A Successful Effort of Corrective Intervention in Deviated Growth by Nature Leading to Survival of an Infant by Pediatric Dental Professionals: A Case Report

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ABSTRACT

Introduction: Pierre Robin Sequence (PRS) is a congenital condition constituting of a classic triad of Micrognathia, Glossoptosis and Cleft palate. The primary defect lies in the arrested development of mandible leading to retro displacement of the tongue and reduction of oropharyngeal airway. In general, mishaps from PRS are due to the combined effects of poorly managed airway and obstructive apnoea. Approximately 3% of child patients with clefts of palate do not have multifactorial inheritance.

Case report: This case report presents the sequence of treatment procedures followed to enable an infant with PRS to breathe freely without any artificial aid. Initially the infant was managed with Nasopharyngeal airway (for 10 days but patient suffered from aspiration pneumonia) after which Glossopexia was attempted (for the next 3 days, mild dehiscence observed) and finally Mandibular distraction was carried out using two parasymphysial orthodontic stainless steel wires (21 Gauge) under local anaesthesia (For 30 days, resulting in reduction in apnoeic attacks frequency, appreciable mandibular growth and above all, improved quality of life).

Conclusion: Though this technique also has its merits and demerits but still it is promising with a success rate of more than 90% enough to replace tracheostomy which has a high morbidity rate.

Keywords: Pierre Robin Sequence, Micrognathia, Glossoptosis and Cleft Palate, Velocardiofacial Syndrome and Treacher Collins Syndrome.

INTRODUCTION

Pierre Robin Sequence (PRS), named after the French dental surgeon Pierre Robin, identifies a group of neonates affected by congenital retrognathia/micrognathia and glossotossis, which may or may not lead to airway obstruction. A cleft palate may be present but it is not required for diagnosis of PRS.[1]

Robin sequence occurs as an isolated defect, as a part of a recognised syndrome or as a part of a complex, multiple congenital anomalies. The most frequent syndromes involving PRS in decreasing frequency are Stickler syndrome, velocardiofacial syndrome and Treacher Collins syndrome.[2]

Cleft lip and palate is one of the most common congenital maxillofacial developmental defects, with a prevalence rate of 0.28-3.74 per 1,000 live births.[3] Cleft of the lip and palate are more common in American Indians (3.7/1000 live births). Children born with a cleft lip and palate suffer from a number of problems that must be attended to for complete rehabilitation of the patient. Clefts of the palate, alveolus and lip may be syndromic or non-syndromic. The syndromic types are by definition associated with other malformations, and include the Pierre Robin sequence, Treacher Collins Malformation, trisomies 13 and 18, Apert's syndrome, Stickler's syndrome, as well as Waardenburg's syndrome. Nonsyndromic Clefts are of polygenic/multifactorial inheritance.[4]

Infants with craniofacial anomalies (CFA) may experience airway obstruction and feeding difficulties requiring medical and surgical intervention. It has been reported that up to 65% of this population will require some form of airway intervention (Perkins et al., 1997). The range of airway interventions includes both nonsurgical and surgical procedures such as positioning of the infant, nasopharyngeal airway (NPA), endotracheal intubation, tongue lip adhesion, mandible distraction, and tracheotomy (Wagener et al., 2003) each has benefits and morbidities.[5] The ultimate aim of the concerned cleft management team is restoration of different parameters of physiological functions of the body and not to fall as the victims of the disability

CASE REPORT

The present clinical report describes a male infant, born in a government hospital of Guntur, Andhra Pradesh, India. After taking history from the parents of the infant it was found that they had consangious marriage and the child was born at pre- term, weighing 2Kgs 100 Gms (normal range) and

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How to cite this article: Anita Chauhan, Amit Kishor, Rupesh Kumar Singh, Krishnapriya V, Shilpa G, Jay Kishore. A successful effort of corrective intervention in deviated growth by nature leading to survival of an infant by pediatric dental professionals: a case report. International Journal of Contemporary Medical Research 2018;5(9):I26-I29.

DOI: http://dx.doi.org/10.21276/ijcmr.2018.5.9.27

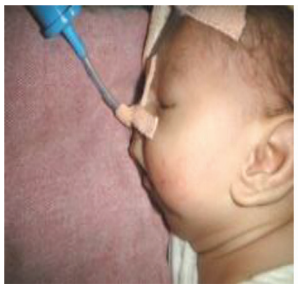
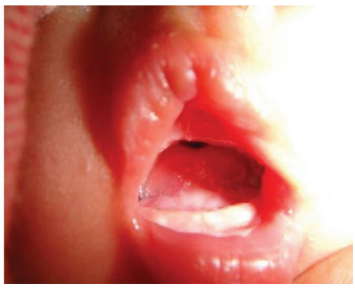

Figure-1: Profile view showing micrognathia; **Figure-2:** Retruded position of the tongue and cleft in the soft palate.

