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Case Report

Management of Feeding Problems in Infants with Cleft Palate and Review on Pierre-Robin Syndrome

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ABSTRACT

Pierre Robin syndrome is a congenital disorder with triad of features micrognathia, glossoptosis which result in airway obstruction and feeding difficulties and cleft palate. Cleft palate is the most common congenital anomalies of craniofacial region seen in Pierre-Robin syndrome. Infants with cleft palate have to face various problems. Difficulty in feeding is the most common problem faced by cleft palate neonates that made them difficult to maintain adequate nutrition, result in failure to thrive. There are many methods given in literature to overcome this problem including special type of nursing bottles. In this case report, fabrication of feeding plate to obturate the defect in palate and its use in managing feeding problem is described.

Keywords: Cleft palate; Feeding plate; PierreRobin Syndrome

1 INTRODUCTION

Cleft palate is the most common congenital anomaly having incidence of 0.28 to 3.74 per 1000 live birth globally.[(1)] Cleft lip and/or cleft palate are structural abnormalities that occur in the embryonal period of life between the 4th and 10th weeks.[(2)] The etiology of cleft is still unknown, but both genetic and environmental factors may be responsible for many congenital malformations that can result from the developmental process failing to reach some developmental end point or threshold.[(3)] The cause of the familial aggregation of the disease may be genetic, environmental, or both. Poor nutrition, tobacco smoke, alcohol, viral infection, and medicinal drugs are among the most important etiological factors. The environmental risk factors are also to be equally important. Interactions between maternal and foetal genes are significant in the etiology of the disease.

Cleft palate also affects several systems and functions that include facial growth, dentition, speech, hearing and genetic aspects because of the complex mode of inheritance. Child born with cleft have many problems that need to be solved for successful rehabilitation by a multidisciplinary team approach.[(4)] Management of the patient with a cleft begins with immediate attention to the need of the newborn. Feeding problems associated with cleft make it difficult for the infant to maintain adequate nutrition. These problems include difficulty in sucking, excessive air intake during feeding, choking, nasal discharge, and excessive time required for feeding.

The feeding plate / feeding appliance obturates the cleft and closes the defect between oral and nasal cavities. It creates a rigid platform towards which the baby can press the nipple and extract the milk.[(5)] According to GPT-9, Feeding Appliance is defined as "a medically prescribed prosthesis that closes the oronasal cavity defect, thus enhancing sucking and swallowing, and maintains the right and left maxillary segments of infants with cleft palates in their proper orientation until surgery is performed to repair the cleft."[(6)]

The obturator also acts as a barrier between the tongue and the cleft so that there will be no interference with the

spontaneous growth of palatal shelves towards the midline. It also helps to position the tongue in correct position so that it can facilitate normal jaw growth and speech development.

The obturator prevents nasal regurgitation thus reducing the incidence of otitis media and nasopharyngeal infections.[7] Feeding plate restores the basic functions of feeding, swallowing and speech production until the cleft lip and/or palate can be surgically corrected. The steps involved in fabrication of feeding appliance are described in this case report along with review on Pierre-Robin syndrome.

2 PIERRE-ROBIN SYNDROME

In 1923, Pierre Robin, a French stomatologist, documented a disorder with a triad of features micrognathia (which he termed "mandibular hypotrophy") and glossoptosis (an abnormal posterior placement of the tongue), which results in airway obstruction and feeding difficulties labeled as "Pierre Robin Sequence" (PRS) on his name.[8,9] The small mandible is thought to be due to an inherent genetic problem when intrauterine growth is altered. Some controversy exists in the literature regarding the cleft palate as a feature of the triad of Pierre Robin. In Pierre Robin sequence, the primary developmental disturbance is the micrognathia (true hypoplasia of the mandible should be distinguished from the retrognathia i.e. backward positioning of the mandible; Shprintzen, 1992) leading to a backward positioning of the tongue, obstructing the fetal closure of the secondary palatal shelves, and further causing respiratory problems by mechanical obstruction of the oropharynx (Hanson and Smith, 1975; Cohen, 1999). Rather than a syndrome (multiple anomalies from a single pathogenesis), it is important to note that PRS is a sequence, where multiple anomalies result from a sequential chain of malformations. In PRS, the micrognathia leads to glossoptosis, which in turn results in airway obstruction and difficulty to feed.[10]

