RECONSTRUCTION OF RARE CRANIOFACIAL CLEFTS: AN EARLY EXPERIENCE

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ABSTRACT

Objective: *To share our experience regarding the management of rare craniofacial clefts.*

Methodology: *The study was and conducted at the department of plastic and reconstructive surgery, Hayatabad Medical Complex Peshawar and Al-Shifa Health Care Center Peshawar, from January 2008 to December 2009. A total of 08 patients with rare craniofacial clefts were treated. Diagnosis was based on clinical examination and radiographic findings on CT scan. Tessier's classification was used to describe these clefts. There were two case of unilateral cleft 7, two cases of bilateral cleft 7, two cases of cleft 30, one case of bilateral cleft 4 and a single case of proboscis lateralis. All the patients were treated surgically and the different clefts were repaired or reconstructed executing different procedures.*

Results: *We achieved good results in all the cases regarding soft tissue defects. No surgical intervention was performed for bony defects.*

Conclusion: *Proper assessment and referral of these rare craniofacial clefts to plastic surgeon in necessary for the optional surgical outcome.*

Key Words: *Craniofacial cleft, reconstruction*

INTRODUCTION

Craniofacial clefts are major clefts affecting the face, the cranium, or both. These congenital clefts cause distortion of the face and cranium with deficiencies or excesses of tissue that cleave anatomic planes in a linear fashion[1]. They are the most disfiguring facial anomalies. Craniofacial clefts exist in varying degrees of severity and almost all of them occur along the predictable embryologic lines. These clefts can be either complete or incomplete and can appear alone or in association with other facial clefts. The severity of craniofacial clefts varies considerably, ranging from a barely perceptible notch on the lip or on the nose or a scar-like structure on the cheek, to a dramatic separation of all layers of facial structures. In addition one cleft type can manifest on one side of the face while a different type is present on other side[1,2].

Craniofacial clefts are much rarer than simple cleft lip/palate. The exact incidence of craniofacial cleft has not been identified because of their rarity. Most often the milder forms are likely to go unrecognized. However the reported incidence of rare craniofacial clefts is 1.5 to 6.0 per 100,000 live births. The incidence of rare craniofacial clefts compared with common cleft lip and palate malformations may range from 9.5 to 34 per 1000[2]. The etiological factors are genetic factors, radiation, infection, maternal metabolic disorders, and use of drugs or chemicals by mother during pregnancy and dietary deficiencies (folic acid) [1-3].

Varieties of separate classifications are projected with regard to cleft lip and palate[4]. The clefts falling under the category of "Rare Craniofacial Clefts" are also classified by many authors differently. One of the popular classifications is that proposed by Paul Tessier's as "Rare Cranio Facial Clefts"[4,5]. The central point of reference in this classification is the orbit[5].

Paul Tessier's classification system based on the basis of clinical findings in which the clefts are numbered from 0 to 14 depending upon the relationship to the orbit. The orbits divide the face into upper and lower hemispheres and separate the

cranial clefts from the facial clefts. At times, facial clefts extend through the orbit to become cranial clefts. The clefts are numbered so that the facial component of the cleft and the cranial component always add upto 14: 0 and 14, 1 and 13, 2 and 12, 3 and 11, 4 and 10, 5 and 9, 6 and 8. Tessier cleft number 7 is lateral most craniofacial cleft. The soft tissue and skeletal components of a cleft are seldom affected to the same extent. The skeletal landmark tends to be more constant and reliable than the soft tissue landmarks[5].

Apart from the common cleft lip and palate deformity the treatment of other types of craniofacial defects need much more complex diagnostic and treatment modalities[4-6]. Due to their complexity, the individual degree of cleft formation and the different structures and organs involved, successful reconstruction and rehabilitation in almost all the case demand a multistep and multi-professional procedure[6].

Besides careful examination, imaging techniques are necessary to assess the individual degree of skeletal involvement in the cleft formation. For correct diagnosis modern imaging techniques such as CT, MRI and 3-D CT allow better pre-op understanding of the problem and planning of the surgical procedures.

Considering these requirements we would like to present our experience in the surgical management of 08 patients with rare facial clefts.

