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Case report

TESSIER No. 7 CLEFT WITH MACROSTOMIA: CASE REPORT

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Abstract

Craniofacial clefts are rare conditions with an incidence of 2-7 per 100,000 births. Because of the limited experience with such cases, they pose a great challenge for the head and neck reconstructive surgeon. The Tessier no. 7 cleft is the most common and is found in various clinical presentations starting from a small notch in the oral commissure to a significantly disfiguring entity, sometimes as a part of Hemifacial microsomia or Goldenhar's Syndrome. We present a case of a young girl with a unilateral Tessier no. 7 cleft with macrostomia and bilateral facial skin tags, the surgical options, as well as the follow-up results of the repair.

Keywords: lateral facial cleft, macrostomia, Tessier no.7, surgical treatment

Introduction

Transverse facial clefts, sometimes called macrostomia are rare. Their exact etiology is unknown, but the pathogenesis involves failure of fusion of the maxillary and mandibular prominences where normal migration of mesenchyme is aimed at reducing the wide primitive stomatodeum[1]. Failure to achieve fusion around the corners of the mouth leads to macrostomia, while excessive fusion can cause micro- or astomia. The incidence has been estimated to be 1:80,000 live births[2]. This anomaly can be isolated or a part of a larger clinical syndrome, most often Hemifacial microsomia. It can present usually unilaterally, but bilateral cases have been reported, with variable severity[3].

Many classifications exist that try to encompass the vast variety of possible variations of craniofacial clefting. However, Tessier's classification of all possible meridians along which facial clefting occurs, has stood the test of time[4]. It uses the horizontal axis that connects the medial and lateral orbital canthi as an equator, while all the fourteen "northbound" and "southbound" meridians, or time zones, represent the types and directions of possible craniofacial clefts. All of these conditions have many forms, and involve the facial soft tissues, as well as the underlying facial skeleton. Tessier describes the orbital cavity as a key area where most of the clefts intersect and affect its bony continuity.

Cleft no. 7 is the most lateral variation of facial clefting that is found sometimes in hemifacial microsomia and Treacher-Collins Syndrome. It is a temporo-zygomatic cleft, sometimes even with absence of the zygoma, congenital deformities of the mandibular ramus, condyle and coronoid process. Maxilla is short in the vertical plane and the alveolar bone is hypoplastic, with incomplete clefts in the molar region and between the maxillary tuberosity and the pterygoid plates. Soft tissue abnormalities include malformations of the ear, and absence of the temporal muscle. This cleft is both facial and cranial. Facial manifestations include

macrostomia with extension of the cleft laterally from the oral commissure and preauricular skin tags. The lateral cleft itself can range from a small furrow on the angle of the mouth, to complete cleft ranging to the external auditory meatus, even the temporal fossa[5,1].

Case report

We present a case of a young girl with a right-sided unilateral Tessier no. 7 cleft, resulting in a significant macrostomia as well as several preauricular skin tags bilaterally.

She is the second child, born from an uneventful and monitored pregnancy, by a healthy non-smoker mother. Her parents and siblings display no signs of genetic disease and the non-immediate family was negative for any type of clefting disorder or facial malformation.

