ORIGINAL ARTICLE

Management of congenital airway obstruction due to mandibular hypoplasia using distraction osteogenesis: experience from a specialized institute in Lima, Peru

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ABSTRACT

Bilateral mandibular hypoplasia is the leading cause of airway obstruction in children and can result in both respiratory and digestive distress. Treatment often involves mandibular distraction osteogenesis, a technique that increases the anteroposterior length of the mandible. This study aimed to share the experiences of a pediatric specialty institute in Lima, Peru, using distraction osteogenesis to restore airway and digestive function. Data on sociodemographic and clinical (pre-and postoperative) information were collected from patients who underwent bilateral mandibular distraction between 2016 and 2022. Seventeen patients were included, 53 % of whom were between 1 and 11 months old. All patients required care in the Intensive Care Unit, with 58.8 % (9/17) needing orotracheal intubation and 41.2 % (8/17) undergoing tracheostomy. The procedure resulted in anatomical and functional improvements in both the airway and digestive system by increasing the anteroposterior distance of the mandible. These findings suggest that distraction osteogenesis is an effective approach for managing airway obstruction caused by mandibular hypoplasia in pediatric patients.

Keywords: Osteogenesis, Distraction; Pierre Robin Syndrome; Micrognathism; Tracheostomy (Source: MeSH)

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Experiencia en el manejo de obstrucción congénita de vía aérea por hipoplasia mandibular mediante distracción osteogénica en un instituto especializado de Lima, Perú

RESUMEN

La hipoplasia mandibular bilateral es la causa más frecuente de obstrucción de las vías aéreas y puede producir dificultad respiratoria y digestiva. Para su resolución, usualmente se requiere realizar una distracción osteogénica mandibular, que permite incrementar la distancia anteroposterior de la mandíbula. El objetivo de este estudio fue describir las experiencias de un instituto pediátrico especializado ubicado en Lima, Perú, en la aplicación de distracción osteogénica para recuperar la función de las vías respiratorias y del tracto digestivo. Se recopiló información sociodemográfica y clínica (pre y postoperatoria) de pacientes sometidos a distracción mandibular bilateral entre el 2016 y el 2022. Se incluyeron un total de 17 pacientes. El 53 % de los casos tuvieron edades entre 1 y 11 meses. Todos los pacientes permanecieron en la Unidad de Cuidados Intensivos, requiriendo el 58,8 % (9/17) intubación orotraqueal y el 41,2 % (8/17) traqueostomía. Como resultado de la intervención, se produjeron mejoras anatómicas y funcionales de las vías respiratorias y digestivas, mediante el aumento mandibular anteroposterior. Los hallazgos sugieren que la distracción osteogénica es una técnica útil para el manejo de la obstrucción de las vías aéreas causada por hipoplasia mandibular en la edad pediátrica.

Palabras clave: Osteogénesis por Distracción; Síndrome de Pierre Robin; Micrognatismo; Traqueostomía (Fuente: DeCS)

INTRODUCTION

Congenital airway obstruction at the level of the oral or cervical cavity has various etiologies, ranging from anatomical malformations that reduce the airway space to tumors that obstruct airflow either by interposition or by compression of the respiratory tract. Mandibular hypoplasia is one of the most frequent anatomical malformations, with bilateral hypoplasia being the form that causes upper airway obstruction. Additionally, cases of Pierre Robin sequence have been reported, a congenital condition characterized by micrognathia, glossoptosis, and cleft palate (1–3).

Airway obstruction presents clinically as tachypnea, stridor, retractions, or sleep apnea. Diagnostic support studies include computed tomography to evaluate the mandible and airway, fiberoptic endoscopy to assess the airway, and polysomnography to assess the degree of sleep apnea. Airway obstruction compromises the patient's ability to feed, and in some cases, placement of a gastrostomy is required to prevent malnutrition and growth delay (1–3).

The goal of treatment is to improve airway function. Conservative management includes positional therapy and is indicated for patients with weight gain and moderate upper airway obstruction. In contrast, surgical management encompasses several options, such as tongue-lip adhesion (glossopexy), tracheostomy, and distraction osteogenesis. Glossopexy is usually poorly tolerated, as it limits the sucking reflex; it is also not definitive and often requires additional procedures. Tracheostomy offers an immediate solution for airway patency.

However, this technique carries risks such as granuloma formation, stenosis, tracheomalacia, and occasional bleeding. Moreover, it must be maintained until the patient achieves sufficient development for the obstruction to decrease. In contrast, bone distraction offers an alternative treatment by lengthening the mandibular bone in an anteroposterior direction. This gain in dimension results in anterior positioning of the tongue, thereby avoiding the need for tracheostomy and even gastrostomy by expanding the upper airway space. Mandibular distraction is indicated in patients with micrognathia and obstructive sleep apnea (3–8).

