Dental Anomalies in Patients with Pierre-Robin After Distraction Osteogenesis

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INTRODUCTION

Pierre-Robin Sequence (PRS) is a rare congenital birth defect described by a triad of a mandibular hypoplasia, glossoptosis, and airway obstruction. PRS can be either a syndromic or a non-syndromic diagnosis[1]. It is referred to as a sequence because the initial defect, mandibular hypoplasia, leads to the additional clinical findings of glossoptosis and airway obstruction. Some infants may face respiratory challenges associated with the more severe clinical findings. In select cases, surgical intervention, known as distraction osteogenesis, may be indicated. In this intervention, an osteotomy is completed and a titanium device, known as a distractor, is placed bilaterally in the right and left ramus' acting to mechanically pull the boney surfaces apart slowly. The bone then undergoes osteogenesis resulting in additional bone deposition in the space and an increase in bone length. This procedure can help achieve adequate airway patency to facilitate breathing without the alternative of a tracheostomy. Despite the benefit of increasing airway patency, osteotomies of the mandible demonstrated possible long-term consequences to the developing dentition.

ETIOLOGY AND EPIDEMIOLOGY

- Prevalence of Pierre Robin Sequence is approximately 1 in 8,500 to 1 in 14,000 newborns a year[2].
- Non-syndromic cases of PRS have been linked to de novo gene mutations on chromosomes 2, 4, 11, and 17[1].
- Researchers have identified mutations of SOX9 or KCNJ2 on chromosome 17 may lead to PRS[1].
- Retrospective chart review has preliminarily shown that syndromic PRS has been reported to account for 60% of PRS[3].

DIAGNOSIS AND MANAGEMENT

- Early concerns for PRS diagnosis are based on ultrasound findings of micrognathia. Prenatal diagnosis can help in counseling parents on post-delivery expectations, treatments, and complications. Since there is a high association with multiple syndromes; genetic counseling may also be recommended if preliminary ultrasound findings are indicative of PRS.
- Treatment varies based on the severity of condition.
  - Mild cases: Prone and lateral positioning allows positional relief of airway obstruction in 70% of cases. Continuous positive airway pressure is another intervention that can benefit these patients although compliance in this age group is challenging[4].
  - Severe cases: May require surgical intervention such as tongue-lip adhesion, mandibular distraction osteogenesis, or tracheostomy. An estimated 10% of non-syndromic severe PRS patients require surgical management[5].

CASE REPORT

This case report presents a 7 year-3-month-old male who presented to Children's Hospital Los Angeles for a dental exam concerning a morphologically atypical molar. His medical history was significant for non-syndromic Pierre Robin Sequence. He underwent bilateral MDO, with deep internal distraction, from 3 months to 6 months of age to address his severe obstructive sleep apnea and mandibular AP hypoplasia. Extraoral findings were consistent with his history of distraction osteogenesis (**Figure 1**). Multiple dental cavities were identified at his exam, and he was scheduled to be treatment under general anesthesia in the operating room due to extensive dental caries and patient's acute situational anxiety. Upon clinical and radiographic examination, the left developing first permanent molar was malformed in both coronal and root segments (**Figure 2**). Based on patient ability to sit for possible future extraction and poor long-term prognosis of the malformed tooth the tooth was extracted, and all remaining dental caries treated (**Figure 3**). Post-operatively a panoramic x-ray was taken to view developing teeth to which the left second permanent molar was confirmed as missing (**Figure 4**). To prevent supra-eruption of the opposing dentition; A Nance appliance will be provided until more interventional orthodontics is started. Despite the long-term dental findings, his surgery allowed for an improved quality of life, as the patient experienced a decrease in apnea-hypopnea index (AHI) and avoided requiring a tracheostomy as the definitive treatment for his mandibular hypoplasia (**Figure 5**).

