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Feeding Difficulties and Management for Neonate Afflicted with Pierre Robin Syndrome – A Case Report

Dental Science

KEYWORDS: Pierre robin syndrome, cleft palate, feeding bottle, prosthetic attachment.

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ABSTRACT

This paper reports a case of 3-day old neonate with difficulty in feeding, born with Pierre robin syndrome and cleft palate. Babies with cleft palate, faces feeding problem which interfere with their growth and development, also endure daily suffering during feeding. Obtaining a good seal of oral cavity for swallowing and palatal support for feeding bottle can be difficult in such patients. To overcome these difficulties and discomfort a feeding bottle attachment was fabricated and successfully used for feeding the patient. The design of attachment was subsequently refined, improvised by author for further trials.

INTRODUCTION

Pierre Robin Syndrome or Sequence (PRS (MIM 261800)) named after French stomatologist who in 1923 and 1934 described the problems associated with new-born micrognathia with a prevalence of 1:8500 live births. This phenomenon comprises triad of mandibular micrognathia, U-shaped cleft palate, and glossoptosis1. This developmental anomalad was originally classified as a syndrome but reclassified as a sequence in 19822. During fetal development, at nine to eleven weeks of gestation, if head of fetus did not raise at cephalic flexure at proper time, tongue would be positioned between folds of soft palate and prevent their natural union[3].

PRS is a well-defined subgroup of cleft palate population with unknown etiology, often observed as a part of other Mendelian syndromes, such as Stickler's syndrome, Velocardial syndrome and Marshall's syndrome. Some external factors or neurological conditions, which limit the fetus and interfere with growth of lower jaw, may contribute to PRS4. Recent studies on genetics have shown that, association of dysregulation of genes SOX9 and KCNJ2 may be involved in PRS5.

Neonates with PRS suffer from two main problems: airway obstruction and feeding difficulties6. Nursing time is longer, choking and regurgitations are also common. This can lead to anxiety and concern of parents and/or caregivers[7].

Presented here is a case report of a neonate afflicted with PRS for whom a feeding plate attachment was fabricated. Easily available material and inexpensive method was used to connect the attachment with a normal feeding bottle. Condition of patient and subsequent feeding experience was closely followed by authors and treating pediatrician based on visual monitoring and feedback given by the caregiver.

CASE REPORT

A 3-day-old full term male neonate was referred to Department of Pediatric Dentistry, by attending pediatrician of Neonatal Intensive Care Unit in a private hospital, with chief complaint of difficulty in feeding, inability to swallow, nasal regurgitation. Mother was advised to expel milk with breast pump and with the help of a syringe feed child through nasogastric tube. Nasal regurgitation was still evident; and it was difficult for mother to continue feeding child like that and hence there was necessity for customized cleft palate management.

On extra-oral examination, bird face appearance, receded chin, retrognathic mandible and convex facial profile were evident (Figure 1). On intra-oral examination a large isolated palatal cleft with tongue falling back into the palatal defect was seen (Figure 2). Also small whitish pearl like spots were present on either side of palatal defect and on upper gum pads suggestive of Epstein pearls and Bohn's nodules respectively (Figures 3(a), (b)). Medical and family history of child was non-contributory. Based on these clinical findings, a diagnosis of PRS was made. On examination by oral & maxillofacial surgeons, surgical correction of cleft palate was planned at age of 18 months. For purpose of ease of feeding and to prevent regurgitation or aspiration, it was decided to fabricate an "Attachment for feeding bottle" to be used till the time of surgical correction of cleft palate.

Primary impression of palate was made using a small plastic spoon, dipped in self-polymerising resin for strength, as an impression tray, with its convex surface facing defect. Polyvinyl siloxane elastomeric impression material of soft putty consistency was loaded on convex surface of spoon, to make primary impression. Then child was held upright with face towards floor, in order to prevent aspiration in event of vomiting and asphyxiation due to airway obstruction. Spoon carrying impression material was placed into oral cavity and held with very gentle pressure just to make sure to cover periphery of defect and not to record depth of the defect. Once sufficient suction was achieved, spoon was removed from mouth. A dental stone model was obtained from the impression (Figures 4(a), 4(b)). Customized self-cured acrylic feeding plate attachment was fabricated. All borders of feeding plate attachment were rounded and small holes were made to increase retention of lining material. Permanent soft denture relining material was loaded on convex surface of customized plate and final impression was made, using the steps followed during primary impression.

A normal feeding bottle was taken and a hole was made in rim carrying the nipple, hole was adjusted according to handle of feeding plate attachment. Feeding plate attachment was attached to a feeding bottle over the nipple in order to cover intraoral defect while feeding with bottle (Figure 5). Feeding bottle was checked in infant's mouth (Figure 6). Instructions regarding insertion, removal and cleaning of feeding bottle were given to parents. Child was reviewed after 24 hours to observe any signs of ulceration or irritation. During follow-up visit after 24 hour, 1 week and 1 month later, it was observed that child had accepted feeding bottle well and mother reported ease of feeding with significant reduction in nasal

regurgitation.

DISCUSSION

Robin in 1923 published a description of neonates with micrognathia, glossoptosis, and respiratory distress. Later, in 1934 he added cleft palate (CP) as an aggravating factor. Since 1960s, triad of micrognathia, glossoptosis, and CP has been established as PRS9. In this anomalad, primary defect lies in the arrested development, hypoplasia of mandible, producing characteristic bird-facies10,11. According to Lidsky et al, incidence of PRS in general population is rare 1/8500 (0.01%) 12. Thakkar et al reported that majority of neonates with PRS suffer from two main problems: airway obstruction and feeding difficulties. The palatal cleft interferes with nursing and causes insufficient suckling or aspiration/nasal regurgitation of food. Coughing, chocking, vomiting, infection of the nasopharynx and chronic catarrh is frequent. Otitis media may result in 30-40% of afflicted infants, leading to hearing impairment or permanent deafness. Labyrinthitis with disturbance of equilibrium has also been observed. In general, death from PRS is thought to be as a result of poorly controlled combined effects of obstructive apnea and failure to thrive[13].

