Surgical Correction of Cleft Lip Accompanied with Macrostomia: Case Report

Nyoman Ayu Anggayanti¹, Harmaz Yazid Yusuf², Endang Sjamsudin³, Agus Nurwiadhi⁴

¹Resident of Oral and Maxillofacial Surgery Department Faculty of Dentistry University Padjadjaran, Dental Hospital University Padjadjaran, Bandung, Indonesia; Staff of Oral and Maxillofacial Surgery Department Faculty of Dentistry, University Udayana, Denpasar, Indonesia

², ³, ⁴Staff of Oral and Maxillofacial Surgery Department Faculty of Dentistry University Padjadjaran, Indonesian Cleft Centre and Dental Hospital University Padjadjaran, Bandung, Indonesia

Abstract: Introduction: Cleft lip accompanied with macrostomia is a rare congenital disorder. This condition stems from the fusion failure of upper and lower jaw at the first branchial arch and could be syndromic or isolated, unilateral or bilateral. Purpose: To report the management of cleft lip accompanied with macrostomia with good results functionally and esthetically. Case report: A 3-years-old baby boy with cleft at upper lip, right corner of the lip and palate since birth. Case management: Surgical correction was done with Millard technique to correct the cleft lip and double Z-plasty to correct the macrostomia. Conclusion: Surgical correction of cleft lip with macrostomia in infant has to be done simultaneously to obtain satisfying results in both functional and aesthetic aspects.

Keywords: Cleft lip, Macrostomia, Surgical

1. Introduction

Cleft lip accompanied with macrostomia is a rare congenital disorder, known as Tessier cleft type 7, and also one of the rarest facial anomalies. Although the reported incidence of cleft lip is 1 in 3000-5000, the incidence of lateral cleft lip is 1 in 100,000-300,000 and constitutes about 0.3-1% of all facial clefts.¹

The etiology of lateral cleft is the failure of fusion of the maxillary and mandibular processes of the first and second branchial arch or due to disruption in the processes after fusion. It is commonly associated with macrostomia and defects of the first branchial arch. The patients commonly have other deformities such as preauricular tags, sinuses and Goldenhar syndrome bilateral lateral clefts. This cleft can present as slight widening of the mouth to a cleft extending up to the ear. It can be bilateral, but most of the reported cases are unilateral and do not extend beyond the anterior border of the masseter.¹, ²

2. Case Report

A 3 years old baby boy patient came to polyclinic Oral and Maxillofacial Surgery Department Padjadjaran Dental Hospital Bandung with his parents in July 2019, with cleft at upper lip, at right corner of the lip and palate since birth. The main complaints of parents was unacceptable facial appearance as well as disturbed oral functions. There was no history of complication during pregnancy, there was no history of any medications taken and exposure to radiation during pregnancy. Delivery process was natural.

General physical examination found the body weight was within normal limit (around 12 kg). Extra oral examination found an asymmetrical face, there was cleft at right side of upper lip and right corner of the lip. Microtia was seen on right ear. Intra oral examination revealed a complete cleft palate. The patient was diagnosed with complete cleft lip and palate accompanied with macrostomia at right corner of lip.

Figure 1. Preoperative photograph with cleft at right upper lip, right corner of the lip and palate.

3. Case Management

The patient was planned to be treated with surgical correction of cleft at upper lip and right corner of lip under general anaesthesia. Blood count and chest x-ray was performed before the surgery. The patient was consulted to Pediatric Departement for cardiopulmonary examination and to Anaesthesia Departement for preoperative and perioperative preparation. After all examination was completed, it was concluded that no contraindication for the treatment and the informed consent was carried out.

The surgery was perfromed in the middle of July 2019. Surgical correction of cleft lip was done with Millard technique and double Z-plasty was done to correct the macrostomia. The patient was on supine position under general anaesthesia. After the aseptic and antiseptic was done, surgical marking for both cleft was made identically. After the aseptic and antiseptic was done, surgical marking for both cleft was made identically. After the aseptic and antiseptic was done, surgical marking for both cleft was made identically. After the aseptic and antiseptic was done, surgical marking for both cleft was made identically. After the aseptic and antiseptic was done, surgical marking for both cleft was made identically.

The incisions and dissection were made in...
accordance to previous diagramming in the usual manner. The surgical correction was done in three layer closure steps: first, the mucosal, muscle, and skin. Surgical correction of cleft lip with macrostomia in infant has to be done simultaneously in order to obtain satisfying results in both functional and aesthetic aspects.

