Evaluation of treatment on unilateral cleft lip and palate complete dextra to the quality of life: 4-year follow-up

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ABSTRACT
Treatment for the patient with cleft lip and palate (CLP) requires an interdisciplinary team concerned with improving the patient's quality of life. This case evaluates the influence of comprehensive therapy on unilateral CLP complete dextra within a 4-year follow-up. A 4-days-old female patient visited a government hospital in Bandung after being referred to a pediatric clinic in the same hospital. The patient was born at 3.2 kg weight with no underlying medical conditions or syndromes. Intraoral examination revealed cleft soft and hard palate and separated alveolar segment. Extraoral examination revealed cleft lip unilateral dextra and shortened columella. The patient underwent comprehensive therapy including presurgical NAM (PNAM), labioplasty, nasoalveolar molding (NAM), veloplasty, speech therapy, and palatoplasty. It is concluded that early treatment of PNAM reduces the severity of the orofacial deformity, induced the reposition of soft & hard tissue and there's an improvement in intelligibility and production of pharyngeal sounds and velopharyngeal closure when using intravelar veloplasty in palatal repair along with NAM. In the fourth year of life, the patient has capable on communicates with intelligibility pronounce with relative adequate arch.
Keywords: unilateral cleft lip and palate complete dextra, nasoalveolar molding, intravelar veloplasty, comprehensive treatment

INTRODUCTION

Cleft lip-and-palate (CLP) is considered the most common congenital craniofacial birth anomalies in different populations. According to the literature based it has a multifactorial etiology, comprising both genetic and environmental factors. And the relative risk of CLP including environmental influence (smoking, alcohol use, drugs, and dietary factors) will be useful for the development of future preventive measures.[1,3,10] The presence of CLP introduces feeding difficulties, concerns regarding speech development, and the possibility of impaired facial growth. Children with cleft anomalies may experience a multitude of physical and developmental challenges. There also may be psychosocial and emotional concerns for the patients and their families. As such, comprehensive care for the patient with cleft lip and or palate requires an interdisciplinary team.[1,3] The guidelines for team care outlined by the Government Hospital in West Java Indonesia recommend team members that may include oral maxillofacial surgery, prosthodontics, orthodontics, pediatrics, pediatric dentistry, otolaryngology-head and neck surgery, plastic surgery, anesthesiology, audiology, genetics, neurosurgery, ophthalmology, psychology, psychiatry, and speech-language pathology. Management of patients suffering from CLP can start early at infancy with treatment goals of lip segments approximation, nostrils symmetry achievement, increase columellar length, and alveolar segments alignment. In other words, the aim at infancy is to help the surgeons to achieve better surgical result by decreasing the severity of the cleft defect.[10]

The early treatment of CLP is nasoalveolar molding (NAM) therapy, that may be successfully employed in the early management of both unilateral and bilateral cleft anomalies in newborns.[8] Studies have shown that when instituted at 1 week of age and continued 3-4 months, NAM is effective in approximating the cleft as well as improving the nasal deformity. Patients undergoing NAM treatment experienced improved nasal alar symmetry, columella lengthening, and nasal tip projection.[1] Performing intravelar veloplasty (IVV) along with NAM in primarily repaired palates was believed to gain the functional repair of the palate. Early repair of cleft palate yields the best results in regard to adequate velar port closure, less hypernasality of speech, and better development of speech articulation. The velum is known to have a great role in producing intraoral pressure and thereby producing certain sounds by its movement against and down the pharyngeal musculature and maintenance of nasopharyngeal closure during feeding. Adequate velar port closure is essential for the balance of oronasal resonance during speech production.[4] Previous anatomic studies of the palatal musculature concluded IVV would decrease the incidence of post palatoplasty velopharyngeal insufficiency (VPI).[5] So,

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IVV significantly improves velopharyngeal port closure and speech production later in life.[3]

