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Case Report

Pierre Robin Syndrome: A Rare Genetic Disorder and its Dental Management

Chirag M. Raiyani[1*], Ruchi Arora[2], Deepak P. Bhayya[3], Subha Dogra[4]
[1]Postgraduate student, [2]Professor and Head, [3]Professor, [4]Seniour Lecturer, Department of Pedodontics and Preventive Dentistry, Darshan Dental College and Hospital, Udaipur- 313001, Rajasthan, India

***Corresponding author**
Dr. Chirag M. Raiyani
Email: drchiragpedo@gmail.com

Abstract: Relatively very few number of Pierre Robin syndrome patients are affected by genetic syndromes that involve oral structures or the structures associated with first brachial arch. This case report markedly emphasizes the dental management of children with Pierre Robin syndrome. A 7 years old boy had been referred to the outpatient department with complaint of pain and decayed teeth, description and management of which has been described in this case report.
Keywords: Genetic disorder, Micrognathia, Pierre robin syndrome

INTRODUCTION

Pierre Robin Sequence is a well-recognized congenital condition involving a combination of micrognathia, glossoptosis and with or without cleft palate [1]. Pierre Robin Sequence named after the French Stomatologist who, in 1923 and 1934, described the problems associated with newborn micrognathia with a prevalence of 1:20,000 [2]. The exact etiology is unknown. A possible hereditary factor has been pointed out by several authors, but after a long time this has not been striking factor [3, 4]. The primary defect lies in the arrested development of mandible leading to a characteristic bird face appearance [4].

CASE REPORT

A 7 years old boy referred to department of Pedodontics and Preventive Dentistry, Darshan dental college and hospital (Fig. 1) with the chief complaint of pain in upper left back tooth region since 4 to 5 days. The pain was continuous in nature and throbbing in type. There was no history of trauma to the area and patient had undergone the treatment of tongue surgery because of ankyloglossia 2 years back.

On intraoral examination, there was deep proximal caries i.r.t to 65, which was tender on percussion and was vital on vitality test (Fig. 2). Moreover, root stump i.r.t to 72, 74 and 82 was present and 84 were grossly decayed (Fig. 3). Other oral finding includes micrognathia, deep palate was observed. On extra oral examination, Patient had polydactyl in left hand and patient was malnourished with the weight 19.5 kg and 3.2 feet height.

On radiographic evaluation, acute periapical abscess i.r.t to 65 and radicular abscess i.r.t to 74 was found. OPG (panoramic radiograph) showed the absence of permanent tooth buds i.r.t to bilateral mandibular central and lateral incisors (Fig. 4). On laboratory investigation, decreased haemoglobin that is, around 8.02% was found. Depending on all the findings like deep palate, micrognathia, bird face appearance, polydactyl and missing permanent tooth buds were similar to the Pierre Robin Syndrome so we would like to conclude this syndrome as a Pierre Robin Syndrome.