METHODOLOGY AND RESULTS

Over a period of two years i.e. from January 2008 to December 2009 a total of 08 patients with rare craniofacial clefts were treated. Diagnosis was base d on clinical examination and radiographic findings on CT scan. In our study we used the Tessier's classification to describe the craniofacial clefts because it gives the anatomical description of the cleft and is internationally recognized as standard reference (Figure 1). This classification has the merit of permitting all clinicians who deal with patients having these malformations (pediatricians, geneticists, foetal pathologists, surgeons, and obstetricians, sonographers) to readily communicate with one another since it is based on anatomical and clinical considerations[5].There were two cases of unilateral cleft 7, two cases of bilateral cleft 7, two cases of cleft 30, one case of bilateral cleft 4 and a single case of proboscis lateralis.

CASE1:

A female baby at the age of 3 months was referred to our unit with bilateral Tessier cleft 4. The cleft had spared the philtral column and the entire nose. The cleft commenced medial to the oral commisures and passed lateral to the nasal ala. The orbicularis muscle was located in the lateral lip elements and there was no muscle in the midline. The clefts passed through the cheeks into the lower eyelids. The medial canthus and lacrimal systems were normal. Microphthalmia observed in the left eye. The skeletal component involved the maxillary sinuses but the medial walls of the sinuses were intact. The clefts were present medial to the infraorbital formina and terminated at the medial aspect of orbital rim.

Operative Technique:

Only soft tissue reconstruction was performed with local flaps. The grooved or hypoplastic tissue extending in the path of the cleft between the upper lip and the lower eyelid was excised. The rotation advancement repair of the cleft lip was performed. The lateral cheeks advanced over the cleft region to close the soft tissue defect.

Figure 1: Tessier's Calssification

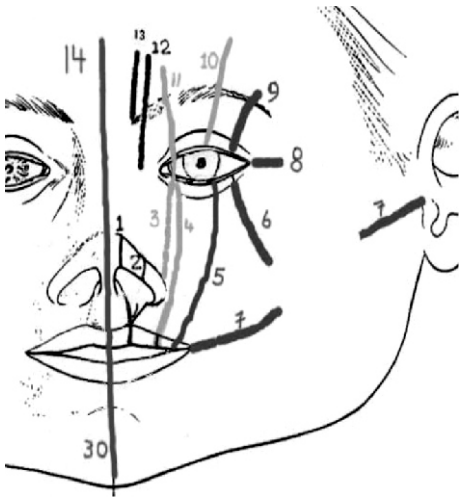


Figure 2 a: Pre-operative bilateral Tessier's # 4 Cleft

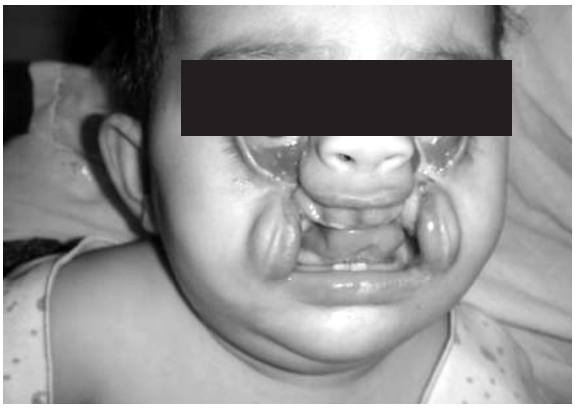
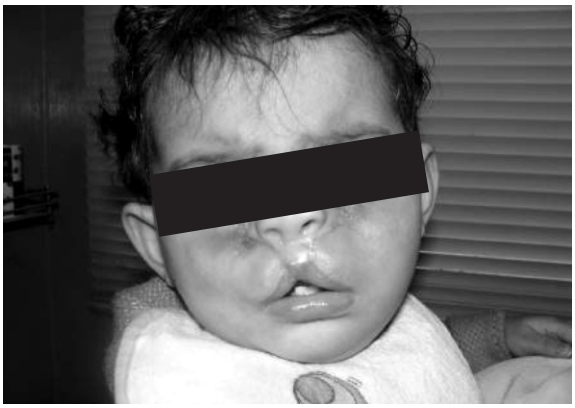


Figure 2 b: Post-operative bilateral Tessier's # 4 Cleft



The incision at the lower lid margin is continued 1-cm lateral to the lateral canthus. The nasal soft tissue is dissected to rotate the nasal ala caudally. The medial canthal ligament is dissected and medial canthopexy is performed with non-absorbable suture.