In this case patient experienced a complete unilateral CLP. The objective of cleft lip repair is to approximate the medial and lateral lip elements with preservation of natural landmarks, align a functional concentric orbicularis, and to establish symmetry and proportionality.[1] From the beginning, the patient's planned for undergone 2 stage closure with soft palate closure at 12 months of age and hard palate closure will be planning at approximately 5 years old. The 2 stage palate repair with delayed hard palate closure was chosen for the patient to mitigate the risk of growth interference. By performing a veloplasty first, the hard palate is encouraged to narrow as it facilitates normal midfacial growth.[1]

CASE

A 4-day-old female patient visited a government hospital in Bandung after being referred to a pediadiatric clinic in the same hospital. The patient is the firstborn with unremarkable family history. The baby was born after a full-term pregnancy at 3.2 kg weight with no underlying medical conditions or syndromes. Intraoral examination revealed cleft soft and hard palate and separated alveolar segment. There is an external and upward rotation of the medial segment of the premaxilla and an internal and posterior rotation of the lateral segment. Extraoral examination revealed cleft lip unilateral dextra and shortened columella. The patient underwent comprehensive therapy including NAM stage 1, labioplasty, NAM stage 2, IVV, speech therapy, and palatoplasty (Fig.1).

Figure 1 The 4-day-old patient

MANAGEMENT

A comprehensive treatment was initially started with NAM therapy. The first phase was to take an impression prior to fabricating the plate under pediatric supervision. The impression is obtained with the infant fully awake and without any anesthesia. The infant was held to prevent the possible aspiration of regurgitated stomach contents and also helped to lay in the upright position during taking an impression to avoid any aspiration of the materials.

Care is taken to ensure that the material has registered the border regions of the maxilla and premaxilla as well as the cleft region. The infant should be able to cry during the impression-making (Fig.2) procedure. If no crying is heard, the airway is blocked. At our institution, all impressions of clefts in infants are made in the hospital setting with a pediatrician present as part of the impression team. The hospital setting also allows a rapid response by an airway team should there be an airway emergency.

Figure 2 An elastomer impression

Figure 3 NAM appliance

The NAM appliance was fabricated using heat-cured polymethyl methacrylate resin. Any part that makes contact with the defect area is the anatomic part and the other side is the mechanical part. After initially inserting the oral molding appliance, the baby must be observed for several minutes while the clinician stabilizes the appliances. The infant must be able to easily suckle without gagging or struggling. Parental caring for the treatment is immensely fundamental. The parents were instructed on the use of the plate for 24 hours despite cleaning the appliances in between. They were also counseled regarding potential complications and were shown pictures to enable them to spot rashes early. They were also shown how to clean the mouth and peri-oral area post-feeding (Fig.3).

To achieve the desired movement, the acrylic is selective removed from the region into which one desires the alveolar bone to move. These minor adjustments are made weekly. The ultimate goal of the addition of materials and selective grinding is to reduce the size of the cleft gap and to have the two segments of alveolus contact with the configuration of a proper maxillary alveolar arch form. On the second visit, an extraoral retentive button is built at the site of the cleft in the lip. This retentive

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button serves to facilitate both the positive seating of the appliance to the palatal tissue and to secure the retentive lip tapes and elastic bands. Proper lip taping between appointments is crucial if the appliance is to be maximally effective. The tape will serve to help retain the appliance and allows the soft tissues of the lip and nasal base regions to become more properly oriented as the nasal stent is developed. A lip tapping force in conjunction with a molding plate yields a controlled movement of the alveolar segments and also serves to improve the alignment of the nasal base region by bringing the columella toward the midsagittal plane (Fig.4). Lip tapping also improves the symmetry of the nostril apertures.[7]

Figure 4 Patient underwent lip tapping

In this report, a nasal stent was incorporated into the molding plate when the cleft gap has been reduced to approximately 6 mm or less (Fig.5).