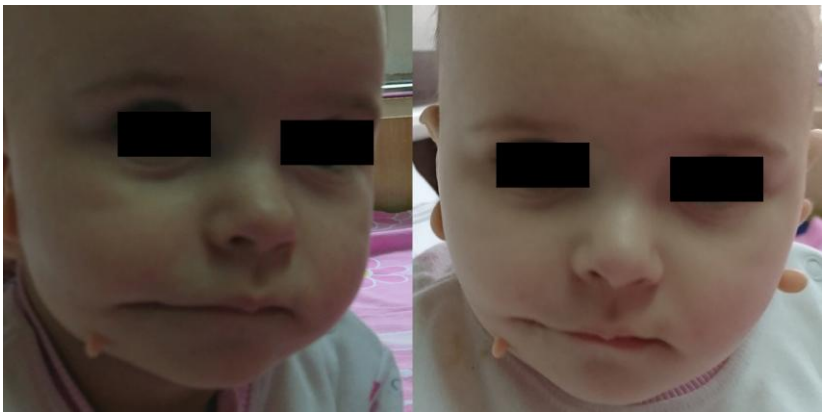

Fig. 1. Preoperative findings

She is otherwise healthy and underwent a repair of the lateral cleft under general anesthesia when she was 15 months old. Anthropometric measurements[6] were taken to ensure that the newly planned commissure was symmetrical to the healthy side. Namely, the distance from christa philtri to cheilion (Cph – Ch) on the healthy side was measured and the distance was transposed to the affected side, as well as the distance from labrale inferior to cheilion (Li – Ch) on the lower lip, thereby identifying the exact length of upper and lower lip vermilion to obtain labial symmetry. Then, a quadrilateral inferomedially based vermilion flap on the lower lip was constructed with its base ending just short of the ideal angle position. The flap was raised, the orbicularis oris bundles ware dissected in the subdermal plane and sutured together to form the new oral muscular modiolus.

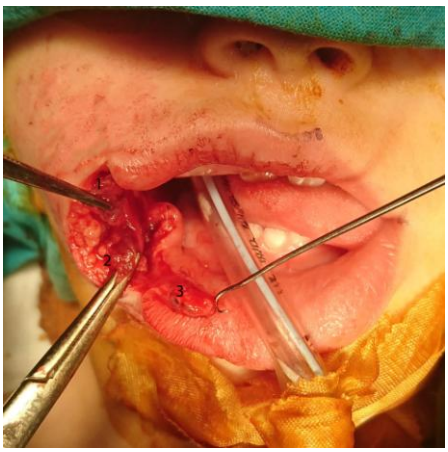

Fig. 2. Surgical repair. 1 and 2 - Orbicularis oris muscle fibers forming the new modiolus. 3 - Lower vermilion flap

Muscle and subcutaneous tissues were closed with 5/0 polyglactin 910 suture (*Vicryl, Ethicon Inc*.). A Z-plasty was planned to break up the axis of the scar, in such a way so that one of the arms corresponded with the nasolabial fold, while the other part was re-approximated in a linear fashion with 6/0 monofilament polypropylene suture (Prolene, Ethicon Inc.).

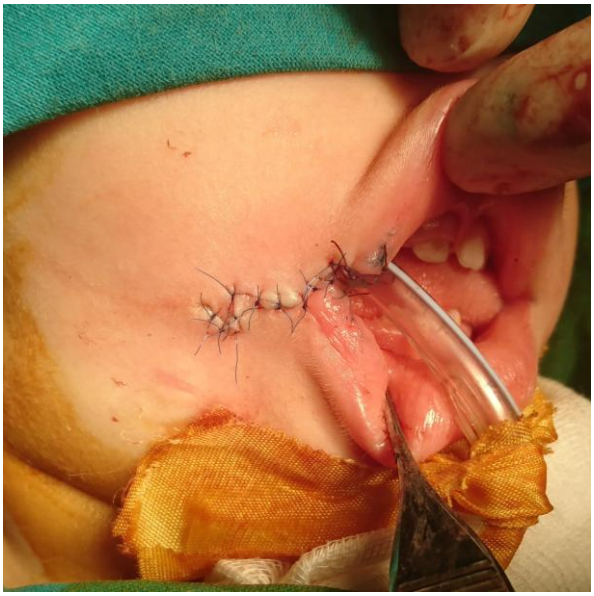

Fig. 3. Z- plasty and final skin closure

Her skin tags were also excised and closed primarily. The immediate postoperative result showed good facial and labial symmetry. A standard skin care regimen was employed; the child was kept overnight and resumed oral feeding with a spoon and bottle on the same day. The parents were instructed for proper wound care [7] and sutures were removed on the sixth postoperative day. Regular check-ups were scheduled and the child remains in good health and satisfactory labial and facial aesthetics. Now, aged 5, she shows only slight commissure migration, almost ideal labial symmetry, no functional deficits and completely normal appearance of the facial skeleton and dentition.