The surgical technique for mandibular distraction involves bilateral osteotomy using osteosynthesis materials (percutaneous Kirschner wires or monocortical screws) at the junction between the body and the angle of the mandible, followed by the placement of a mandibular distractor. According to various publications, parameters such as the duration the distractor must remain in place, the latency period (0 to 7 days), the distraction rate (1–4 mm/day), and the consolidation period (20 to 90 days) are variable (4–8).

A meta-analysis by Breik et al. (5) suggests that mandibular distraction is a highly successful procedure, indicated in neonates and infants with airway obstruction secondary to micrognathia, avoiding tracheostomy in up to 95% of cases. According to the same author, the most common causes of treatment failure include previously undiagnosed airway obstruction, central apnea, undiagnosed neurological abnormalities, and the presence of cardiovascular comorbidities, which have been similarly described in other publications (9–11).

This study aimed to describe the experience of the Instituto Nacional de Salud del Niño San Borja, a pediatric reference center in Lima, Peru, in performing mandibular distraction. Although this technique is known, it is not frequently applied relative to the number of pediatric patients who require it due to the cost of distractors, the shortage of experienced surgeons, and the limited capacity of healthcare institutions. Our study seeks to describe the use of bilateral mandibular distraction as a solution for airway obstruction due to bilateral mandibular hypoplasia, with the added benefit of avoiding or removing a tracheostomy, in the hope that this procedure can be replicated in other institutions for similar cases.

METHODS

The study was conducted at the Instituto Nacional de Salud del Niño San Borja (INSN-SB), a national referral center for the surgical treatment of complex pathologies. We included all patients diagnosed with acute or chronic respiratory failure, tracheostomy, and micrognathia or maxillary developmental abnormalities who underwent bilateral mandibular distraction osteogenesis between January 2016 and December 2022.

Data were collected on age, sex, signs and symptoms, diagnosis, anteroposterior maxillomandibular discrepancy, surgical treatment performed, complications, relapses, and sequelae. The mandibular-maxillary anteroposterior discrepancy (overjet) was calculated from the alveolar ridge of the maxilla to the alveolar ridge of the mandible (or from the incisal edges of the central incisors if present).

Statistical analysis was performed using SPSS (Statistical Package for the Social Sciences) version 25. Quantitative variables were reported as means and ranges, while categorical variables were presented as frequencies and percentages.

The study received approval from the Institutional Research Ethics Committee of INSN-SB prior to execution. The patient's anonymity and confidentiality of the collected data were maintained.

RESULTS

A total of 17 patients underwent bilateral mandibular distraction osteogenesis during the study period. As an example, Figure 1 shows the clinical progression of one of the included patients. Males accounted for 52.9% (n = 9), and the average age was 2.9 years (range = 1–8 years). The age group with the highest number of cases was infants, followed by the 1 to 5 years age group (Table 1). The most frequent sign was micrognathia in 14 cases (82.4%), followed by respiratory distress in 13 cases (76.5%) (Table 2). A total of 12 patients (70.6%) had not undergone any prior surgical intervention, while 3 patients (17.6%) had previously undergone tracheostomy, and 2 (11.8%) gastrostomies

Table 1. Distribution of patients included in the study by age group

Age group	n	%
Neonates	1	6
Infants (2–11 months)	9	53
Preschool children (1–5 years)	5	29
School-aged children (>6 years)	2	12
Total	17	100

Table 2. Clinical characteristics of patients included in the study

Characteristic	n	%
Micrognathia	14	82.4
Respiratory difficulty	13	76.5
Sleep apnea	12	70.6
Microstomia	3	17.6
Tracheostomy	3	17.6
Gastrostomy	2	11.8
Anterior open bite	1	5.9
Intubation since birth	1	5.9
Total	17	-

(surgeries performed at other institutions). Only one patient was admitted to the intensive care unit (ICU) with orotracheal intubation upon arrival. The most frequent diagnosis was Pierre Robin sequence, identified in 14 patients (82.4%). The average overjet was 14.2 mm (range = 10–27 mm) (Table 2).

Preoperative intubation was performed in 13 of the 17 patients. In 52.9% of these cases, airway visualization was achieved, and intubation was completed with the assistance of a flexible fiberoptic bronchoscope. In 4 cases (23.5%), visualization of the airway was not possible via bronchoscopy, and a tracheostomy was performed following the failed attempt to proceed with surgery. Three patients (17.6%) already had a tracheostomy, and one (5.9%) was already intubated upon admission.