Despite the normal size of the tongue, the small mandible provides less volume in the oral cavity and forces the tongue to fit into a smaller space that leads to the blockage of the posterior pharynx results in airway obstruction.[8,9] Infants with PRS may have an airway obstruction at the level of the tongue base.[11] To combat this obstruction, more energy is required to continue breathing and is manifest by suprasternal retractions and the use of accessory muscles of respiration. Some infants may maintain their airway when awake but suffer from obstruction during sleep, especially in the supine position depending on the severity of airway obstruction,[11] Difficulty in breathing, gastro esophageal reflux and aspiration are common sequelae of this process as infants struggle to breathe. The associated cleft prevents the formation of negative intraoral pressure, which is required to suck milk from the breast or bottle; further the micrognathia and glossoptosis also impede mechanical sucking.[11,12] Airway obstruction and resulting negative intrathoracic pressures have been identified as factors associated with increased gastro esophageal reflux.[13] Given the poor caloric intake associated with reflux and difficulty in feeding and the increased respiratory effort driving increased energy expenditure, these infants often fail to thrive and are unable to gain weight during the early postnatal period.

There has been some controversy about whether to include children with other well-known syndromes in the populations of Pierre Robin. For instance, children which can actually have all three features of the Pierre Robin triad, and some authors include these but others do not (Cohen, 1989; Shprintzen, 1992). Cleft palate is also not considered as one of the common feature of PRS. Oligohydramnios has been suggested as the cause of micrognathia by physical compression of the fetal mandible, subsequently leading to cleft palate and respiratory distress. Even generalized intrauterine impairment like fetal alcohol syndrome can include the Pierre Robin triad (Cohen, 1989).[10]

Pierre Robin sequence is related to several other craniofacial anomalies and may appear in conjunction with the findings characteristic of other syndromes. Stickler syndrome, an autosomal dominant condition, is characterized by a short mandibular ramus, antegonial notching of the mandibular body, myopia, and joint problems.[8] Velocardiofacial syndrome is characterized by a retrognathic mandible, palatal abnormalities, hypotonia, impaired thymus development, and cardiac malformations.[14,15] Craniofacial microsomia (or oculoauriculovertebral spectrum) results in anomalies of the external and middle ear along with the involvement of mandible, zygoma, maxilla, temporal bone, facial muscles, and palate.[9] Treacher Collins syndrome is notable for dysplasia affecting the zygoma, temple, ear, and mandible. It is further characterized by downslanting palpebral fissures, lower lid colobomas, facial bone hypoplasia, malformation of the external ear, macrostomia, and a high-arching palate. Some have likened this to Tessier 6, 7, and 8 facial clefts.[8]

3 MANAGEMENT

3.1 3.1 Nonsurgical Management

Prone or lateral positioning will solve the airway obstruction in 70% of cases of PRS.[9,13] With appropriate positions, feeding problems also resolved. If airway is not patent, then placement of a nasopharyngeal (NP) tube is indicated. Chang et al have discussed the technique of creating individualized NP tubes from endotracheal tubes, with size (diameter and length) chosen according to the infant's weight.[16,17]

If the problems are not resolved conservatively with PRS infants, further intervention is required. Supplemental oxygen, nasopharyngeal tubes, laryngeal masks, and prolonged intubation are temporary measures; inadequate in infants with severe respiratory distress and surgical management of airway obstruction is required. The most common methods for surgical management of airway obstruction

include tongue–lip adhesion, distraction osteogenesis, and tracheostomy.

