Surgical repair of macrostomia - cleft 7 - a case report

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ABSTRACT
Macrostomia, also called Tessier 7 or lateral cleft is an uncommon congenital anomaly. It results from the failure of fusion of the maxillary and mandibular processes of the first branchial arch and could be syndromic or isolated, unilateral or bilateral. This paper reports a case of bilateral macrostomia (bilateral lip cleft) in a seven month old baby as a sole entity without any other skeletal and facial deformities. The cleft was repaired by a simple linear flap using extra oral landmarks to locate lip commissures. Patient was followed through a six-month period.

Keywords: Bilateral Cleft Lip; Lateral Cleft; Macrostomia; Tessier 7

Introduction
Congenital macrostomia, a form of transverse facial cleft. This type of anomaly accounts for one in 300 cases of facial fissure. So far, only 21 cases of bilateral macrostomia have been reported by the literatures. The embryonic origin of lateral lip cleft may be related to the seventh 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. It is also claimed to be a post-merging anomaly due to considerable clinical variability in expression. Numerous surgical reconstructive techniques have been developed such as, the straight-line closure, without supero-inferior rotation or positioning of the scar or W-plasty to normalise the appearance of this unsightly defect. Some authors prefer a type of z-plasty for the closure, which may be positioned horizontally or inferiorly. The paper reports the management of a seven month old male child with Tessier no 7 cleft.

Case Report
A seven month old baby was referred to SKIMS Hospital for correction of inferiorly rotated bilateral transverse cleft lip (Figure 1). The main complaints of parents were unacceptable facial appearance as well as disturbed oral functions. There was a history of uncomplicated pregnancy and natural delivery, and no history of any medications and exposure to radiation during pregnancy. The baby was a full term, normocephalic male, with normal fontanels and auditory meatus without any evidence of skin abnormailties. The treatment decision was 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. 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 mucosa and muscle to muscle by interrupted sutures. Finally the skin layer was repaired by simple linear inferiorly rotated triangular incision (Figure 2, 3).

Discussion
Tessier no 7 clefts are more rarely encountered than cleft lip and palate deformities. Physical restricting forces such as amniotic bands may lead to the formation of these unusual clefts. This hypothesis is supported by an experimental study by Stelnicki et al. There are several classification systems, such as that of the American Association of Cleft Palate Rehabilitation, Karfik, Boo-Chia, Demeyer, and Van der Meulen. However, the best known classification system of facial clefts is that devised by Paul Tessier. The clefts are numbered 0 to 14 with cleft number 8 forming the equator. Hence, clefts numbered 0 to 7 of the lower hemisphere represent the facial clefts and the clefts of the upper hemisphere numbered 9 to 14 are their cranial prolongation. According to Tessier, cleft number 7 is a lateral cleft. The side of involvement is not significant. The incidence of Tessier number 7 cleft has been determined to be 1/3000 to 1/5642. The clinical expression of number 7 cleft is highly variable. In addition to the facial findings, hypoplasia of the zygoma, temporal bone, maxilla and mandible, parotid gland and parotid duct, and hypoplasia of the innervation area of the fifth and seventh cranial nerves, palate and tongue have been seen.

The severity of the deformity, the pathophysiology of the disorder with regard to growth potential and the psychological aspect of the deformity all contribute to the decision regarding the surgical intervention. Treatment involves the reconstruction of soft tissue followed by reconstruction of the bony frame. Soft tissue reconstruction should be performed in the preschool period. In this period, excision of skin tags, and the correction of macrostomia and prominent ear are appropriate. When unnecessary, the bone frame is not reconstructed in the early ages. Bone grafting and augmentation must be applied at older ages. At five years of age, cranio-orbito-zygomatic development is at 85% of adults. For this reason, skin and bone reconstruction of the middle face have to be done.
after age six, and that of maxilla and mandibular interventions must be performed in the adolescent period. 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 salivary 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. 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. Initially Z plasty were commonly performed, however it has the shortcomings such as of suboptimal esthetics, Other techniques such as, Wplasty, triangular flap and simple linear 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 medially dynamic counterforce to lateral displacement resulting from contractile force of the linear scar.

Conclusion
In conclusion, bilateral macrostomia as a rare soft tissue deformity can be repaired successfully with minimally visible scar by simple linear triangular flap.

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References

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