Description of the surgical technique

A bilateral submandibular incision was made, followed by dissection down to the mandibular basal and buccal cortex. Subperiosteal dissection was then performed to provide adequate space (approximately 10–13 mm) for the distractor's fixation. In all cases, an oblique osteotomy was made at the junction of the mandibular body and angle. The osteotomy was partially performed using a saw or surgical bur and completed with a chisel for the lingual cortex and alveolar ridge, using pivoting movements to avoid entering the oral cavity.

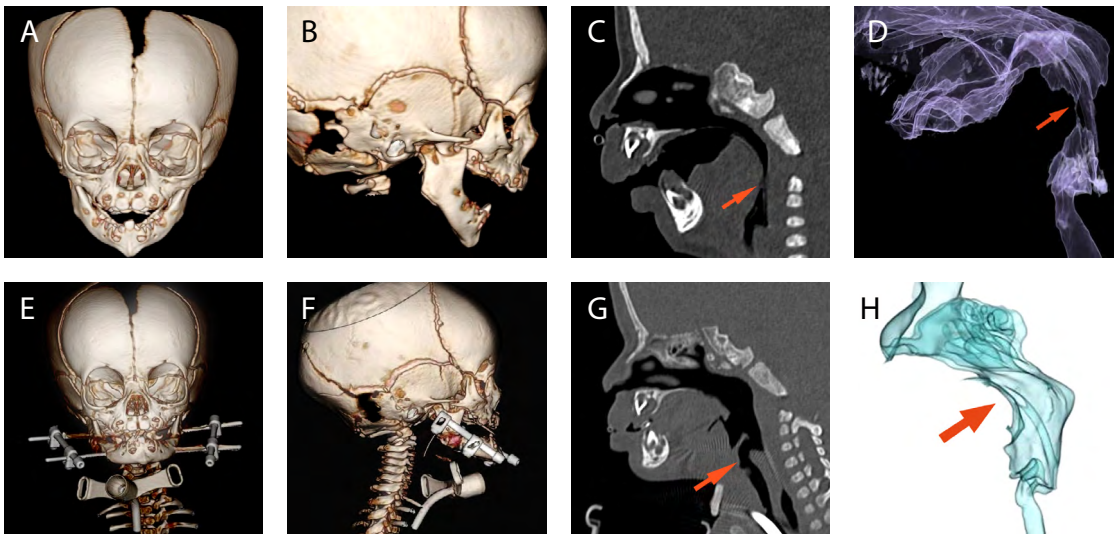
Figure 1. Postoperative progression of a 4-year-old female patient included in the study

Left: photographs taken before the operation. Frontal view (A) and lateral view (B) of the patient.
Right: photographs taken after the operation. Frontal view (C) and lateral view (D) of the patient.

For external distractors, 1 or 2 Kirschner wires were placed, depending on the size of the selected distractor and the patient's mandible. The wires had a threaded tip with a diameter of 2.0 mm for fixation. Kirschner wires were placed proximally and distally to the osteotomy, maintaining a distance of 5 mm from the osteotomy edge and an additional 5 mm if a second wire was inserted on the same side of the osteotomy (Figure 2).

For distractors fixed adjacent to the mandibular cortex, three 1.8 mm screws were used proximally and two 1.8 mm screws distally, with the activation pin emerging percutaneously at the intra-auricular level. Distractors with lengths of 30, 40, and 50 mm were used, depending on the mandibular size and patient age. The latency period was 3–5 days for children under 1 year old and 5–7 days for those older than 1 year. The average distraction length was 7.6 mm, with a mean overcorrection of 7.6 mm and a range of 6–10 mm (Figure 3).

