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Feeding and mandibular distraction osteogenesis in children with Pierre Robin sequence: A case series of functional outcomes

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ABSTRACT

Introduction: In addition to upper airway obstruction, many patients with micrognathia and Pierre Robin sequence also have swallowing abnormalities and reflux. Many studies have demonstrated the effectiveness in alleviating the airway symptoms with mandibular distraction osteogenesis, but very few studies have focused on feeding and reflux outcomes.
Methods: A retrospective chart review was performed to identify patients with Pierre Robin sequence who underwent mandibular distraction osteogenesis with completed pre- and post-operative upper gastroesophageal series and videofluoroscopic swallow assessments.
Results: All six children in our series demonstrated significant improvements in both airway obstructive symptoms and feeding abnormalities. More specifically, all patients showed clinical and objective improvements in reflux and swallowing function after distraction surgery.
Conclusion: Objective and symptomatic improvements in swallowing function and reflux disease can be seen after mandibular distraction osteogenesis in children with Pierre Robin sequence.

1. Introduction

Upper airway obstruction secondary to micrognathia was first widely described by Pierre Robin in 1934 [1]. He described a constellation of findings, which included micrognathia, glossoptosis, and in some patients, cleft palate. These findings are now commonly referred to as Pierre Robin sequence (PRS). Some craniofacial syndromes were later recognized to be associated with PRS. Most notably they include Stickler syndrome, Treacher Collins syndrome, and Nager syndrome [2].

Micrognathia can cause upper airway obstruction due to posterior tongue collapse and physical obstruction of the oropharyngeal and hypopharyngeal regions. Although the majority of children born with micrognathia or PRS can be treated with conservative management, some patients may have significant respiratory issues, necessitating more aggressive interventions [3–5].

Traditionally, tracheostomy has been the most effective and definitive treatment option for these patients [6]. Tracheostomy, however, is associated with frequent morbidity, high cost, and occasional mortality [7–9].

Distraction osteogenesis of the mandible is a relatively new treatment option in children with PRS, which has been shown to be very effective in relieving the upper airway obstruction by gradually lengthening the mandible, which leads to the correction of the posterior tongue base position.

Feeding and swallowing problems, along with gastroesophageal reflux disease (GERD), have commonly been associated with PRS [10]. The exact pathophysiology behind this association is currently unknown but the prevailing theory involves the feeding issues being caused by the upper airway obstruction and the subsequent respiratory difficulties [10,11]. In relation, GERD is thought to originate from the negative intrathoracic pressure generated by the upper airway obstruction, which can cause a "suction" effect on the gastric contents [10–12].

Although many studies have demonstrated the efficacy of mandibular distraction on improving the symptoms related to airway obstruction, very few articles have shown any objective evidence of swallowing and GERD changes after mandibular distraction osteogenesis.

In this study the effects of mandibular distraction osteogenesis on feeding and GERD are reported in a small series of infants with PRS. More specifically, the effectiveness in relieving the swallowing dysfunction and reflux by comparing the pre- and post-operative

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objective assessments (videofluoroscopic swallow test and upper gastroesophageal (GI) series) is presented.

2. Study methods

2.1. Patients

Research Ethics Board approval was obtained for this study. A retrospective chart review was performed to identify all children who underwent bilateral mandibular distraction osteogenesis with complete preoperative and postoperative videofluoroscopic swallow tests (modified barium swallow) and upper GI series (barium swallow).

Consultation with a feeding team, consisting of a nurse and an occupational therapist, was carried out in all children. Patient characteristics, perioperative investigations, operative details, complications, and postoperative functional outcomes were documented. More specifically, the changes noted in the modified barium swallow tests and the upper GI series after the operation were reviewed for each patient. The radiologist's report was the primary outcome measure utilized in our analysis and if there was any uncertainty regarding the reports, it was reviewed again with the aid of a radiologist. The same procedure was carried out with a speech language pathologist when reviewing the videofluoroscopic swallow assessments.

