

Interesting Case Series

Bilateral Macrostomia

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DESCRIPTION

A 16-month-old boy with bilateral macrostomia, cleft palate, bilateral cleft lip repair, and developmental delay presents to a medical mission surgical center in Valladolid, Yucatan, Mexico. The picture demonstrates inadequate closure of the mouth bilaterally.

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QUESTIONS

- 1. What is the epidemiology of this condition?**
- 2. What other symptoms may be present in a patient presenting with macrosomia?**
- 3. What surgical techniques are used for reconstruction of this defect?**

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DISCUSSION

Macrostomia is most often associated with a transverse facial cleft, classified by Tessier as a number 7 cleft. It is found in 1 out of 80,000 births and occurs more frequently in males. Most cases are unilateral, but bilateral involvement can be seen in 10% to 20% of cases. More than 50% of bilateral cases are isolated without additional ear or skin deformities. However, the defect is often larger than in unilateral macrostomia.

The severity of the cleft can vary, and thus, additional symptoms may or may not be present. Examples include preauricular tags; microtia; zygomatic arch deficiency; deformities of the mandibular ramus, condyle, and coronoid process; and other craniofacial clefts. The cleft can also be part of an underlying syndrome and present with an absent orbital rim and eyelash hypoplasia in Treacher Collins syndrome, facial asymmetry in hemifacial microsomia, and vertebral anomalies in Goldenhar syndrome.

Surgical repair is more complicated in bilateral macrostomia than unilateral, because the surgeon lacks a comparative normal side and must define the location of the commissure without oral anatomical aide. Fortunately, the vermilion often thins at the approximate location of the natural commissure and can align with a line from the pupils to the infraorbital and submental foramina.

Three layers must be considered for repair: mucosal, muscular, and skin layers. The mucosal layer is closed with simple interrupted absorbable sutures, with the commissural sutures attaching the mucosal layer to either the overlying muscle or skin. Function for speaking, eating, and blowing is dependent upon correction of the muscular layer. Involved muscles consist of the orbicularis, buccinator, and zygomatic muscles. The superior orbicularis muscle is sutured at a 90-degree angle over the inferior orbicularis muscle. The remaining muscles are similarly overlapped and sutured together.

The skin layer is the final layer to consider and many techniques have been described in the literature, including simple linear closure, Z-plasty, and W-plasty for the lateral portion of the cleft. The simple line closure can result in a noticeable long, linear scar that can contract or become hypertrophic. The Z-plasty, positioned to lie within the nasolabial fold, and the W-plasty minimize this and prevent lateralization of the commissure. Final consideration must be given to the vermilion. Although direct suturing of mucosa to skin is most common, various flaps have been described which result in superior repositioning to further minimize scar visibility.

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