Case Report

Pierre robin syndrome: Rehabilitation of a neonate for nursing with palatal obturator

N. V. Thakkar, M. J. Bhatt[1]
Ahmedabad Dental College, Santej, [1]Consultant ENT Surgeon, Ahmedabad, India

For correspondence
Nipa V. Thakkar, Benivila, Near Swaminarayan Temple, Vasna Barrage Road, Vasna, Ahmedabad - 380 007, India.
E-mail: drnipabhatt@yahoo.co.in

Pierre-Robin Syndrome or anomalad is a congenital abnormality characterized by cleft, micrognathia and glossoptosis. Feeding of a neonate with a complete cleft palate is a very difficult task. A multidisciplinary approach is required to manage the complex features involved in the case of such children and their families. Here, we are presenting a case of a one day old neonate having Pierre-Robin syndrome with complete cleft palate in whom we constructed a palatal obturator for feeding.

Key words: Pierre-Robin syndrome, cleft palate, micrognathia, feeding plate, congenital defects, ppalatal obturator

INTRODUCTION

Pierre-Robin syndrome is considered to be a non-specific anomalad which may occur either as an isolated defect or as a broader group of malformations. The isolated defect is considered a sporadic or non-genetic condition with a very low recurrence risk in the family. In contrast, Pierre-Robin syndrome, in association with other genetic syndromes, may carry a very high recurrence risk. In the case of Pierre-Robin anomalad, the primary defect lies in the arrested development and ensuing hypoplasia of the mandible, ultimately producing the characteristic 'bird facies'.[1] This in turn prevents the normal descent of the tongue between the palatal shelves, resulting in cleft palate.[2] Because of this mechanism cleft lip is not associated with cleft palate. A single midline cleft of the palate is due to lack of fusion of the palatal processes; it may vary in extent from a bifid uvula to a cleft which involves both the soft and hard palate resulting in a complete cleft palate. Other systemic findings may also be present including congenital heart defects, other skeletal anomalies and ocular lesions. In addition, mental retardation also may be present in a significant number of these patients. Here we are presenting the case of a one day old neonate with Pierre-Robin syndrome,in whom we constructed a palatal obturator for feeding required for the proper growth and development and also to prevent other associated complications.

CASE REPORT

A one day old male child with Pierre-Robin syndrome was referred from ENT and pediatric dept. to Dental dept. On extra-oral examination, the neonate showed receded chin, ocular lesion and club feet [Figure 1]. On intra-oral examination, the palate was almost totally absent with only the alveolus present in right and left sides, with the cleft extending to the soft palate. The nasal septum and chonchae were visible. Feeding of the patient was carried out with the help of a Ryles tube [Figure 2].

The child had been examined by ENT and plastic surgeons and surgical correction was planned at the age of 18 months for cleft palate. For the purpose of feeding the child intra-orally, a palatal obturator was necessary to prevent nasal regurgitation or aspiration of milk into the trachea.

For the construction of an obturator, impression of the palate was taken with dental impression compound. First, the impression compound was softened and placed on the finger and allowed to lose a little heat. The child was made to cry and the finger was inserted into the oral cavity with the compound. Once the child started sucking on the finger and the compound hardened preliminarily, the finger was removed from the mouth and the compound chilled with tap water. A Plaster of Paris model was prepared by pouring the impression. Wax pattern was prepared on the plaster model. S.S. wire bent in the form of a 'U' was

inserted in the wax pattern for the purpose of obtaining a handle. Then, the wax-pattern was invested and converted into heat-cure acrylic. All the borders of the obturator were rounded and polished in order to avoid trauma [Figure 3]. Post-insertion adjustments were done by trimming excessive amounts of acrylic from the nasal surface inducing gagging until the obturator was comfortably accepted by the child and polishing on all the sides was completed [Figure 4]. Proper instructions regarding the use were given to the mother.

DISCUSSION

The anomaly is due to failure of various embryonic processes of the upper jaw to unite. Of various other theories, faulty pre-natal nutrition and mechanical interference are given the greatest prominence. Palatal defects are associated even with other syndromes.[3] The palatal cleft interferes with nursing and causes regurgitation of food through the nose. Infection of the nasopharynx and chronic catarrh are frequent. Often, through the involvement of the Eustachian tubes, the ears are affected. Otitis media may result and in 30-40% of the afflicted, hearing is impaired and the patient may become permanently deaf and dumb. Labirinthitis with the disturbance of the equilibrium has been observed. Bronchitis and Pneumonia may complicate local infections. The infant becomes undernourished and underweight and often is emaciated. The impediment in speech becomes more and more marked as time progresses, the patient having difficulty with guttural and labial sounds. On account of deafness, the speech defect is not easily corrected in older patients even if the operation is successful. Hence, the construction of palatal obturator for nursing is very important until the surgical correction of the defect is carried out. The time of operation for the best result is closely related to the method and technique employed.[4-6]