Figure 5 Alveolar gap was reduced to approximately 4.5 mm

Figure 6 An incorporated nasal stent to the molding plate

The stent serves as a custom tissue expander that slowly corrects the flattening of the cleft lip nasal deformity. It also serves to bring the columella into a more midline position when the lip has been taped with the nasal stent in place. Nasal stent positioned in nostril aperture to support the dome and reposition lower nasal cartilage. The nasal stent exerts a reciprocal intraoral molding force against the alveolar segments (Fig.6).[7]

Figure 7 1-month post labioplasty

The patient underwent labioplasty on 15 weeks of age after regaining the exact improvement of the soft and hard tissue reposition. Sufficient weight according to triple ten is mandatory. Following evaluation, an impression of the intraoral cleft defect was made using an elastomeric material in an acrylic custom tray for stage two NAM appliance. The infant underwent speech therapy provided by a rehabilitation medic at 11 months of age (Fig.7).

Figure 8 A stage two of NAM appliances

According to Taylor, the palate repair is performed at our institution when the infant shows evidence of phoneme speech development at approximately 11-13 months of age. Several palate-repairs surgery at our institution were held initially with IVV. IVV is the procedure of reconstruction of the levator muscle sling in cleft palate patient in order to gain velopharyngeal (VP) closure. It is achieved by detaching the levator muscles from their abnormal attachment to the hard palate and repairing them in the midline with the muscle fibers oriented more normally. The main objective of IVV surgery was to avoid the undesirable VP closure insufficiency. The term VP insufficiency refers to a structural deficit in VP spinchter. The patient underwent IVV surgery on 13 months of age (Fig.8).

As along with speech therapy the patient regularly visits the department of prosthodontics for

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Figure 9 Four months post IVV

observation (Fig.9). Selective grinding was done to the NAM in order to reduce the size of the palate defect (Fig.10). As the years goes by, the NAM has been regularly rebuilt and also reduced. The patient was well adapted to the NAM and highly capable to chew and also swallow (Fig.11). The patient spoke in good articulation and good word spelling.

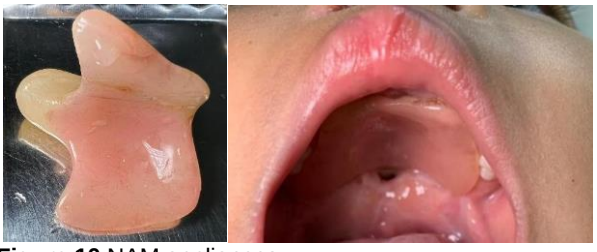

Figure 10 NAM appliances

Figure 11 A 3 years old patient

Figure 12 A 4 years old

In the fourth year of the patient's life, she's capable on communicates with intelligibility pronounce with relative adequate arch (Fig.12). The patient was managed gradually to discharge of appliance by learning to talk, sing, drink and swallow without NAM. Until today the patient is under monitoring still by the rehabilitation medic therapist to improve the pronunciation structures. At five years of age, the patient will have been planning to undergo palatoplasty surgery.

DISCUSSION

The unilateral cleft lip and palate defect will yield a less than ideal esthetic result when addressed only through a surgical correction. The basic goal of any approach to cleft lip, alveolus, and palate repair is to restore normal anatomy. Ideally, deficient tissues should be expanded and malpositioned structures should be repositioned prior to surgical correction. This provides the foundation for a less invasive surgical repair.[7] NAM serves two purposes. In addition to closing the hard tissue gaps to enable the surgeon to better close the gap, it was also hoped that the plate would help her feed and gain weight to reach the proper and safe weight to perform surgery under general anesthesia. NAM includes not only the reduction of the size of the intraoral alveolar cleft through molding the bony segments, but also the active molding and positioning of the surrounding soft tissues affected by the cleft, including the deformed soft tissues and cartilage in the cleft nose. This is accomplished through the use of a nasal stent that is based on the labial flange of a conventional oral molding plate and enters the nasal aperture. The stent provides support and gives shape to the nasal dome and alar cartilages.[7,8] The infant should be carefully monitored on a weekly basis to check the progress of the movement of the ridges and avoid a situation where there is *locking out* of the lesser segment by the posteriorly directed greater segment. When the cleft gap has been reduced to approximately 6 mm or less, a nasal stent may be added to the molding plate. The proper sequence of molding (alveolar followed by nasal) is followed to avoid the production of a *mega-nostril*. According to Latham, the first surgical procedure is usually performed between 12 and 16 weeks of age to address the defects of the entire nasolabial complex. The palate repair, if indicated, is usually performed once the infant shows evidence of phoneme speech development and occurs at approximately 11-13 months of age.[7]