The conjunctival fornix is reconstructed with mucous tissue within the cleft, and orbicularis oculi muscle is preserved and reconstructed. The cheek flap is advanced to the medial and cephalic direction and tightly sutured to the periosteum at the piriform aperture. A small rectangular flap, designed on the medial margin of the cheek flap, is inserted into the medial canthal area. Lip repair was performed by using the bilateral rotation-advancement technique, and the white skin roll and vermilion on the central portion of the lip were reconstructed with both lateral vermilion flaps, which were sutured in the midline (Figure 2a & b).

CASE 2 & 3:

Two patients aged 3 years and 4 years had unilateral cleft 7. Both were males & the cleft was on left side. On clinical examination there was no hemifacial microsomia. The soft tissue manifestation of cleft 7 in these patients was a variable degree of macrostomia. Malformation of the external ear was not found in any of the patient.

Surgical Technique:

The surgical technique consists of the creation of a straight-line closure of the mucosa, a red-lip commissural flap rotated from the superior part of the lip (which, as a rule, is an extended part of the natural red-lip component) , a modiolus muscle reconstruction at the confluence of the orbicularis oris, zygomatic major, risorius and depressor anguli oris muscles performed and modified z-plasty flap for skin closure.

The distance from the midpoint of the cupid's bow to the normal commisure is used for reference. The position of the new commissure on the cleft side is marked at the vermillion-cutaneous junction on the upper lip and by sighting a perpendicular line, the corresponding point is noted at the vermillion- cutaneouis line on the lower lip. This proposed position of the commissure is confirmed by measuring the peak of cupid's bow on each side. Another mark is made on the upper lip vermillio-mucosa approximately 3-4 mm medial to the proposed commissural point as the inset for the inferiorly based vermillion mucosal flap. The cutaneous-mucosal margin of the cleft is marked and the critical points are tattooed with gention violet dye. After infiltration of local anesthetic with epinephrine, the cutaneous mucosal margins are incised to expose the underlying orbicularis oris musle. The splayed muscular elements are dissected in the subdermal plane. An inferiorly based rectangular vermillio-mucosal flap is raised off the underlying muscle (pars marginalis) on the lower lip extending the base just lateral to the lower labial point of the new commissure. Next the orbiculris oris muscle is approximated side to side from lateral to medial. Near the position of the anticipated commissure, the pars marginalis is opposed end to end to complete construction of the sphincter. The cutaneous points of the new commissure are approximated by a dermal suture and the skin is closed in two layers. After the mucosal dog-ear is excised, the buccal mucosal cleft is closed, lateral to medial, toward the commissure and a Z plasty is performed. The vermillion – mucosal flap is draped into the commissure and ecured to the pars marginalis (Figure 3,4 a & b).