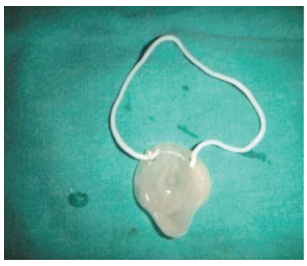
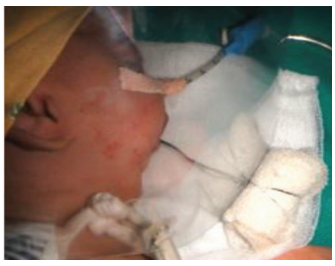

Figure-3: Feeding plate with a supporting thread; **Figure-4:** Modified Glossopexia

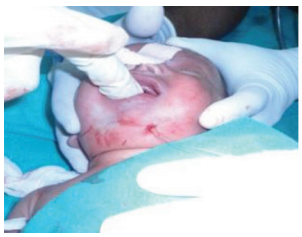


Figure-5: Traction being performed; **Figure-6:** Antiseptic applied on the chin and the wires joined


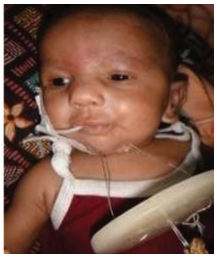

Figure-7: Mandibular traction done and pully being applied; **Figure-8:** Improved growth 15 days after the traction

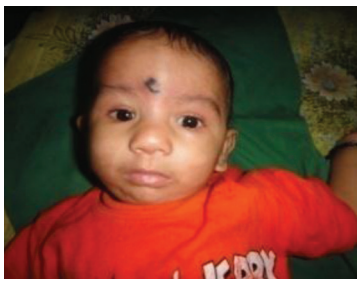

Figure-9: Frontal view after 3 months showing improved growth of the mandible

had no family history of clefting or any other congenital defect. Maternal age at the time of delivery was 17 yrs. and paternal age was 25yrs. After complete general and intra-oral examination the clinician diagnosed the infant with a soft palate defect (Figure 1- 5).

The infant was referred at the age of 35 days to the paediatric ward, Military hospital, Secunderabad, Andhra Pradesh, India as he had frequent spells of breathlessness, turning blue as reported by the parents. The infant also suffered with difficulty in taking nourishment and showed certain skin eruptions.

After complete examination the following details were recorded –Micrognathia, Glossoptosis, Cleft in Soft Palate (Group 1 according to Veau's classification[3]), Respiratory difficulties and Pneumonitis. The child was diagnosed as a case of Isolated PRS.

The child was hospitalised and managed with the non-surgical techniques of airway management. He was put on Antibiotics for Pneumonitis and Nasopharyngeal airway (NPA) and a feeding plate was fabricated.

Initially NPA was used to manage the patient with airway obstruction related to tongue position (Glossoptosis). The main goal of using NPA is to offer a nonsurgical means of airway management that will allow for the infant to be discharged home safely.[5] NPA involves placement of a modified endotracheal tube into one nasal passage, with the tip positioned in the distal oropharynx. It was also planned to construct a feeding plate as an adjunct to help in feeding and also relieving airway obstruction by mechanically pushing the tongue to a normal position.

A written Consent was taken from the parents for making the impression of the defect and fabrication of an appliance. For the impression the infant was positioned facing downwards to prevent airway obstruction and aspiration of the impression material and choking. The impression was made with silicone rubber base impression material with a stock tray that covered the lateral wall of the defect and posterior wall of the pharynx.[6] The internal surface of the completed impression was verified as accurate and sufficient to cover the defect. The impression was boxed and poured in with Type-V dental stone and a cast was poured for the fabrication of a feeding plate. The appliance was fabricated with clear autopolymerising acrylic resin. Two elastics were tied to it in the canine region which was extended outwards to the back of the head for support of the appliance. After checking for the optimal fit, the response of the infant to the new appliance was observed showing improved feeding and breathing patterns. The parents were educated about the appliance insertion and removal, feeding technique and cleaning of the appliance and oral cavity. The child was managed with this feeding plate along with the nasopharyngeal airway 10 days after which the child again suffered from severe apnoeic attacks and there was no appreciable improvement. Though Nasopharyngeal Airway improved his oxygen saturation to some extent, but inspite of the above measures child suffered with Chronic Hypoxemia and failure to thrive.