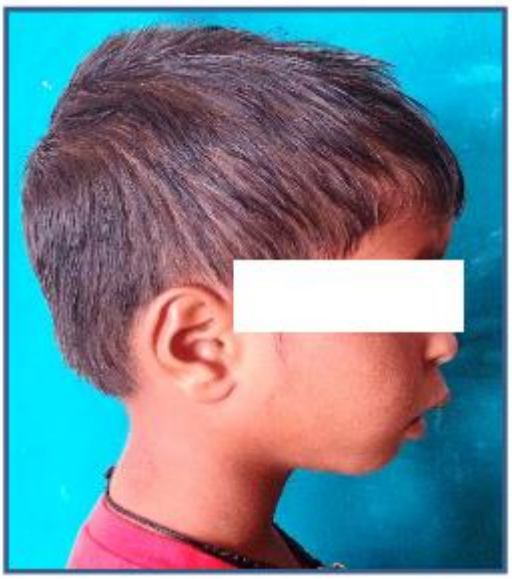

Fig. 1: Lateral view of the patient

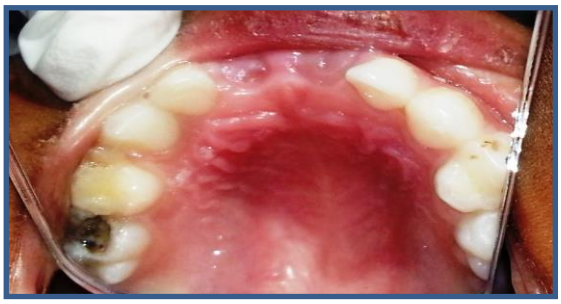

Fig. 2: Pre-operative view of Maxillary arch

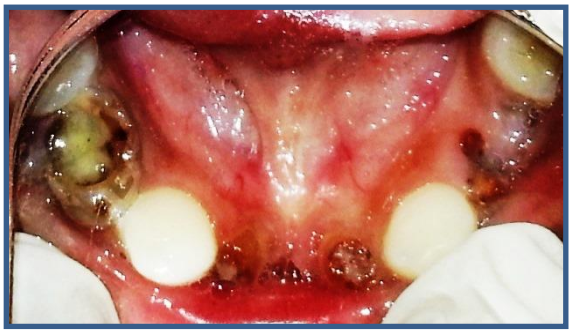

Fig. 3: Pre-operative view of Mandibular arch

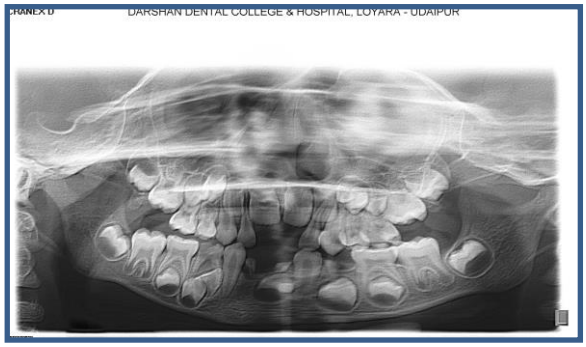

Fig. 4: OPG

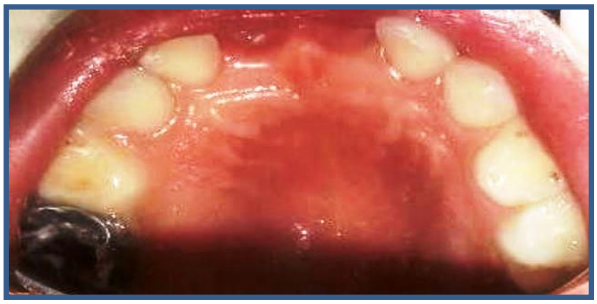

Fig. 5: Post-operative view of Maxillary arch

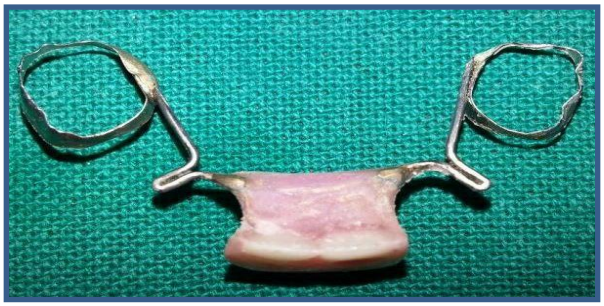

Fig. 6: Modified lingual arch with embedded acrylic teeth

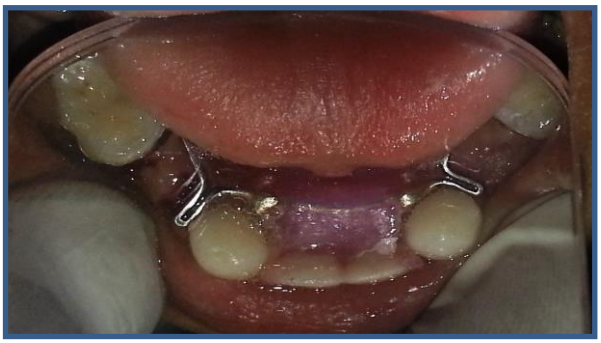

Fig. 7: Post-operative view of Mandibular arch

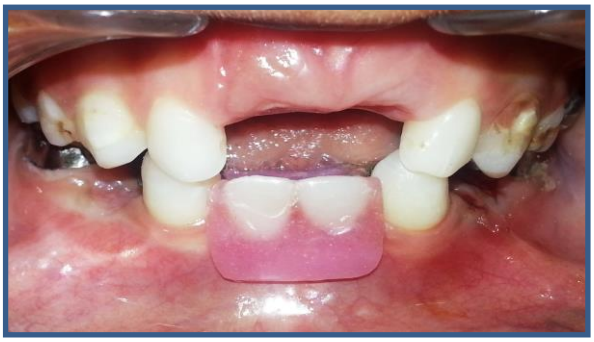

Fig. 8: Post-operative front view

After the clinical and radiographic evaluation, pulpectomy procedure was performed using metapex with 65 followed by placement of stainless steel crown (Fig. 5). Extraction was preferred with 72, 74, 82 and 84 after taking parent's consent. After the extraction of the teeth, fabrication of space maintainer with the acrylic teeth i.r.t to 71 and 81 was done to maintain space in dental arch and to address the aesthetic concern. Band pinching was performed on both 75 and 85. Crib was soldered with the anterior part of wire for better retention between acrylic portion and teeth (Fig. 6). Finally, wire component was soldered with band. These appliances serve as a space maintainer and maintain the canine in present position. Moreover, this appliance helps for aesthetic purpose (Fig. 7, 8).

DISCUSSION

The Pierre Robin Sequence is characterized by several degrees of micrognathia, glossoptosis, palatal malformation and with cleft palate [4]. However, in our case, patient shows micrognathia associated with deep palate. Shprintzen RJ [5] reported that many of the patients that carry the PR diagnostic label might in fact represent various other syndromes (Eg. Stickler's, velocardiofacial) those are associated with a genetically induced mandibular micrognathia. Bird face appearance was found in case report presented by Hegde RJ [4] and that finding was similar in our case. Moreover, polydactyl were observed in left hand and decreased haemoglobin (8.02%) was found in our case report.

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