Figure 3 a: Pre-operative left Tessier's # 7 Unilateral Cleft

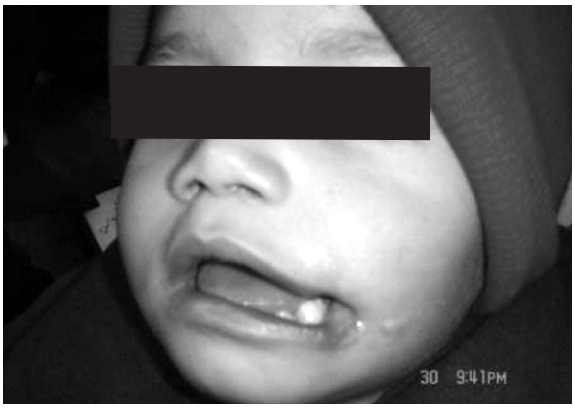


Figure 3 b: Post-operative left Tessier's # 7 Unilateral Cleft

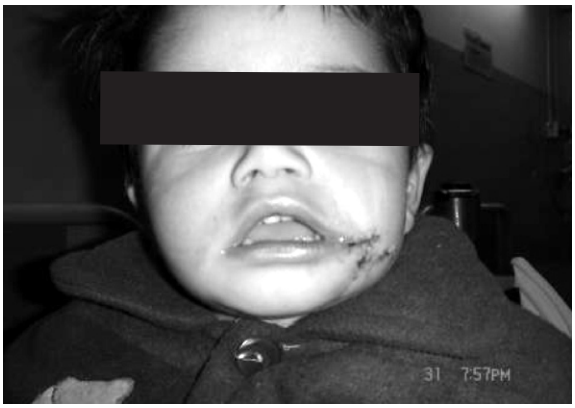


Figure 4 a: Pre-operative left Tessier's # 7 Unilateral Cleft

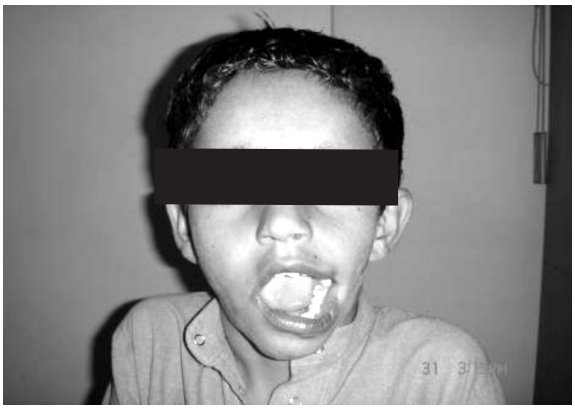
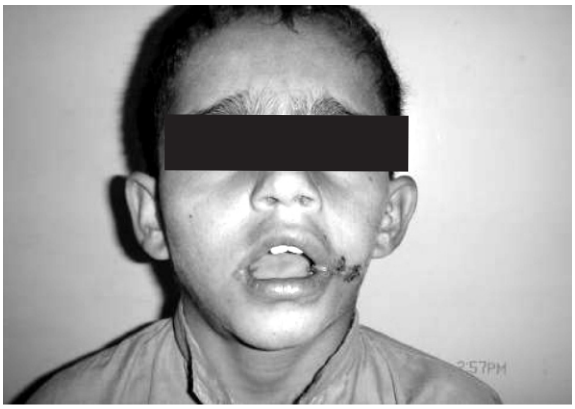


Figure 4 b: Post-operative left Tessier's # 7 Unilateral Cleft



CASE 4 & 5:

Two patients aged 3 years each had bilateral cleft 7. One of them was male and the other female. Right sided hemifacial microsomia was noted in female patient. In both the patients the eaxternal ears were normal and the clefts extended from the oral commissure toward the ear. Tongue and soft palate were normal in both the patients but the muscles of mastication were underdeveloped (Masseter). Parotid glands and ducts were present in both the patients. The posterior maxilla and mandibular rami were hypoplastic in the female patient.

Surgical Technique:

Soft tissue repair performed for the correction of macrostomia. The same surgical technique applied as for the unilateral cleft 7 but on both sides [8,9] (Figure 5,6 a & b).

CASE 6 & 7:

Two patients presented with cleft 30. Both were males. One patient was 18 years and the other was 28 years old. The cleft involved the lower lip in the midline and there was no skeletal involvement. In one patient the lower lip was notched only and in the second patient there was midline involvenement of the more than two thirds of lower lip. Tongue and mandible were spared in both the patients.

Surgical Technique:

In both the patients a W shaped incision designed at the vermilion coetaneous junction of the cleft lip. The skin flaps dissected and elevated. The orbicularis oris muscle dissected separated from the mucosa and stitched in midline. A three layer wound closure done by stitching the mucosa, muscle and skin separately[10,11] (Figure 7,8 a & b).