![Figure 2](image_url)

**Figure 2.** Durante operation (A). Marking side, (B). After Surgery

Post operative therapy was managed by injection of amoxycillin 100 mg intravenously and suppositoria analgetic. At the first day after surgery the patient’s lip showed a minimum edema, no bleeding on the site of surgery and there was no complaint of fever due to dehydration state. We instructed the parents to give milk or water using spoon for a month. The patient’s state was observed for two days after surgery. Bandage was replaced every day until day seventh. The suture removal was performed on day seventh. Clinically seen, there was no wound dehiscence nor infection at the site of surgery. Two weeks after surgery the cleft on the right side of upper lip and right corner of lip has closed and the healing was good.

![Figure 3](image_url)

**Figure 3:** (A). First day after surgery, (B). Seventh day after surgery the suture was removed, (C). Two weeks after surgery

4. Discussion

Transverse clefts or the lateral cleft lip is more common in the male child and, although most of the reported cases are unilateral, there are quite a few bilateral occurrences as well. The additional anomalies reported with the lateral clefts are hemifacial microsomia, preauricular skin tags, microtia, absence of the Eustachian tube, temporomandibular joint, zygomatic arch, polydactyly and renal anomalies. The database of the cleft lip and palate clinic was analysed for the incidence of lateral facial clefts, as well as its various associated presentations: cleft lip unilateral left-sided, unilateral right-sided cleft or bilateral as well as the superiorly rotated, middle-positioned, inferiorly rotated and agenetic lateral cleft lip.1,3

The etiopathogenesis of macrostomia remains unknown, many theories have been proposed. The mandibular dysplasia and transverse soft tissue deficiency could be explained by a lack of growth of the mandibular process. Others have suggested a vascular etiology, explained by the presence of a hematoma in the territory of the stapedia artery preventing fusion of the maxillary and mandibular processes.4

Problems associated of cleft lip accompanied with macrostomia include aesthetic disharmony and functional problems like feeding difficulties, drooling, speech incoherence, and difficulty in blowing. Surgery should be done in young age to avoid unwanted anxiety and psychological impacts on both child and family as well as correcting sialorrhea, speech problems and compromised chewing ability.1,4

Various surgical techniques have been evolved over a period of time with revisions to the existing ones to attain harmony between function and aesthetics.5 Two basic techniques are universally in use for unilateral cleft lip closure: the Tennison-Randall procedure and the Millard procedure. Both techniques recognize the importance of repositioning the lip muscle orbicularis oris in a correct anatomic orientation that results in an aesthetic as well as a functional improvement. The Millard procedure is known as the rotation-advancement technique. It is a more flexible technique cut-as-you-go but needs more experience and artistry. The technique camouflage the violation of the philtrum column near the nose. With the Millard technique, one easily gets a vertical scar contracture with vermilion notching of the lip and a notorious tendency towards a small...
nostril. Excessive narrowing of the nostril is never far from reality and the surgeon simply should aim for a slightly larger nostril on the cleft side.

Surgical technique for the correction cleft lip and macrostomia should address skin, muscle, and mucosa. There should be natural blending of the mucosa with the skin at the commissure. For commissure, triangular mucosal flaps or triangular skin flaps are used. For skin closure, straight line or Z- or W-plasty is used. Straight line muscle closure or overlapping myoplasties are used for muscle reconstruction. In a lateral cleft, the skin, muscles and mucosa may be reconstructed in such a way as to create a scar resulting in a good facial appearance. This can only be done, if a modified cutaneous Z plasty, rotated superiorly, with modiolus reconstruction is performed.

Complications observed with surgical techniques include asymmetric closure, hypertrophic scar, drooping of oral commissure, and fish mouth deformity resulting from flaccid commissure. One should consider the symmetry both in vertical and in horizontal plane as improper techniques might result in asymmetry. Dehiscence is not very common in unilateral cleft lip repair. The main reason is either tension in the repair or improper suturing of the orbicularis muscles. Trauma or infection also may be possible causes.

5. Conclusion

Cleft lip accompanied with macrostomia is a rare case that best treated with surgical correction. The combination of Millard and double Z-palst sy technique was an option for this condition, and it has to be done simultaneously to obtain satisfying results in both functional and aesthetic aspects.

References