So surgical intervention was planned with a mutual understanding and discussion with a team comprising of a Pediatriation, Pedodontists, Oral surgeons, Plastic surgeons and Orthodontists and the parents. A technique known as,

tongue lip adhesion (TLA) or Glossopexy, was performed in which the tongue is temporarily attached to the lower lip, bringing the base of the tongue forward.[7] This procedure showed positive results but after 3 days the tongue showed dehiscence which resulted in failure of the procedure.

Finally it was decided to perform mandibular distraction. The traction was decided on the basis of the guidelines and inclusion criteria stated by Schaefer et al[8] in 2004

Mandibular traction was performed under local anaesthesia by using 2 circum-mandibular orthodontic stainless steel wires (gauge 21) on both sides of symphysis.[9] The 2 traction wires were passed through 2 drilled holes in a plate made of rigid plastic and the wires were tied together and total sterilization was maintained. The plate acting as a spacer was used to keep the wires parallel to each other, thus maintaining the direction of traction symmetric, parallel and equal for both the wires. A loop was made by bending the 2 wires, which had been previously joined. This loop served as a point for hanging the weight, through an apparatus of hangers and an orthopaedic pulley attached to the child's bed. The weight applied for orthopaedic traction was approximately 95 grams.

Immediately after applying the wires, a continuous traction of mandible started. The nurses were taught to alternate the position every 2 hrs. The procedure was accomplished successfully with no major complications. The patient's respiratory problems and apnoea disappeared after beginning the traction. The oxygen saturation rates, pulse rate and heart rate were monitored and were found to be improved. The patient was kept in the hospital under supervision for 30 days after which the wirings were removed and the patient was discharged in good health.

DISCUSSION

This case report is a classical example of never giving up hopes especially when it is a matter of life and death. Needless to say that, our dentistry is the mirror of the ever growing medical science. This has been proved in the present case report in which a team of doctors both from medicine and dentistry worked together for complete rehabilitation of the patient. The most important and prime objective in the care of such patients is to provide improved airway with minimum trauma. The oldest established and traditional method of treatment of Upper Airway Obstruction (UAO) in newborns is keeping the infant in downward positioning and nursing in the prone position (Robin, 1934).[10] Cogswell and Easton (1974) measured tidal volume airflow and esophageal pressure in babies with PRS. They found that resistance to airflow was lowest in the prone position with the face straight down.

A little or no improvement was seen in the present case with positioning. Moreover Sjolin (1950) and many others have proved that there is broadening of the hypopharynx and epipharynx in prone versus supine. While prone positioning may be of short-term benefit in both airway maintenance and deglutition, there are no data to support it as a long-term or definitive therapy method. The use of nasopharyngeal Airway (NPA) is suggested by different authors as a useful approach either isolated or in combination with other treatment methods (Schaefer et al., 2003; De Buys Roessingh et al., 2007).[11] One of the benefits of NPA was seen in negative pressure being released in the pharynx during acute obstructive attacks (Fletcher et al., 1969). Stern et al. (1972) suggested that the nasopharyngeal tubes are of greatest benefit during the initial days of life when the infant is most likely to develop severe airway obstruction requiring emergency interventions.

However, although this method is simple, it is difficult to keep the tube in an appropriate position in the throat because of the constant movement of the infant's head.

Coughing or swallowing actions could change the position of the end of the tube. If the tube moves upward, the base of tongue could fall back again causing an uninvited obstruction of the airway. If the tube moves downward, it could irritate the larynx and stimulate the gag reflex. Due to these observations some authors propose that placement of nasopharyngeal tubes should be limited to short periods during a stay in the hospital only.

Since conventional and non-surgical approaches didn't bring much improvement so, surgical intervention was planned.

Tongue lip adhesion was performed with the help of a team of doctors. This was first popularized by Douglas (1946).[12] But because of the high complication rate such as dehiscence's, persistent airway obstruction, tongue lacerations, injury to Wharton's ducts, and scar deformation of the lip and the floor of the mouth, this procedure was modified by different surgeons (Routledge, 1960; Randall, 1977; Argamaso, 1992; Kirschner et al., 2003).[13,14] Finally after no improvements in the oxygen saturation of the infant mandibular distraction was planned out. This method is becoming the treatment of choice for the surgical correction of mandibular hypoplasias. This is less invasive and has significantly decreased morbidity rate compared with the traditional methods.