Figure 1: Pre-operative photograph

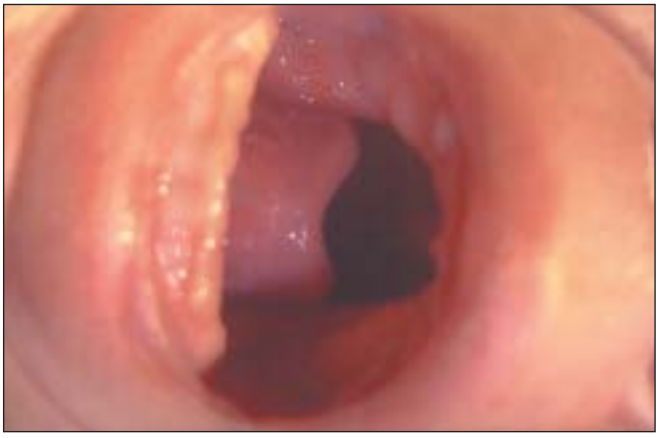

Figure 2: Internal defect

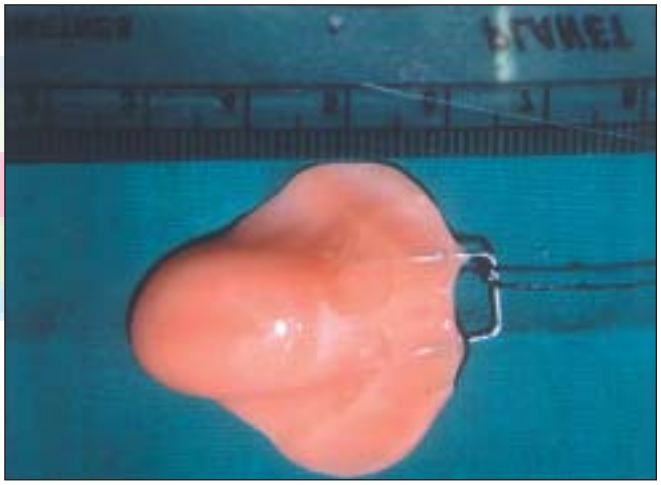

Figure 3: Palatal obturator

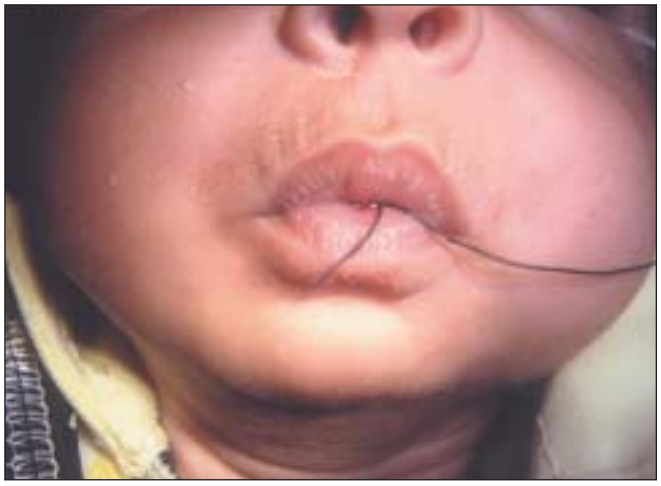

Figure 4: Post-operative photo of patient Pierre-Robin syndrome

REFERENCES

1. Kennedy JM, Thompson EC. Hypoplasia of the mandible (Pierre–Robin syndrome) with complete cleft palate: Report of a case. Oral Surg 1950;3:421.
2. Maue–Dickson W, Dickson DR. Anatomy and Physiology related to cleft palate: Current research and clinical implications. Plast Reconstruct Surg 1980;65:83–90.
3. Cohen MM Jr. Syndromes with cleft lip and cleft palate. Cleft Palate 1978;15:306.
4. Lewis MB. Timing and technique of cleft palate repair: *In*: Mash JG. editor. Current Therapy in Plastic and Reconstructive Surgery.Vol.1, Mosby year book: 1989.
5. Peet E. The Oxford technique of cleft palate repair. Plast Reconstruct Surg 1961;28:282–99.
6. Perko M. Two stage palatoplasty. *In*: Bordach J, Morris HL, editors Multidisciplinary Management of Cleft lip and Palate. 1st edn. W.B. Saunders & Co; 1991. p. 317–20.

Abstracts

Effect of water storage on the silanization in porcelain repair strength

T. Berry, N. Barghi, K. Chung

Source: Journal of Oral Rehabilitation, Volume 26, Number 6, June 1999, pp. 459-463(5)
Publisher: Blackwell Publishing

This study examined the long-term water storage affect of silanization on shear bond strength of composite resin to porcelain. One hundred and sixty square-shaped specimens were fabricated and sanded flat sequentially with silicone carbide papers. The specimens were then placed into four groups and 16 subgroups of 10 specimens each randomly. Four commercially available silane systems, two one-mix and two two-mix, were tested in this study. Teflon tubes with an internal diameter of 2·97 mm and 2 mm in height were filled with a dual cure composite resin (Mirage® FLC), placed on the silanated surfaces and light-cured for 120 s. Specimens were stored in room temperature water and subjected to shear bond strength testing after 24 h, 1 week, 1 month and 3 month periods of immersion. An Instron Universal testing machine with a crosshead speed of 0·5 mm/min was used for the testing. The mean values of the shear bond strengths ranged from 4·38 MPa (24-h period) to 23·90 MPa (3-month period). anova and Scheffe' tests were used to analyse data with confidence level at 95%. All groups recorded an increase in bond strength after one week as compared with the 24-h period (P0·05). With the exception of a one-mix system, all systems showed significantly higher bond strength at 3 weeks as compared with the 24-h and 1-week water storage periods. In conclusion, bond strength of composite resin to porcelain resulting from silanization of porcelain increased during the experimental period. The bond strength also varied for different silanes used in this study.