Staged Reconstruction of a Tessier Number 4 Facial Cleft

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Tessier number 4 cleft is an extremely rare facial cleft. We report a case of Tessier number 4 facial cleft recently managed in our center. The approach to facial clefts is multi-disciplinary and may differ between centers. Multiple surgical interventions are required as well as a good psychosocial support. The child underwent a craniofacial reconstruction at 6 months of age, followed by a left macrostomia repair 9 months later and transcranial correction of the right orbital dystopia with eyelid reconstruction.

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The craniofacial cleft is a rare congenital anomaly with a reported incidence of only 1.5 to 5 per 100,000 births[1]. One of the methods for classification of the facial cleft is by the Tessier classification. This method was first described by Paul Tessier in 1976 and is still widely used today. It is based on 15 different anatomical positions of the cleft in relation to the orbit[2]. They are numbered from 0 to 14, whereby clefts 0 to 7 are facial defects from the lower hemisphere, clefts 9 to 14 are from the upper hemisphere and cleft eight forms the equator.

A multidisciplinary approach is essential in the management of these cases as the facial cleft involves defects of not only the skin and muscles, but also all the other structures surrounding it. Multiple surgical procedures need to be performed, one at a time as the child grows. It is important to distinguish between functional issues and cosmetic or aesthetic issues to decide which procedure to perform first.

The aim of this presentation is to report the multidisciplinary sequence of procedures to manage Tessier Number 4 Facial Cleft.

THE CASE

A male infant presented with facial anomalies at day one of life. He was born at term gestation with a birth weight of 2.88 kilograms. The plastic and reconstructive surgery team found that the baby had right Tessier number 4 cleft and termed it oro-ocular cleft. It begins laterally to the cupid bow and skirts around the nose to end in the lower eyelid medial to the punctum. Pediatric surgery, otorhinolaryngology (ORL), ophthalmology, neurosurgery as well as oral and maxillofacial surgery (OMFS) were consulted.

The right facial cleft was noted to be extending intra-orally to the right pre-maxillary region and orbital floor. The right lower eyelid was missing. No anomaly was seen within the nasal cavity. Left macrostomia was also noted. A computed tomography of the orbit revealed a unilateral right paramedian cleft lip and palate extending to the inferior-medial part of the right orbital floor. The feeding was established initially via nasogastric tube and later bottle-feeding and breastfeeding.

The patient was discharged from the neonatal ward at day 7 of life. Six months later the patient was admitted for the first stage of craniofacial reconstruction, the patient's weight was 6.8 kilograms see figure 1. Intraoperatively, a large coloboma of the right lower eyelid with orbital dystopia was found. The lower eyelid lacrimal apparatus was missing. The right upper eyelid was missing and the orbital floor was defective. Also, a complete right cleft lip and alveolus was found. His soft and hard palate were intact. The edges of the cleft were excised and the lower eyelid margin was reconstructed by

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approximating the cleft edges and buccal mucosa, see figures 2 and 3. The orbicularis oris muscle was reorientated and repaired. The right facial muscles were anchored medially to fill the infraorbital defect and the skin was approximated. The oral mucosa and lip vermillion were sutured. A temporary right tarsorrhaphy was performed, see figure 4. Postoperatively, the patient was stable and was discharged four days later.

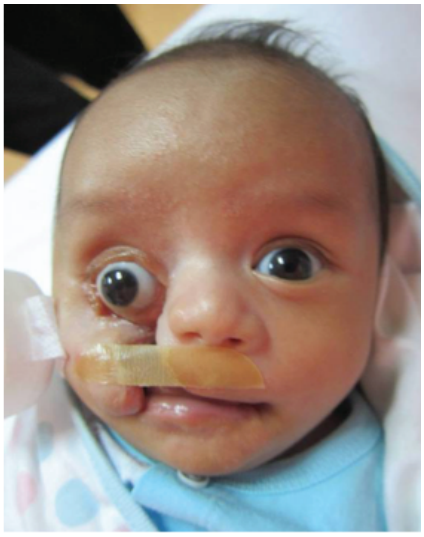

Figure 1: Six Months Old, with External Splinting and Moisture Chamber for the Right-Eye