Schaefer et al. (2003) has suggested that mandibular distraction among neonates is reserved for severe isolated tongue-base airway obstruction cases.[12] The mandibular distraction produces tongue–base advancement in infants. The most important drawbacks of this technique are damage to the tooth buds resulting from injury to the inferior alveolar nerve and the ensuing skin scars externally at the site of traction.[15]

CONCLUSION

The concept of inducing remodelling of mandible and the technique of wire traction were first reported successfully by Callister in 1937. On the basis of this case report and other similar studies which have already been done it can be concluded that the wire traction can be considered as the first choice in cases of severe respiratory distress in newborns affected with isolated PRS.

REFERENCES

1. Shafer's Textbook of Oral Pathology; 6th edition. Chapter 17:717-718.

2. Van Den Elizen AP, Semmekrot BT, Bongers EM, Huygen PL, Marres HA. Diagnosis and treatment of Pierre Robin Sequence: results of a retrospective literature. Eur J Pediatr 2001;160: 47-5.
3. McDonald R, Avery D, Dean J. Dentistry for the Child and the Adolescent. 9th Ed St.: Mosby; 2011:614.
4. Chandna P, Adlakha V.K, Singh N. Feeding obturator appliance for an infant with cleft lip and palate.Journal ofIndian Society of Pedodontics and preventive dentistry.2011;29:71-73.
5. Nooshin Parhizkar, Babette Saltzman, Kellyn Grote, Jacqueline Starr, Michael Cunningham, Jonathan Perkins, Kathleen Sie, Nasopharyngeal Airway for Management of Airway Obstruction in Infants With Micrognathia. Cleft Palate–Craniofacial Journal, July 2011; 48:478-482.
6. Banu Karayazgan, Yumushan Gunay, Bahadir Gurbuzer, Mustafa Erkan, Arzu Atay. A Preoperative Appliance for a Newborn with Cleft Palate Cleft Palate–Craniofacial Journal, 2009; 46:53-57.
7. Janka Kochel, Philipp Meyer-Marcotty, Johannes Wirbelauer, Hartmut Bo¨hm, Michael Kochel, Wolfgang Thomas, Ute Bareis, Helge Hebestreit, Christian Speer, Angelika Stellzig-Eisenhauer. Treatment Modalities of Infants With Upper Airway Obstruction—Review of the Literature and Presentation of Novel Orthopedic Appliances Cleft Palate–Craniofacial Journal, January 2011; 48:44-55.
8. Schaefer RB, Stadler III JA, Gosain AK. To distract or not to distract:an algorithm for airway management in isolated Pierre Robin Sequence. Plast Reconstr Surg. 2004; 113: 1113-1125.
9. U. Baciliero, S. Spanio di Spilimbergo, M. Riga, E. Padula. Respiratory distress in Pierre Robin Sequence:an experience with mandible traction by wires.Int. J. Oral Maxillofac. Surg. 2011;40:464-470.
10. Robin P. Glossoptosis due to atresia and hypotrophy of the mandible. Am J Dis Child 1934; 48:541-547.
11. Schaefer RB, Gosain AK. Airway management in patients with isolated Pierre Robin sequence during the first year of life J Craniofac Surg. 2003;14:462-7.
12. Douglas, B. The treatment of micrognathia associated with obstruction by a plastic procedure. Plast Reconstr Surg 1946; 1: 300-8.
13. Faye Huang, MD; Lun-Jou Lo. Tongue-Lip Adhesion in the Management of Pierre RobinSequence with Airway Obstruction: Technique and Outcome. Chang Gung Med J 2005; 28: 90-96.
14. Kirschner RE, Low DW, Randall P, Barlett SP, McDonald-McGinn DM, Schultz PJ, Zackai EH, LaRossa D. Surgical airway management in Pierre Robin Sequence: is there a role for tongue – lip adhesion? Cleft Palate Cran J. 2003;40: 13-18.
15. McCarthy JG, Schreiber J, Karp N. Lengthening of the human mandible by gradual distraction. Plast Reconstr Surg 1992; 89:1-8.

Source of Support: Nil; **Conflict of Interest:** None

Submitted: 07-08-2018; **Accepted:** 09-09-2018; **Published:** 20-09-2018