Figure 2. CT images of a 6-month-old patient included in the study

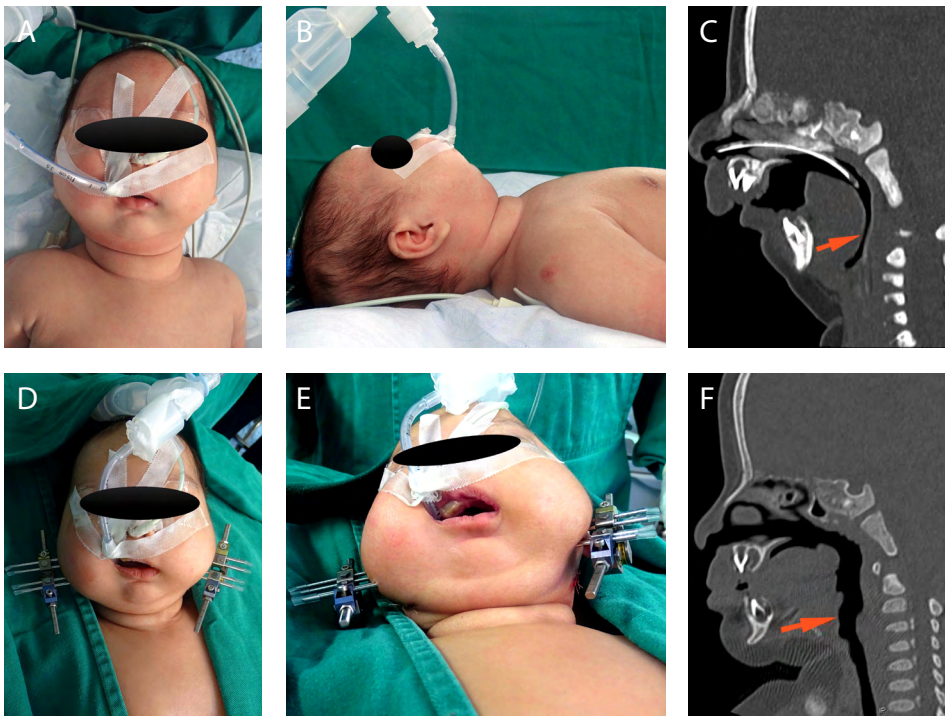


Top: Frontal (A), lateral (B), sagittal (C) views, and 3D airway reconstruction (D) (arrow indicates supraglottic anteroposterior narrowing).

Down: Frontal (E), lateral (F), and sagittal (G) views with distractors, and 3D reconstruction showing anteroposterior airway increase after mandibular distraction (H).

Figure 3. Postoperative progression of a 3-month-old female patient included in the study

Left: photographs taken before the surgery (A and D).
Center: postoperative photographs showing the patient without distractors (B and E).
Right: photographs taken at the second postoperative follow-up (C and F).



Top: Frontal (A) and lateral (B) views of the patient intubated via bronchoscopy, and lateral view of the airway before surgery (C).
Bottom: Frontal (D) and lateral (E) views of the patient with mandibular distractors, and lateral view of the airway after surgery (F).

Postoperative care and follow-up

All patients were admitted to the ICU postoperatively, 10 with orotracheal tubes (58.8%) and 7 with tracheostomies (41.2%). Following the latency period, the distraction phase was initiated according to the previously described protocol. The latency period was defined as the time from the day after surgery to the start of distraction. The distraction period involved daily lengthening of the bone callus using a device turned around a pin on the external end of the distractor. During distraction, 1 mm per day was achieved through two 0.5 mm turns. Overcorrection was defined as the mandibular ridge or lower incisors surpassing the maxillary ridge or incisors. Fourteen patients (84.2%) reached 20 mm of distraction, while the remaining three achieved 25 mm, 30 mm, and 35 mm (Table 3). In the eight patients (47.1%) who had tracheostomies, the tube was removed simultaneously with the distractors. The remaining nine (52.9%) only had orotracheal intubation, which was removed postoperatively in the ICU (Table 4). The bone consolidation period was equivalent to twice the sum of the latency and distraction periods before removing the distractors.

Patients were followed up every six months for the first two years post-surgery and annually thereafter, with a maximum follow-up of seven years in one case. In nine patients, the orotracheal tube was removed during distraction based on the progressive withdrawal of sedation, initiation of respiratory therapy, and clinical evidence of increased mandibular projection. Patients with tracheostomy remained in the ICU

Table 3. Diagnosis, age, mandibular–maxillary anteroposterior discrepancy (overjet), and distraction values in the patients included in the study

Diagnosis	Age (months)	Overjet (mm)	Distraction (mm)
Pierre Robin sequence	12	13	20
Micrognathia	48	18	25
Pierre Robin sequence	12	12	20
Pierre Robin sequence	6	12	20
Pierre Robin sequence	9	12	20
Pierre Robin sequence	12	13	20
Pierre Robin sequence	11	12	20
Pierre Robin sequence	12	14	20
Pierre Robin sequence	8	13	20
Pierre Robin sequence	0.03	10	20
Pierre Robin sequence	7	13	20
Pierre Robin sequence	2	11	20
Pierre Robin sequence	3	13	20
Hanhart syndrome	96	23	30
Treacher Collins syndrome	96	27	35
Pierre Robin sequence	2	13	20
Pierre Robin sequence	2	12	20

Table 4. Procedures performed in the patients included in the study

Procedures	n	%
Tracheostomy removal + distractor removal at 4 months	1	5.9
Tracheostomy removal + distractor removal at 3 months	7	41.2
Orotracheal extubation in ICU at 20 mm distraction + distractor removal at 3 months	4	23.5
Orotracheal extubation in ICU at 10 mm distraction + distractor removal at 3 months	5	29.4
Total	17	100

only during the latency period, as the tracheostomy ensured airway patency. In those without tracheostomy but with prior respiratory distress or apnea, these episodes resolved according to parental reports.