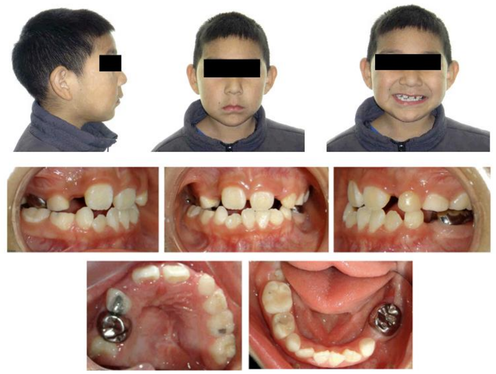

Figure 1. Extra-oral and intra-oral photographs

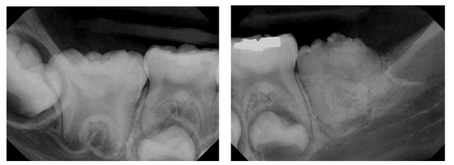

Figure 2. Right and Left periapical radiographs, respectively.

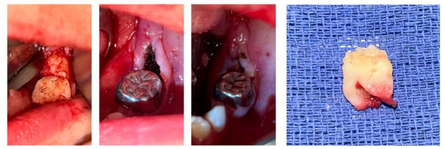

Figure 3. Extraction process; and presentation of affected permanent 1st molar.

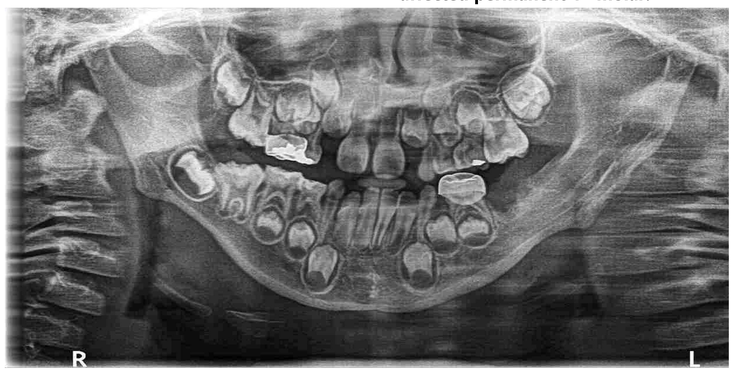

Figure 4. Panoramic X-ray taken post-operative; showing missing 2nd permanent molar

DENTAL IMPLICATIONS OR CLINICAL FEATURES

Although not all cases of Pierre-Robin Sequence with distraction osteogenesis result in dental complications the following have all been recorded in current literature. The following statistics are from a systematic review by I. Shuman and V.A. Cardo Jr (2021), regarding documented long-term outcomes of mandibular distraction osteogenesis.

- Positional changes (35%)
- Shape alteration/fracture (24%)
- Missing Teeth (15%)
- Root malformation (14%)
- Follicle/bud perforation (9%)
- Delayed eruption (2%)
- Dentigerous cyst formation (1%)

This review additionally identified that permanent first molars are the most affected tooth; followed by the second primary molar [6]. Once a tooth injury is identified; long-term prognosis of involved teeth will need to be evaluated and incorporated into a patient's orthodontic treatment plan. Possible treatment to these teeth varies dependent on malformation and include, but are not limited to, orthodontic re-positioning, full-coverage restorations, implant treatment, and extraction.

	Age	AHI
Pre-Mandibular Distraction	3 mos.	**18.3** per hour
Post-Mandibular Distraction	5 mos.	**11.6** per hour
	8 mos.	**7.8** per hour
	18 mos.	**1.9** per hour
	29 mos.	**0.7** per hour

Figure 5. Relationship of mandibular distraction to improved AHI.

CONCLUSION

This case report presents a child with Pierre-Robin Sequence who underwent bilateral mandibular distraction to address his severe sleep apnea and the ramifications. The report discusses the possible presentations and management of dental injuries as well as the importance of a multi-disciplinary team in addressing dental concerns. Although these dental injuries are not guaranteed, proper planning with a dental team should include the discussion of possible outcomes to injured teeth. If tooth removal is deemed necessary, orthodontic preservation of the arch and future prosthodontic treatment may be necessary.

References

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