Pierre Robin sequence and the pediatric dentist

B. N. RANGEETH, JOYSON MOSES, N. VENUGOPAL REDDY

Abstract

This article on the dental management of a neonate with Pierre Robin sequence describes the clinical and laboratory procedures for construction of a feeding plate due to the presence of a cleft palate. Emphasis has also been laid on a few literatures to describe medical complications associated with this condition. A 56-day-old neonate had been referred to the outpatient department with the complaint of difficulty in feeding, description, and management of which has been described in the case report.

Keywords: Cleft palate, feeding obturator, feeding plate, Pierre Robin syndrome, syndromic cleft

Introduction

Pierre Robin sequence (PRS (MIM 261800)) named after the French stomatologist who, in 1923 and 1934, described the problems associated with newborn micrognathia with a prevalence of 1:8500 live births. The phenomenon comprises the triad of mandibular micrognathia, U-shaped cleft palate, and glossoptosis.[1] The developmental anomaly was originally classified as a syndrome but was reclassified as a sequence in 1982.[2] PRS is a clinically well-defined subgroup of the cleft lip-palate population with an unknown etiology, often observed as a part of other Mendelian syndromes, such as Stickler’s syndrome, velocardiofacial syndrome, and Marshall’s syndrome.[3]

Recent studies on genetics have shown that that the association of dysregulation of the genes SOX9 and KCNJ2 may be involved in PRS, evidenced by a familial translocation with a breakpoint located in the gene empty region between SOX9 and KCNJ2, and by reduced expression of SOX9 and KCNJ2 in non-translocated patients with PRS.[4] Airway obstruction in PRS has been classically described to develop soon after birth.[5] However, this view was challenged by Ogborn and Pemberton, who reported that 5 of 16 (30%) of their cases presented with upper airway obstruction between 2 and 21 days of age.[6] Bull et al noted that upper airway obstruction may appear over the first month of life, specific clinical details were not provided.[7] The late onset of upper airway obstruction, particularly after initial hospital discharge, has potentially serious consequences. Caouette-Laberge et al reported deaths by sudden deterioration in the degree of upper airway obstruction in the PRS.[8] Clinical grounds alone are clearly not sufficient to predict whether an infant will subsequently present with upper airway obstruction, suggesting that there is a role for polysomnography as a diagnostic tool or continuous overnight pulse oximetry. All infants with PRS clearly need close clinical monitoring with particular attention paid to weight gain.[9] This report highlights an infant diagnosed with PRS, for whom we constructed a palatal feeding obturator for feeding required for the proper growth and development and also to prevent other associated complications.

Case Report

A 56-day-old full-term male infant [Figure 1] diagnosed with PRS was referred to the outpatient department with the complaint of difficulty in feeding. The child was born out of non-consanguineous marriage to healthy parents. The attending physician of the Pediatric Intensive Care Unit in a private hospital referred the patient to the Department of Pedodontics because of poor swallowing ability due to which a nasogastric intubation was advocated. Following a period of using the nasogastric intubation, the mother was advised to expel the milk into a feeding bottle and then feed the child as the oroantral communication could not facilitate breast feeding. Nasal regurgitation was still evident on bottle feeding; hence, the necessity for dental management arose.

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Extra-oral examination revealed a retruded chin and ocular lesions. Intra-oral examination of the baby revealed a large median U-shaped cleft of the hard palate that extended to the soft palate with only the alveolus in the right and left side [Figure 2]. The nasal septum and conchae was visible. Since the baby was not scheduled for corrective surgery in the near future, it was decided to construct a feeding obturator.
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Following approval from the parents so as to prevent nasal regurgitation or aspiration of milk into the trachea.

Preliminary impression was done with impression compound using the index finger to mold the impression material. The process of the child’s suckling reflex when the index finger was placed also aided in the molding of the impression compound. A special tray [Figure 3] was constructed using self-cured acrylic resin that was finished and polished to prevent any injuries. The tray was coated with a tray adhesive and the impression was made using addition silicone impression material [Figure 4]. The impressions were made with the infants face held toward the floor, in order to prevent aspiration in the event of vomiting and asphyxiation due to airway obstruction.

The feeding obturator [Figure 5] was constructed using heat polymerizing acrylic resin with two holes for tying a dental floss for handling and positioning the obturator. The infant was made to wear the obturator, with the floss around the ears [Figures 6a and b] following trimming and polishing to correct the extent as well as to prevent injuries due to sharp margins. The parent was made to breast feed the child and as advised review after 24 h, 1 week, and 1 month later. The parent was advised to clean the obturator after every feed. The parent and child showed good prognosis using the feeding obturator and nasal regurgitation of milk was not to be noticed.
Discussion

Historically, cleft palate has not been regarded pathognomonic of the PR population, and even today some authors regard this feature as optional. At the other end of the spectrum, some authors regard only children with a typical long, U-shaped cleft as being truly PR (Shprintzen, 1988). PRS, in association with other genetic syndromes, may carry a very high recurrence risk. In the case of PR anomalad, the primary defect lies in the arrested development and ensuing hypoplasia of the mandible, ultimately producing the characteristic “bird facies.” This in turn prevents the normal descent of the tongue between the palatal shelves, resulting in a cleft palate. Airway obstruction and feeding difficulty associated with PRS require invasive therapeutic measures. Upper airway obstruction and feeding difficulties not resolved by prone positioning and placement of an indwelling nasogastric tube should be considered before an invasive surgical procedure is undertaken. The use of feeding tubes is started early but are said to be responsible for most of the recorded perforations as they are soft at first and become hard and stiff after several hours of use. Rare complications such as urinary bladder perforation, pericardial sac perforation, and Enterobacteriaceae colonization on prolonged feeding expose the risks associated with using intubation for feeding.

Considering the risks, it is best to advocate other techniques to aid in long-term solutions for feeding. We had decided upon one of the successful techniques of using a feeding obturator for the infant. The use of the feeding obturator has also been described in a preterm infant born at 37th week of pregnancy showing the effectiveness and safety in its use. Other feeding facilitation techniques in the form of suction of a pacifier (non-nutritive sucking technique), massage to anteriorize and relax the tongue, support for sustaining the mandible, techniques changes in the feeding bottle nipple, rhythmic movement of nipple in the oral cavity during nutritional sucking, positioning of bottle nipple exactly on the tongue have been analyzed and have been effective in improving the oral feeding acceptance, increase in the volume of milk ingested and greater safety during the process of oral feeding. These techniques have been used during admission following requirement of post-delivery care.

The palatal cleft interferes with nursing and causes regurgitation of food through the nose. Infection of the nasopharynx and chronic catarrh are frequent. Some of the features noticed are involvement of the Eustachian tubes, Otitis media, hearing impairment in 30–40%, and the patient may become permanently deaf and dumb. Labirintitis with the disturbance of equilibrium has been observed. Bronchitis and pneumonia may complicate local infections. Difficulty in speech becomes more and more marked as time progresses, the patient having difficulty with guttural and labial sounds. Hence, the construction of the palatal feeding obturator for nursing is very important until the surgical correction of the defect is carried out. The time of corrective surgery for the best result is closely related to the method and technique employed.

Conclusions

The development of cleft palate no longer seems to be a pit fall in the development of the child. A multispecialty approach that our patient had undergone for management has proven to be successful in improving the health for surgical correction. The risks associated with general anesthesia have also been reduced with techniques such as capnography-guided awake nasal intubation to deal with difficult airway situations. The basic criterion of physical health is necessary prior to the surgical approach, achievable only with intake of mother’s feed in infants and our approach was based on this principle.

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