Bilateral submandibular scars evolved favorably in all patients, with no cases of hypertrophic or keloid scarring. All patients received orogastric feeding during their ICU stay, which was discontinued once oral feeding could safely resume.

A patient diagnosed with Pierre Robin sequence experienced a postoperative complication unrelated to the surgical procedure. This case involved a two-month-old infant who underwent fiberoptic-guided intubation preoperatively and required a tracheostomy on the seventh postoperative day due to a distal orotracheal tube leak.

DISCUSSION

Surgical intervention in patients with congenital upper airway obstruction should be performed as early as possible to avoid the need for tracheostomy, to allow its removal if already present, to improve swallowing, to address respiratory distress and sleep apnea, and, if needed, to initiate respiratory and swallowing therapy. Without bilateral mandibular distraction, tracheostomy would be required to resolve the airway obstruction; however, this entails specific care and training for the patient's caregivers and does not provide a short-term solution without the tracheostomy. Bouchard et al. (2) recommend distraction osteogenesis for pediatric patients with obstructive apnea, as it allows for significant mandibular advancement without the need for bone grafts and is associated with a lower risk of recurrence.

Given the presence of micrognathia (with an average discrepancy of 14.2 mm), measures must be taken to ensure a safe airway for surgical intervention. In cases requiring intubation, flexible fiberoptic-guided intubation is the first procedure attempted, with a success rate of 69.2% in this study. Intubation is performed while keeping tracheostomy as a backup procedure in cases where orotracheal intubation is unsuccessful. In this series, tracheostomy was performed as a secondary procedure in four patients.

Removal of internal distractors tends to be more laborious than removal of external distractors. When internal screws placed on both sides of the osteotomy are removed, they detach and shift away from the incision site due to the distraction process, requiring a wider surgical field. Distractors do not interfere with breastfeeding, oral formula intake, or the progression to soft or full diets in children older than one year. In this study, patients with tracheostomy resumed oral feeding after the postoperative recovery period without any physical limitations caused by the distractors.

While Miloro (4) implemented no latency period, using daily advancements of 3 to 5 mm in his infant series, Diep et al. (6) adopted standardized latency periods of 3 to 5 days with daily advancements of 1 to 1.2 mm. Our established consolidation period ranged from 48 to 52 days, aligning with findings from Diep et al. (6) and Resnick et al. (9). However, most patients in our study experienced delays in returning for distractor removal.

Pierre Robin sequence was the most frequent diagnosis in our series compared to other syndromes or isolated micrognathia, a finding consistent with the global literature. Since Pierre Robin sequence includes micrognathia, glossoptosis, and airway obstruction, bilateral mandibular distraction is essential to prevent upper airway collapse and its associated complications (1, 6–8).

A significant limitation of this study is that some patients were not referred to INSN-SB because the tracheostomy had already temporarily resolved the airway obstruction without further surgery at the referring institution.

The results of this study suggest that bilateral mandibular distraction is an effective surgical treatment for neonates and infants with respiratory compromise, as evidenced by respiratory distress and supported by imaging, bronchoscopy, or polysomnography. Bilateral mandibular distraction enabled anteroposterior mandibular advancement through complete osteotomy and strict adherence to distraction protocols, increasing airway diameter and anterior displacement of the tongue and hyoid bone; key factors in determining the removal of the tracheostomy and/or orotracheal tube and the resumption of oral feeding. Surgical success must be supported by respiratory and swallowing therapy. Based on our findings, we conclude that distraction osteogenesis improves both the anatomical structure and function of the airway in cases of congenital airway obstruction due to mandibular hypoplasia.

Author contributions

Conceptualization: JFOA.

Data collection, management, and curation: JFOA, RUQ.

Data analysis: JFOA, RUQ, KCCC.

Visualization: JFOA, RUQ, KCCC.

Writing – original draft: JFOA, RUQ, KCCC.

Writing – review and editing (final version): JFOA, RUQ, KCCC.

Conflicts of interest

The authors declare no conflicts of interest associated with the material presented in this manuscript.

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