Case Report / Olgu Sunumu

Congenital macrostomia: a case report

Doğuştan makrostomi: Olgu sunumu

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Macrostomia is often associated with the first and second branchial arch syndrome. Depending on the involvement area, appearance may vary. In isolated cases of macrostomia, the cleft usually terminates at the medial border of the masseter muscle. The goal of macrostomia reconstruction is to achieve functional, symmetrical and accurate mouth commissure with minimal scar. In this article, we present an eight-year-old girl case with isolated bilateral macrostomia treated with vermillion-square flap method. We recommend this method for patients with mild to moderate macrostomia.

Key Words: Macrostomia; vermilion-square flap; W-plasty.

Makrostomi genellikle birinci ve ikinci brankiyal ark sendromu ile ilişkilidir. Dış görünüş tutulum alanına bağlı olarak değişebilir. İzole makrostomi olgularında, yarık genellikle masseter kası medial sınırında sona erer. Makrostomi rekonstrüksiyonunundaki amaç, fonksiyonel simetrik ve doğru ağız komissürünü en az yara ile elde etmektir. Bu makalede, izole iki taraflı makrostomisi olan ve vermillonu-kare flep yöntemi ile tedavi edilen sekiz yaşında bir kız olgu sunuldu. Biz hafif ve orta derecede makrostomili hastalarda bu yöntemi önermekteyiz.

Anahtar Sözcükler: Makrostomi; vermilion-kare flep; W-plasti.

Congenital macrostomia, also called transverse facial cleft, is a rare anomaly and accounts for every one of 100-300 cases of facial cleft.[1] The cleft is thought to result from interruption of mesoderm migration, which enables union of the maxillary and mandibular processes in the fourth and fifth weeks of embryonic development.[2] Macrostomia usually accompanies the first and second branchial arch syndromes, though it may rarely appear as unilateral or bilateral isolated deformity.[3] The clinical picture of macrostomia ranges from a slight dislocation of the commissure to a cleft extending towards the temporal bone and absence of ears in patients with the first and second branchial syndromes, while isolated macrostomia is characterized by a cleft extending from the mouth commissure to the anterior edge of the masseter muscle.[4]

The treatment of macrostomia varies with severity and extension of the cleft, and presence of accompanying deformities, but is directed towards reconstruction of the commissure with

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Figure 1. *Planning vermilion square flap and lazy W-plasty around the cleft.*

minimal scar and functionally and aesthetically good results.[5] There have been various treatment alternatives to achieve the desired results. In this report we present our reconstruction of macrostomia in a case using the vermilion square flap described by Eguchi et al.[6] and the obtained results, review other treatment alternatives and discuss advantages and disadvantages of macrostomia repair with the vermilion square flap.

Surgical technique

The transverse cleft was reconstructed bilaterally under general anesthesia using the vermillion-square flap described by Eguchi et al.[6] First, bilateral upper and lower lip mucosa points where normal mucosa ends and the cleft starts were marked. Second, a square flap was designed on the vermilion mucocutaneous line of the lower lip and a lazy W-plasty was performed around the cleft (Figure 1). After removal of dermal and mucosal remnants, the superior and inferior fibers of the orbicularis oris muscle were placed on the anterior and posterior sides respectively and were sutured one top of the other to obtain a natural appearance of the commissure and to preserve sphincter function (Figure 2). Then, the inferior based flap was rotated and placed on the upper lip achieving an appropriate location of the commissure without a suture line on the commissure. Last, a straight suture was put on the oral mucosa and the buccal skin was closed with lazy W-plasty (Figure 3).

Figure 2. *After removal of dermal and mucosal remains, superior and inferior fibers of the orbicularis oris muscle were placed on the anterior and posterior sides respectively.*

CASE REPORT

An eight-year-old girl presented with bilateral congenital deformity of the commissure and was diagnosed with macrostomia and prominent ear (Figure 4a). Macrostomia was limited to tissues around both commissures and did not extend beyond the masseter muscle, the oral commissure was moved to the lateral side and normal and pathological vermilion margins were pronounced bilaterally. The rest of the face was normal. Examinations of other systems and genetic investigations showed no accompanying abnormality.

Macrostomia was reconstructed with Eguchi's vermillion square flap and prominent ear deformity was reconstructed with bilateral modified Mustarde technique. There was no postoperative

Figure 3. *Appearance of the patient following completion of unilateral reconstruction.*

Figure 4. **(a)** *Preoperative appearance of the patient with prominent ear and macrostomia caused by bilateral facial cleft.* **(b)** *Appearance of the patient one year after repair of macrostomia and prominent ear.*

complication and the patient was discharged within two days of operations. One year after the operations, the result was both functionally and aesthetically satisfactory (Figure 4b).

DISCUSSION

Mild or moderate congenital macrostomia causes only cosmetic problems, whereas severe macrostomia causes nutritional and speech problems also. It is widely accepted that macrostomia should be corrected as early as possible.[7] Although macrostomia is rarely encountered, there are a wide variety of techniques for its repair.[8-10]

The goals of macrostomia repair are to achieve a symmetric, natural oral commissure and lips with minimal scar and to form integrity of the orbicularis oris muscle and thus restore oral sphincter function. As in the repair of lip clefts, there is a general agreement that reconstruction of the orbicularis oris muscle is important in all surgical techniques used for treatment of macrostomia. May, who was the first to describe macrostomia repair, emphasized that a web may occur around the oral commissure when repair of the muscle was not done properly. Boo-Chai called this deformity "Gold Fish Mouth".[11,12] Another important point in repair of the muscle is that orbicularis oris muscle fibers of the upper and lower lips should overlap each other in order to be able to achieve a natural mouth contour.

A disadvantage of most of the surgical techniques described for macrostomia repair is that the resultant scar tissue is perpendicular to the tension lines of the face and is therefore pronounced, and the oral commissure moves laterally due to scar contracture.[13] Even though Z-plasty performed on the edge of the oral commissure allows appropriate localization of the commissure, it may not prevent lateral movement of the commissure. The disadvantage of Z-plasty is that it causes more scar tissue on the cheeks.[14,15]

It is recommended that flaps should not be placed exactly on the commissure to obtain a natural appearance. A flap transposed from the upper and lower lips to the commissure may provide a natural appearance of the commissure and prevent displacement of the commissure to the lateral side. Eguchi's technique, which we used for reconstruction of macrostomia in a case reported here, is an improved form of a triangular skin and mucosal flap described by Onizuka.[16] Kaplan and Sugihara modified the triangular skin and mucosal flap, but they both observed marked scar tissue.[17,18] Later, Eguchi et al.[6] described elevation of a square flap from the lower lip and its combination with W-plasty. W-plasty was first used by Bauer for treatment of macrostomia and is known to provide better cosmetic results than Z-plasty in reconstruction of facial scars.[4,19]

Combination of a square flap and W-plasty, described by Eguchi et al.[6], provides

reconstruction of the commissure with success and it is advantageous because it does not cause contractures or dislocation of the commissure. In addition, it leads to less facial scars than Z-plasty. In fact, we observed that the combination of the square flap and W-plasty caused less scar tissue but it did not cause any contractures or dislocation of the commissure within one-year follow-up. The patients' families were satisfied with the results. In view of its advantages, we recommend the technique for macrostomia repair.

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