2.2. Distraction osteogenesis surgery

All patients were placed under general anesthesia and intubated orally. Internal mandibular distraction devices were used for all patients. One patient had an absorbable distractor device placed through an intraoral incision while the rest had non-absorbable devices with an external cutaneous approach (modified Risdon incision). A buccal corticotomy (including the cephalic and caudal borders) was first performed with a reciprocating saw. The remainder of the osteotomy was performed with an osteotome placed along the lingual cortex. The overall osteotomy was performed in an inverted L-shape and the inferior alveolar nerve was identified and preserved in all cases. The vector of distraction in each case was planned during the preoperative phase using clinical and imaging assessments.

Distraction was initiated on the first postoperative day at a rate of 1.5 mm to 2.0 mm per day. Distraction was ended when there was no maxillomandibular discrepancy but the device (non-absorbable) was left in place for the consolidation period, which lasted between 6 and 8 weeks. Please refer to Table 2 for surgical details pertaining to the mandibular distraction osteogenesis procedures.

3. Results

Six children were identified who underwent bilateral mandibular distraction osteogenesis and who had complete documentation of both pre- and post-operative videofluoroscopic swallow assessments and upper GI series. Four boys and two girls underwent surgery at a mean age of 65 days with a range of 28–94 days. All patients were found to have PRS (micrognathia, glossoptosis, and airway obstruction) and all had a cleft palate. Four of the six patients had associated genetic syndromes (Table 1).

Table 1
Summary of the patient characteristics.

Patient	Age[a] (Days)	Gender	PRS[b]	Syndromes	Reflux	Feeding[c]
1	89	M	Present	Otopalatodigital	Severe	ND
2	69	F	Present	Stickler	Yes	NG
3	94	M	Present	4p deletion	Yes	NG
4	78	M	Present	No	Yes	NG
5	32	M	Present	No	Yes	NG
6	28	F	Present	Stickler	Yes	NG

[a] Age at the time of operation.
[b] Pierre Robin sequence (micrognathia, glossoptosis, +/− cleft palate).
[c] ND, nasoduodenal tube; NG, nasogastric tube.

All patients had clinical symptoms and physical examination findings of severe upper airway obstruction that was not adequately managed with conservative therapy. Half of the patients required intubation prior to the distraction procedure to secure the airway. Briefly, newborns were considered to have severe airway obstruction requiring surgical intervention if they were intubated at birth and later failed extubation and/or presented with significant oxygen desaturations with signs of respiratory distress despite conservative measures, such as positioning.

All patients were found to have GERD in the preoperative setting that required anti-reflux medical therapy (see below). One patient had intractable GERD with severe episodes of emesis requiring a placement of nasoduodenal feeding tube for nutrition and pro-motility medication, in addition to the standard anti-reflux management (patient 1). All patients required gavage feedings due to feeding and swallowing difficulties and subsequent failure to thrive. Aspiration was documented in all children in our series, further necessitating the gavage tube feedings (see below).

Table 2 summarizes the clinical information during the perioperative period. Preoperative investigations included flexible pharyngolaryngoscopy, bronchosocopy, and imaging of the craniofacial skeleton. In addition, all but one patient underwent a sleep study to rule out the presence of central apneas. All of the patients tested with a sleep study had documented obstructive sleep apnea with a mean apnea/hypopnea index of 6.4 and lowest oxygen saturation levels of 72%. More importantly, none of the patients had evidence of central apneas. Bronchoscopies did not reveal any significant distal airway abnormalities (e.g. tracheomalacia), but in one patient some erythema possibly due to uncontrolled reflux was identified (patient 1).

None of the patients had any significant complications related to the mandibular distraction surgery. Two patients had local erythema and tenderness around the activator site, which was treated successfully with antimicrobial ointment and oral antibiotic

Table 2
Summary of the preoperative investigations and operative details.

Patient	Bronchoscopy	Sleep study	Imaging[b]	Distraction length (mm)	Distractor device
1	Mild erythema[a]	None	3-D CT	20	Internal nonresorbable
2	Normal	OSA	Cephalogram	16	Internal resorbable
3	Normal	OSA	3-D CT	18	Internal nonresorbable
4	Normal	OSA	3-D CT	18	Internal nonresorbable
5	Normal	OSA	3-D CT	15	Internal nonresorbable
6	Normal	OSA	3-D CT	20	Internal nonresorbable

[a] Attributed to severe GERD.
[b] 3-D CT: 3-dimensional computed tomography.