The jaw and palate develop along normal lines if the soft palate is closed during infancy by means of primary veloplasty. The residual cleft in the hard palate remains. The cleft becomes narrower with

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growth of the palate without causing compression of the jaw.[9] The success of palatal repair is directly related to the adequacy of velar muscle repair. Adequate VP closure is essential for the balance of oronasal resonance during speech production. The complete VP closure occurs from the simultaneous movement of the soft palate and the lateral and posterior pharyngeal walls, which ensures the complete separation between the oral and nasal cavities during the production of oral speech sounds.[3,4] Previous anatomic studies of the palatal musculature concluded IVV would decrease the incidence of post palatoplasty VPI. VPI is highly undesirable and would lead to the appearance of characteristic symptoms such as hypernasality because there is a communication between the oral and nasal cavities, so part of the air current diverges to the nasal cavity during the production of oral speech sound.[4-6] As far as speech development is concerned, the intelligence and the temperament of the child play an important part as do family attitudes. Communicative children in a socially favorable environment will find it easier. In any case, most children are able to enter normal schools. Follow-up investigations into the comprehensibility of speech have shown that speech improved after school entry as it is true for most children. In terms of enabling the jaw and face to develop normally and facilitating the development of normal speech, the primary veloplasty is really justified and recommended for the rehabilitation of patients suffering from CLP.[9]

It is concluded that in the case of the unilateral cleft, the infant should be seen on a weekly basis for continuous follow-up prior to surgery. The desirable achievement therapy is highly determined by supportive parents to accomplish all the sequence therapy. In all cases of clefts, the final assessment of clinical treatment is possible only after the patients have reached the adolescence.

REFERENCES

1. Shaye D, Liu CC, Tollefson TT. Cleft lip and palate an evidence-based review. Fac Plast Surg Clin N Am 2015:357-72
2. Muhamad A, Azzaldeen A, Watted N. Cleft lip and palate; a comprehensive review. Int J Basic App Med Sci 2014;4(1):338-55
3. Malki HK, Mustafa SO. The role of intravelar veloplasty in primary cleft palate repair. Zanco J Med Sci 2018;22(1):56-64.
4. Barbosa DA, Scarmagnani RH, Fukushiro AP, Trindade IE, Yamashita RP. Surgical outcome of pharyngeal flap surgery and intravelar veloplasty on the velopharyngeal function. CoDAS 2013;25(5):451-5
5. Marsh JL,Grames LM, Holtman B. Intravelar veloplasty: a prospective study. Cleft Palate J 1989;1(26):46-9
6. Marsh JL Velo-pharyngeal dysfunction: evaluation and management. Indian J Plast Surg 2009;42(suppl):129-36
7. Taylor TD. Clinical maxillofacial prosthetics. Illinois: Quintessence publishing.; 2000.p.63-78
8. Almotawah FN. The impact of presurgical nasoalveolar molding on the surgical and quality of life outcome: A case report with 1-year follow-up. Saudi J Oral Sci 2020;7(3):207-9
9. Schwekendiek W. Primary veloplasty: long-term results without maxillary deformity.A twenty-five year report, 3rd int'l Congress On Cleft Palate. Toronto; 1977.
10. El-Ghafour MA, Elkordy SA. Parents' acceptance to alveolar and nasoalveolar molding appliances during early cleft lip and palate care: a call for high-quality research. Maced J Med Sci 2020;8(F):58-64

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