Various literature reviews, methods adopted to overcome feeding difficulties and upper airway obstruction in cleft lip and palate babies, with some adjustments to feeding methods and positioning14. Reid et al. reviewed and stated that feeding modalities should proceeded by altering the positioning of the baby. In all feeding practices, baby should be kept in an upright position, with head in one hand and bottle in another, and fed at an angle of 450 with frequent burping of the child. This allows the milk to flow down and helps to prevent choking; otherwise, milk may enter the respiratory passage15. Devi et al discussed different feeding methods like; breast, bottle, spoon or cup feeding or a combination may be chosen with lactation instructions pertaining to correct positioning of the child. Speciality feeding products like supplement nursing system or special feeding bottles like Mead Johnson, Haberman feeder, soft squeeze baby bottle. But these special products have drawbacks like high-cost, leakage around the nipple and difficulty in sterilization16. Prodoehl DC and Shattuck KE stated the placement of an in-dwelling nasogastric tube in case upper airway obstruction and feeding difficulties not resolved by prone positioning. The use of feeding tubes is 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 17. Habel A et al reported that feeding obturators have been a major area of controversy18. According to Savion I et al. there are two schools of thoughts. The first one advocates its advantages as ease in feeding, facilitation of cleft repair, enhancement of speech and facial growth. The other group claims its disadvantages such as cumbersome impression taking procedure, difficulties associated with sterilization and hygiene maintenance, expensive manufacture and effort involved in training the caretaker about effective intra-oral attachment of prosthesis19. Lehman JA et al suggested that in cases where positioning is not helpful, tongue-lip adhesion or mandibular distraction is done. In severe cases, tracheostomy is also advocated[20].

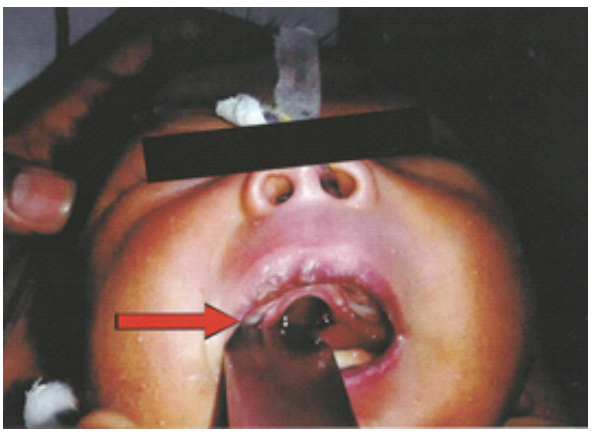
The feeding plate attachment designed and refined by the author is easy to attach to a normal feeding bottle for feeding infants having cleft palate disorder. This attachment may find its advantage by preventing flow of milk from mouth to nasal cavity, as it covers the defect. It also eliminates the need for caretaker to hold the infant in upright or near upright position while feeding. It also helps in enabling the infant to create enough suction to draw milk from feeding bottle. This device may be attached to feeding bottles of different dimensions. Further, this device is under the process of refinement in terms of material and size for trials.

CONCLUSION

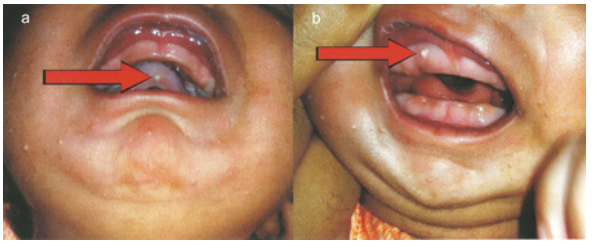
This case illustrates the feeding difficulties faced by PRS infants and others afflicted with cleft palate. Till date, not much data is available in literature about clinical efficiency of various specialized feeding modalities available in market. In this case, a feeding bottle attachment was fabricated and successfully used for feeding which was well accepted by patient and caretaker. Subsequently, design has been refined by the author for further trials as an attachment to a normal feeding bottle.



Figure 1- extra-oral examination showing, bird face appearance, receded chin, retrognathic mandible and convex facial profile



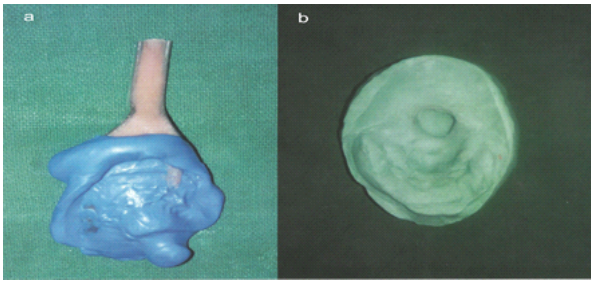
Figures 2- intra-oral examination showing a large isolated palatal cleft with tongue falling back into the palatal defect



Figures 3(a) – arrow showing small whitish pearl like spots were present above the palatal defect, in mid-palatine region, suggestive of Epstein pearls.

3(b) - arrow showing small whitish pearl like spots on upper gum pads suggestive of Bohn's nodules.

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Figures 4(a)- Elastomeric impression in plastic spoon strengthened with acrylic resin.

4(b)- Dental stone cast.



Figures 5- Feeding plate attachment with normal feeding bottle.



Figures 6 - Feeding plate attachment with bottle checked in infant's mouth.

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