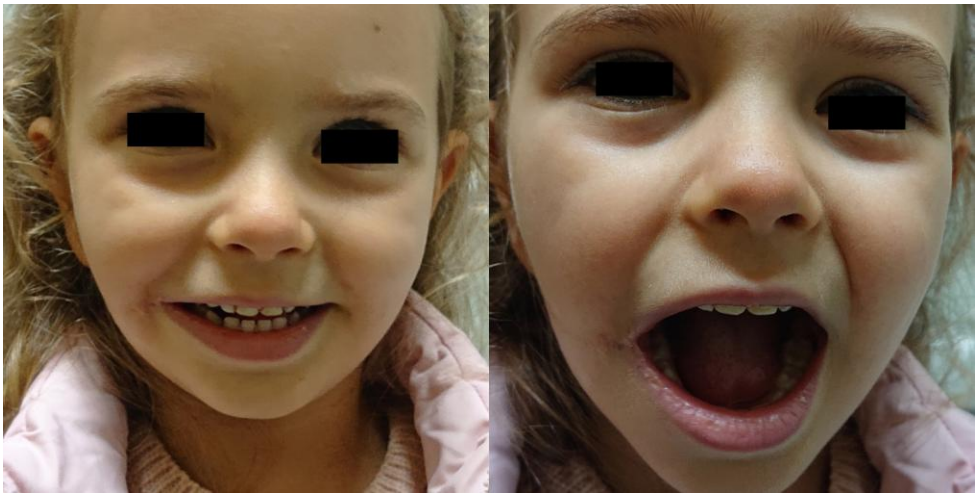

Fig. 4. Postoperative appearance at age 5

Some minor refinements of the commissure and lower lip vermilion would have to be done after growth caseation.

Discussion

Lateral facial clefts are a rare clinical entity and can manifest in varying severity. The orbicularis oris muscle has no continuity at the site of the cleft. The upper fibers are attached to zygomaticus major muscle, and the inferior fibers to the risorius and lower labial depressors. In the extreme cases, a cleft exists in the buccinator muscle and parotid duct and gland can be absent. Because of the unopposed action of the labial depressors, there is pulling of the lip angle downward and laterally, which can cause trouble during feeding, speech and poses an aesthetic problem[1]. Standard of care is an early closure of the cleft as to avoid these issues.

Many techniques exist that can address this deformity, all with unique advantages and drawbacks. Most of them involve the use of a vermilion flap, either from the lower or upper lip, excision of part of the redundant tissue around the commissure, and either using some type of a Z-plasty, W-plasty, wavy line technique, broken geometric line closure, or a linear closure[8]. The advantage of the linear closure is in its simplicity, however there are major drawbacks, such as the contracture deformation and subsequent lateral migration of the labial angle. Z-plasty technique has the tendency to provide a more desirable scar, with one of the arms of the Z running parallel to the nasolabial fold, thereby providing some camouflage. Running W-plasties have a tendency to increase the length of the scar. Geometric methods have also the disadvantage of creating unexpected lines in the cheek and nasolabial area, especially during facial animation[9]. Whichever technique is used, and all authors agree that the most important step is the exact alignment and re-approximation of the muscle bundles from the orbicularis oris[10]. This forms the new muscular modiolus and gives the lip its functionality. Precise intraoperative anthropometric measurements will serve as a surgeon`s guide, so that whichever techniques are employed, the now commissure is placed properly.

Conclusuion

It is difficult to create a new corner of the mouth which has normal function and appearance. We emphasize the need for familiarization with this rare facial cleft, because there is hardly a center with a major series of such cases. Cases need to be evaluated and treated by a multidisciplinary team experienced in craniofacial surgery and congenital clefts to obtain the most optimal outcomes. The Z-plasty technique is simple and can be adapted to the need of almost every clinical scenario. Further follow-up will show the need for technical refinements and further revisions of the result, thus enabling us to better understand the most appropriate technique for addressing this type of cleft.

Conflict of interest statement. None declared.

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