

Treatment of Bilateral Macrostomia (Lateral Lip Cleft): Case Report

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Received: Apr 16, 2011; Final Revision: Oct 26, 2011; Accepted: Jan 06, 2012

Abstract

Background: Macrostomia as a rare facial deformity is classified among facial clefts. It originates from failure in union of maxillary and mandibular prominences of first brachial arch during 7th embryonic week.

Case Presentation: We report a case of bilateral macrostomia (bilateral lip cleft) in a female newborn as a sole entity without other skeletal and facial deformities. The cleft was repaired by a simple linear triangular flap using extra oral landmarks to locate lip commissures. Patient was followed through a six-month period. Acceptable results were gained in mouth appearance as well functional aspects.

Conclusion: Commissural repair through a linear flap can result in minimal visible scar with satisfying results in both esthetics and functional aspects.

Iranian Journal of Pediatrics, Volume 22 (Number 3), September 2012, Pages: 425-427

Key Words: Cleft Lip; Macrostomia; Maxillofacial Abnormalities; Congenital Structural Deformity

Introduction

Macrostomia or lateral lip cleft, which is known as Tessier cleft type 7, is one of the rarest facial anomalies^[1]. Its occurrence is reported 1 in 225,000^[2] or 0.02%^[3] of all live births and constitutes about 0.3% to 1% of all facial clefts³. The anomaly may be found as a part of syndromes like Oto-mandibular dysostosis, Hemifacial microsomia, Treacher-Collins, Goldenhar, or in conjunction with additional facial symptoms such as preauricular tags, zygomatic arch deficiency, deformities in different mandibular parts such as ramus, condyle, or coronoid process, or ear anomalies^[4] (deformities in external, middle or inner parts), and other facial clefts such as Tessier

[2-5]. However, it is uncommon to find macrostomia as an isolated asyndromatic entity.

The embryonic origin of lateral lip cleft may be related to the 7th embryonic week, when the maxillary and mandibular processes of first branchial arch merge laterally to fuse in a posterior to anterior manner and construct the corners of mouth and cheeks. Any disruption in this process may lead to lateral clefts^[5]. It is also claimed to be a post-merging anomaly due to considerable clinical variability in expression^[6].

The manifestations of lateral facial cleft may vary from a slight commissural involvement and macrostomia, that sometimes remains undiagnosed specially when combines with other facial anomalies to a gross combination of soft and

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hard tissue extended from mouth to tragus [6].

Case Presentation

A 4-month female infant was referred to Mofid Pediatric training Hospital for correction of inferiorly rotated bilateral transverse lip cleft (Fig. 1). The main complaints of parents were unacceptable facial appearance as well as disturbed oral functions specially sucking and breastfeeding abilities, and nose vomiting. There was a history of uncomplicated pregnancy and natural delivery in a G1P1 mother. Mother had not received any medications and was not exposed to radiation during pregnancy. Parents were relatives, with a history of cleft presented in the family. The baby was a full term, normocephal female, with normal fontanels and auditory meatus without any evidence of skin abnormalities. The treatment decision made based on the clinical findings relying on the fact that bilateral lip cleft is a soft tissue entity without involving the bony components. The position of new commissures was determined by the point that vermilion texture changes from normal mucosa to cleft mucosa [7]. Correctional surgery performed on lip with simple linear triangular incision and full thickness flap in lower one third of the lip. After releasing the muscle from skin and mucosa, the cleft area was repaired in three layers of muscular, mucousal and skin tissue mucosal layer was sutured to muscle by interrupted sutures. Finally the skin layer was repaired by simple linear inferiorly rotated triangular incision.

The facial appearance immediately after surgery (Fig. 2) and seven days later are presented in Fig. 3.

Discussion

Transverse lateral facial clefts are usually found unilaterally most often in males. Bilateral inferiorly rotated cleft is a rare occurrence [4]. Skin and facial deformities are usually absent in

bilateral types, that is in concordance with our findings [3,11].



Fig. 1: The cleft appearance before surgery



Fig. 2: The facial appearance immediately after surgery



Fig 3. Facial appearance one week after surgery

The cause of anomaly may not be defined with certainty. The mother reported an uneventful pregnancy and labor without any exposure to medications or radiation; however, the parents were relatives and a familial history of other types of clefts was present in this case that intensifies the genetics as the most probable etiological factor. A lateral cleft is considered as a multifactorial congenital anomaly (interactions of several genes and environmental factors), or postzygotic mutation [5]. Various mutations may change in the neural crest or surface proteins which may lead to lateral facial clefts [2]. It may also be attributed to disruption in stapedial artery blood flow during the rapid embryonic growth and development.

Esthetics as well as function, especially in subjects without other anomalies, is a great concern. 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. The aim of surgery is to place symmetric lip commissures with normal contour and minimally visible scar. Reconstruction of orbicularis oris is the key of normal appearance of lip commissure [7]. In this case cleft was an inferiorly rotated type. In contrast to superiorly or middle rotated types, inferiorly rotated lateral lip cleft does not involve the bony component, however separation of the muscles such as the risorius and depressor anguli oris can be observed. In repair process reconstruction of commissural part needs precise technique. The natural corners of upper and lower lip form small triangular flap [10]. Initially Z plasty were commonly performed, however it has the shortcomings such as of suboptimal esthetics [7]. Other techniques such as simple linear flap, W-plasty, and triangular flap which was the one used in this patient have shown good results. In point of scar contraction in linear incision, it should be noted that precise repair of orbicularis oris muscle is the key point in transverse lip clefts which restores muscular balance and prevents inferior displacement of the commissures. A well repaired orbicularis oris would provide a medial dynamic counterforce to lateral displacement resulting from contractile force of the linear scar [8].

Conclusion

Bilateral macrostomia as a rare soft tissue deformity can be repaired successfully with minimally visible scar by simple linear triangular flap. It seems that Z-plasty or W-plasty techniques are unnecessary in repairing transverse cleft lips

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