 0.5% to 5.0% at major centers of the newborn are reported to be high. Tongue–lip adhesion has been used for the relief of upper airway obstruction. Problems with this surgical treatment option are dehiscence, scarring, and feeding difficulties. The success rates were reported to be 18%, and 10 of the 11 patients required additional procedures.

The widely practiced airway management technique is the use of distraction osteogenesis to advance the mandible, which was first described by McCarthy et al. The distraction procedure treats the etiology of the problem by lengthening the mandible, advancing the base of the tongue off the posterior pharyngeal wall, and enlarging the hypopharyngeal region. This technique has been used both to achieve early decannulation and to avoid a tracheotomy in infants and children with craniofacial anomalies. On the other hand, late complications, ie, permanent dentition loss or malformation by the damaged tooth buds, have been reported.

Therefore, long-term follow-up observation after mandibular distraction is important.

Disclosure: The authors have no financial interest to declare in relation to the content of this article.
In this case report, we present 17 years of follow-up after mandibular distraction performed in a 4-month-old infant with Robin sequence.

CASE REPORT

The patient was a male with Robin sequence with cleft palate and was born with normal birth weight (2,875 g). He had mild dyspnea in supine positioning with mandibular hypoplasia. Because oral feeding became difficult and cyanosis occurred during feeding and crying 1 month after birth, he was admitted to the hospital emergently. The tracheotomy and tongue–lip adhesion were performed because the percutaneous oxygen saturation decreased to 70% during feeding and crying. However, postoperative X-ray examination revealed that he could not have obtained sufficient airway by tongue–lip adhesion. We decided to perform the bilateral mandibular distraction when he was 4 months old. The osteotomy line was set perpendicular to the occlusal plane. The bilateral corticotomy was performed on the mandibular body in front of the angle by using a reciprocating saw and a fine chisel, and the mobility was recognized by mobilizing the mandibular body using hands gently. An internal traction device (Zurich distractor R) was then placed (Fig. 1). We started the distraction after 2 days of latency period. The range was 1 mm per day. After 20 days, the mandibular was bilaterally lengthened by 17.5 mm and expansion of the airway was confirmed by X-ray. The distraction devices were removed after 6 months. We reported a part of the above-mentioned clinical course in a Japanese journal in 2003.

A cleft palate operation was performed at the age of 17 months. At the age of 3 years, the tracheostomy was closed and speech/language training and orthodontics were begun. The profilograms from lateral cephalograms show the favorable mandibular growth (Fig. 2). No additional operation was performed.

He was in Angle Class III condition before starting orthodontics, and his bilateral second molars were pulled in the correction of malocclusion under the orthodontist’s control.

Finally, his orthodontic treatment was finished without any complications, including nerve injuries when he was

---

Fig. 1. An internal distraction device fixed on the mandibular body. The arrow shows the mandibular angle. The osteotomy line was perpendicular to the occlusal plane.

Fig. 2. Profilograms of the patient from 5 years to 17 years showing the favorable mandibular growth according the facial growth.
The most serious manifestation in Robin sequence is mechanical obstruction of the upper respiratory tract by depression of the tongue. There are some treatments including tongue and lip adhesion that are popular as technique to relieve these symptoms. Recently, mandibular distraction has become popular to perform on young patients. Genecov et al. evaluated in 67 micrognathia patients with bilateral mandibular distraction performed at the age of 1–2 years enabled decannulation in 92%. Miloro reported performing distraction at an earlier age averaging 3.5 months could avoid tracheotomy and other surgical intervention. On the other hand, Scott et al. reported dentition loss or malformation as the most common complication after mandibular distraction in infants younger than 3 months and found that it could be avoided by osteotomy posterior to the tooth buds. They preferred to perform the osteotomy either superior or anterior to the angle of the mandible.

Moreover Weichman et al. concluded that mandibular distraction is successful, provided that there is a comprehensive clinical program emphasizing adequate mandibular bone stock, proper vector selection, planned overcorrection, and comprehensive orthodontic management. Davidson, et al. recommend that an internal device is indicated in single-plane distraction (especially for a vertical vector) of native bone with a mandibular width greater than 12 mm. External devices have a higher risk of scar burden and displacement with rollover. In our case, we succeeded in obtaining good bone osteogenesis.

However, an internal device is indicated in single-plane distraction. The outcome depends more directly on the vector. Furthermore, remodeling contributed to achieving normal occlusion in our case. Planned overcorrection allowed remodeling and favorable mandibular growth during 17 years of growth in our patient. We observed that permanent molars were formed normally.

**CONCLUSION**

Here we have shown the results after 17 years with earlier invasive surgery and orthodontics. Mandibular distraction treats the etiology of the disease process and does not disturb future growth.

Koichi Ueda, MD, PhD

Department of Plastic and Reconstructive Surgery
Osaka Medical College
Takatsuki, Osaka 569-8686, Japan
E-mail: pla007@osaka-med.ac.jp

**REFERENCES**

1. Paletta CE, Dehghan K, Hutchinson RL, et al. A fall of the base of the tongue considered as a new cause of nasopharyngeal respiratory impairment: Pierre Robin sequence, a translation. **Plast Reconstr Surg.** 1994;93:1301–1303.
2. Miloro M. Mandibular distraction osteogenesis for pediatric airway management. **J Oral Maxillofac Surg.** 2010;68:1512–1523. doi:
3. Boston M, Rutter MJ. Current airway management in craniofacial anomalies. **Curr Opin Otolaryngol Head Neck Surg.** 2003;11:428–432.
4. McCarthy JG, Schreiber J, Karp N, et al. Lengthening the human mandible by gradual distraction. **Plast Reconstr Surg.** 1992;89:1–8; discussion 9.
5. Williams JK, Maull D, Grayson BH, et al. Early decannulation with bilateral mandibular distraction for tracheostomy-dependent patients. **Plast Reconstr Surg.** 1999;103:48–57; discussion 58.
6. Scott AR, Tibesar RJ, Lander TA, et al. Mandibular distraction osteogenesis in infants younger than 3 months. **Arch Facial Plast Surg.** 2011;13:173–179.
7. Fujimori Y, Ueda K, Otani K, et al. A case of Robin sequence with dyspnea performed distraction osteogenesis in a baby. **J Plast Reconstr Aesthet Surg.** 2003;25:332–336.
8. Genecov DG, Barceló CR, Steinberg D, et al. Clinical experience with the application of distraction osteogenesis for airway obstruction. **J Craniofac Surg.** 2009;20(suppl 2):1817–1821.
9. Weichman KE, Jacobs J, Patel P, et al. Early distraction for mild to moderate unilateral craniofacial microsomia: long-term follow-up, outcomes, and recommendations. **Plast Reconstr Surg.** 2017;139:941e–953e.
10. Davidson EH, Brown D, Shetye PR, et al. The evolution of mandibular distraction: device selection. **Plast Reconstr Surg.** 2010;126:2061–2070.