Tongue–Lip Adhesion (TLA) was first described by Shukowsky in 1911 and popularized by Douglas in the mid-20th century.[18] This procedure corrects the problem of glossoptosis by pulling the base of the tongue forward and suturing it to the lower lip. Once healed, this mucosal attachment serves to tie the tongue anteriorly until the infant develops a more stable airway with growth. TLA can only be performed on infants who have not developed any lower teeth, as they could otherwise bite through the repair inadvertently. After adequate growth has occurred, the TLA released with a second procedure.

Distraction Osteogenesis of the Mandible - Distraction osteogenesis (DO) of the mandible, first described in 1989 by McCarthy, has become popular as the definitive technique to address the issues associated with PRS by relieving airway obstruction, improving facial cosmosis, and correcting malocclusion.[19] The process of mandibular distraction lengthens the jaw in a forward direction, and also indirectly pulls the tongue base anteriorly. Thus, this technique reverses the sequence of PRS by correcting the micrognathia, which in turn improves the glossoptosis, and thereby relieving the obstruction of the airway.

4 CASE REPORT

14 days old infant (Female) with her parents reported to the Department of Prosthodontics, Faculty of Dental Sciences referred from Department of Paediatrics. Her mother complains of baby's inability in feeding milk and nasal regurgitation during feeding milk. The infant's medical history was taken from her parents, who reported no similar congenital or genetic anomaly in the family. On examination, it was found that patient having cleft palate involving hard & soft palate both (Group-II cleft palate class according to Veau's classification 1932) [Figure 1].

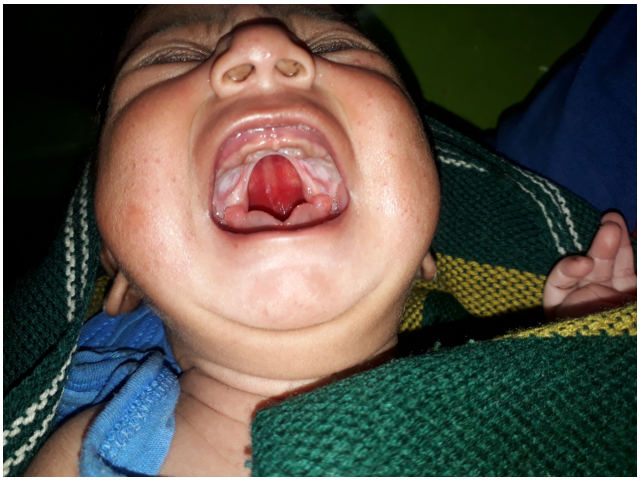

Fig. 1: Pre operative view (Group-II cleft palate class according to Veau's classification 1932)

After complete examination of the infant, fabrication of a feeding plate was decided on to reduce feeding problem. The proposed treatment plan was explained to the parents and informed consent was taken. Preliminary impression of the palate was made with soft modelling wax. First the modelling wax was softened in warm water. With a finger it was adapted into the baby's mouth and pressed the soften wax against the hard palate and into the buccal and labial vestibules, while the baby was held in prone position in the mother's lap, in order to prevent aspiration in the event of vomiting and asphyxiation due to airway obstruction. The impression was inspected thoroughly. A primary model was prepared by pouring the impression in dental plaster (type-II gypsum product). The special tray was fabricated using auto polymerizing acrylic resin. [Figure 2] Special tray was evaluated intraorally, adjustment was made and determined the easiest path of insertion; final impression was made using heavy body Addition Silicone impression material. A master cast was poured using die stone [Figure 3]. Then, processing was done by following the protocol of compression moulding technique and conventional heat cure acrylic resin material was used. After completion of curing cycle, the feeding plate was retrieved from the flask and proper finishing & polishing was done. Dental floss was attached to the feeding appliance to prevent aspiration and easy retrieval of appliance. [Figure 4]. Finally, appliance was placed in infant's oral cavity and infant was easily fed with the appliance. (Figures 4 and 5).