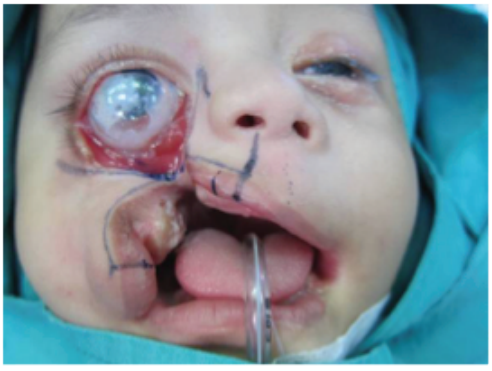

Figure 2: Soft Tissue Repair Preoperative Markings

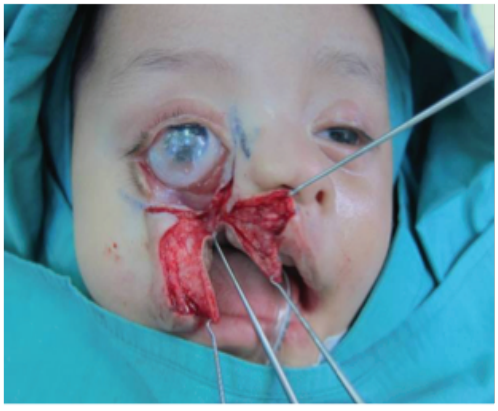

Figure 3: Raised Surrounding Flaps

Figure 4: Temporary Right Tarsorrhaphy

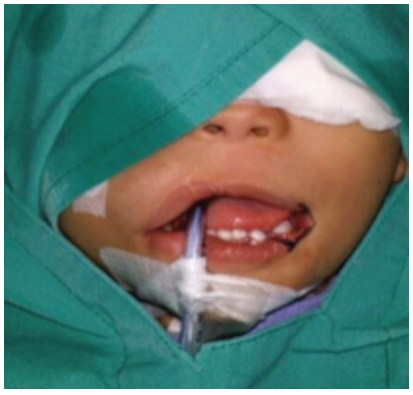

Figure 5 (A): Left Macrostomia Repair Preoperation

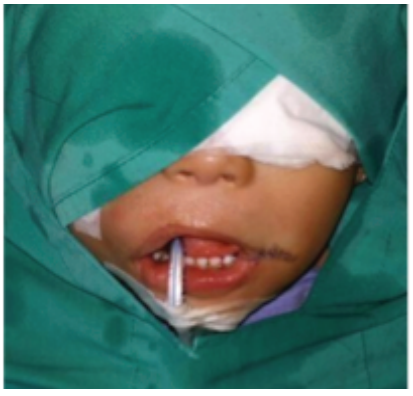

Figure 5 (B): Left Macrostomia Repair Postoperatively

Figure 6: Three Weeks Post Macrostomia Repair

At the age of one year and three months, the patient was readmitted for repair of his left macrostomia. Intra-operative findings revealed that the left part of the orbicularis oris muscle was attached to the corresponding alveolus with adjacent muscles. An incision was made from the left angle of the mouth extending laterally until the orbicularis oris muscle was identified. The muscle was released from its abnormal attachment at the alveolus superiorly and inferiorly. It was then reconstructed to recreate the modiolus. The skin flap on the upper lip was rotated inferiorly to form the angle of the mouth. The mucosal layer was then repaired and the skin sutured, see figures 5A and 5B. He had an uneventful postoperative period and was discharged the following day, see figure 6.

The patient was readmitted three months later for another craniofacial reconstruction and transcranial correction of right orbital dystopia with calvarial bone graft followed by right eyelid reconstruction.

DISCUSSION

Tessier number 4 cleft is a technically challenging craniofacial malformation to repair. The abnormalities of the soft tissues include a cleft lip extending to the cheek, orbital dystopia with reduction of the oculo-alar and oculo-oral distance[4]. These abnormalities impede the normal physiological functions, such as feeding and speech. Correcting the functional issues is the main aim of initial craniofacial surgery during infancy[5].

Alonso et al described the different clinical features in 21 cases; all showed a cleft upper lip and 19 patients had lower eyelid coloboma. Ten out of the 19 cases had dystopia[6]. The management is difficult because the incidence is extremely rare[2,7,8]. Resnick main concern was the protection of the eye; soft tissue deformities and bone grafting should be corrected early followed by the lower eyelid reconstruction[8]. In our center, tissue repair was performed first followed by bone grafting a few months later. CT might help in further assessment of the patient before any definitive surgical correction is performed[9]. Facial clefts may be both non-syndromic and syndromic. Syndromes associated with facial clefts include Goldenhar syndrome, amniotic band syndrome and many others. This patient, however, was developmentally normal for his age and had no neurological deficit[10,11].

CONCLUSION

The management of facial clefts involves the correction of functional issues while attending aesthetic outcomes. The structural facial anomaly that results from the cleft could be debilitating to both the patient and the parents. A multi-disciplinary approach in managing such patient is paramount due to the complexity. Besides the obvious issues, the psychosocial aspect of this matter must also be looked into.

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