Fig. 2: (A) Primary impression made using soft modelling wax (B) & (C) primary cast in dental plaster (type-II gypsum product) and fabrication of special tray was done using auto polymerising acrylic resin

Instructions were given to the parents on how to insert, remove and clean the prosthesis. Parents were given instructions regarding how to use the obturator during feeding time, remove it afterwards, and thoroughly clean the baby's oral cavity and cleft with a soft cloth/ gauge/ cotton soaked in warm water. The patient was seen after 24 hour later for adjustment. Then, patient was followed up regularly after 3 month interval.

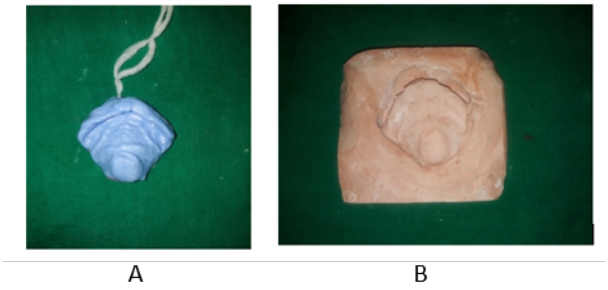

Fig. 3: (A) Final impression was made using vinylpolysiloxane (addition silicone putty) and (B) poured in die stone (type IV gypsum product)

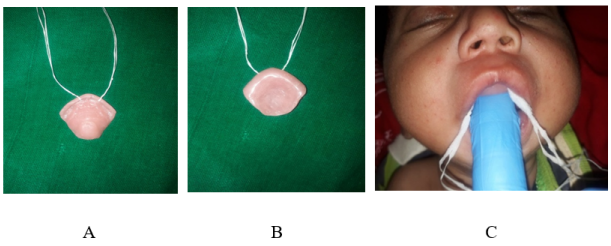

Fig. 4: (A) & (B) Feeding plate fabricated using heat cure acrylic resin (C) feeding plate installed

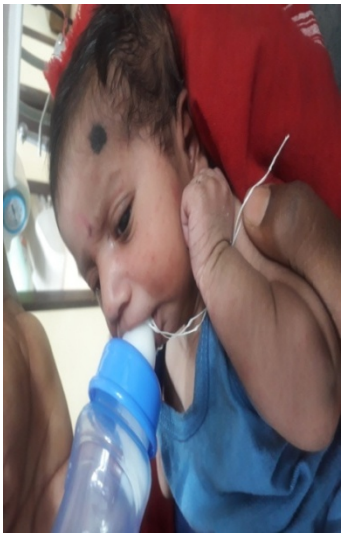

Fig. 5: Infant feeding using nursing bottle.

5 DISCUSSION

The management of infants born with cleft palate is best accomplished by the multidisciplinary team approach in which a Prosthodontist plays an important role in the team which is working closely with medical and allied health specialties. A feeding plate is used as an adjunct in nourishment and prevention of infections for the already debilitated infant. It creates a rigid platform, against which the infant can press the nipple and feed, reduces nasal regurgitation, reduces the time required for feeding, and helps in positioning the tongue away from the cleft area in the correct position so that there will be no interference in the growth of palatal shelves towards each other, it further reduces parent's frustration as a result of feeding problems.

The impression procedure carried out here is easy to follow. Proper care should be taken by the operator during the impression making procedure to prevent aspiration. It was also ensured that the infant made sucking motions during impression-making as this helps in better moldability. The feeding plate should be thoroughly examined for the blebs or any rough surfaces to prevent discomfort to the baby. The feeding appliance should be light in weight and good fit to palate and ridges and decreased possibility of soft tissue injury. [20]

6 CONCLUSION

A multidisciplinary team approach is required in managing cleft palate patients so that the problems experienced by cleft palate patients get reduced and keep the patient under regular review till the final management. This case report describes the role of Prosthodontist in fabrication of feeding appliance for the cleft palate patient that helps the infants in feeding, stimulate oral-facial development along with palatal shelves and expand the collapsed maxillary segment. It also provides psychological help to the parents.

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