Figure 5 a: Pre-operative bilateral Tessier's # 7 Cleft

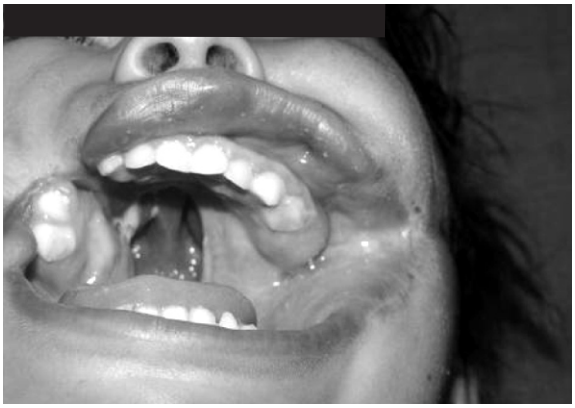


Figure 5 b: Post-operative bilateral Tessier's # 7 Cleft

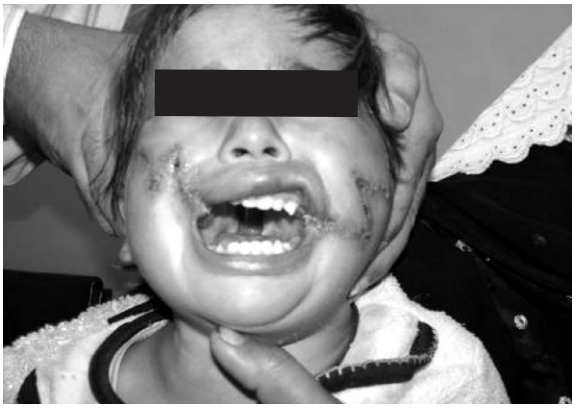


Figure 6 a: Pre-operative bilateral Tessier's # 7 Cleft

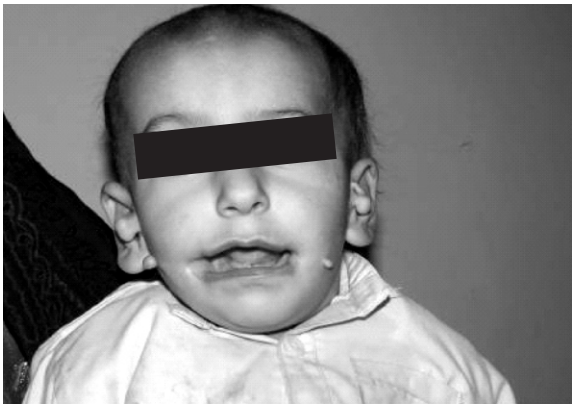


Figure 6 b: Post-operative bilateral Tessier's # 7 Cleft

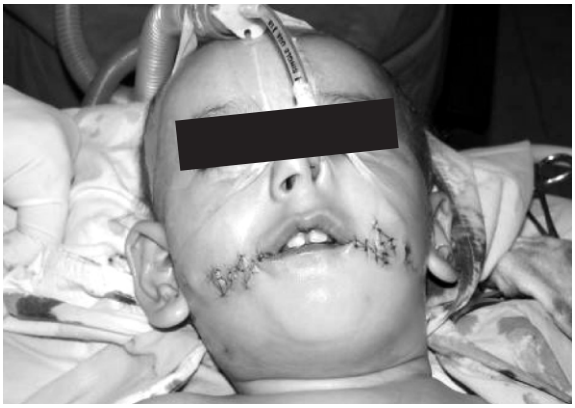


Figure 7 a: Pre-operative Tessier's # 30 Cleft

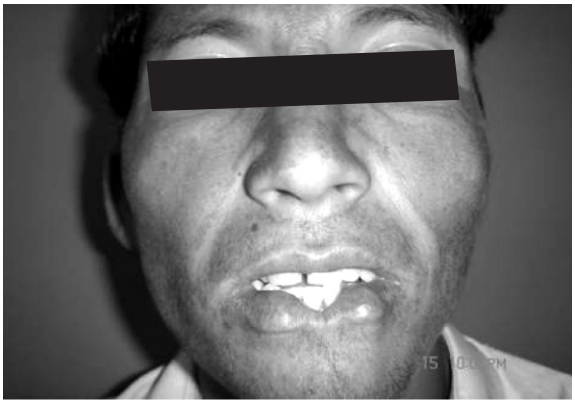


Figure 7 b: Post-operative Tessier's # 30 Cleft

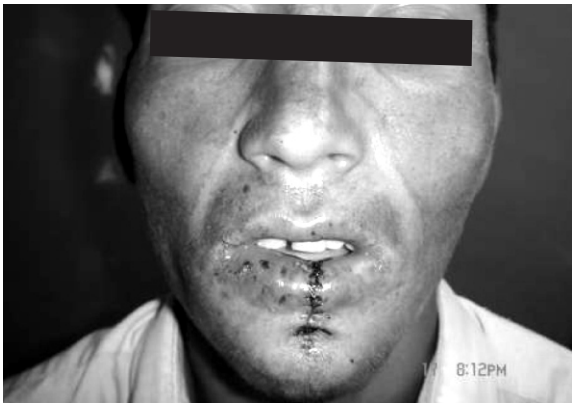


Figure 8 a: Pre-operative Tessier's # 30 Cleft

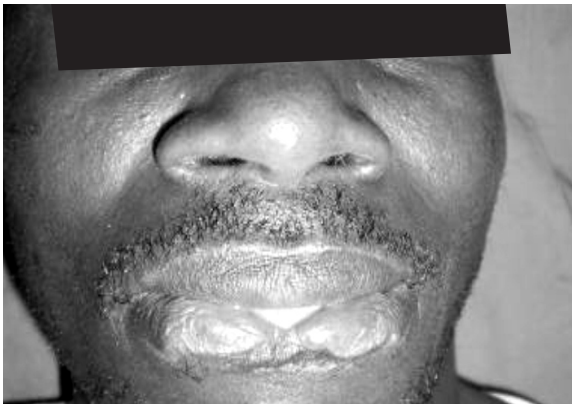
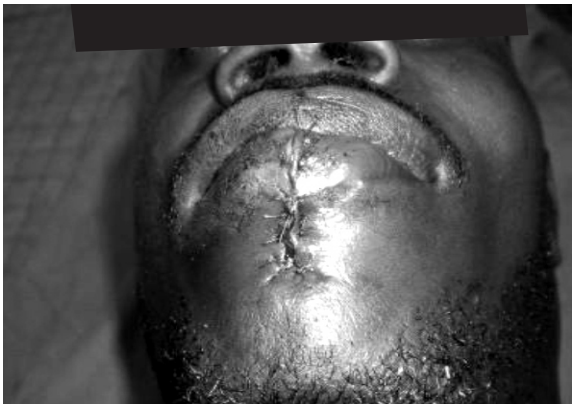


Figure 8 b: Post-operative Tessier's # 30 Cleft



CASE 8:

A young male of 15 years presented to us with proboscis lateralis. It's a very rare congenital anomaly related to the median facial cleft and may be associated with other anomalies of the eye and its adnexa and cleft lip or cleft palate. The incidence of proboscis lateralis is <1 per 100,000[12,13]. The proboscis may be unilateral or bilateral. There is a tube like rudimentary nasal structure which is attached along the embryonic fusion lines between the anterior maxillary process and the frontonasal process. The length of proboscis may vary from 1 to 4 cm in length[14-16]. It has a stratified columnar lining and sometime ends as a blind tract or communicated to the nasal cavity.

The patient which we received had group 2 proboscis on right side. Nasal airway was patent on right side and the patient complained of discharge from proboscis. The patient had no other associated anomaly.

Figure 9 a: Pre-operative probosis lateralis

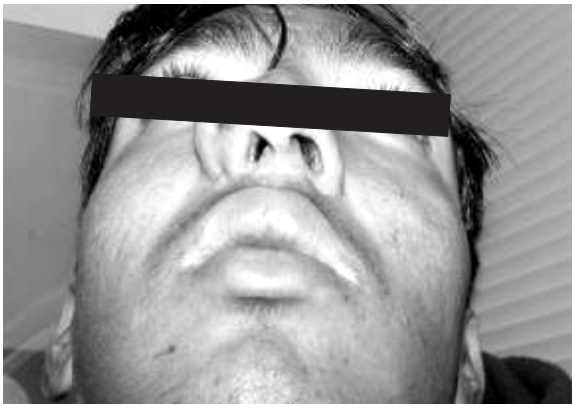
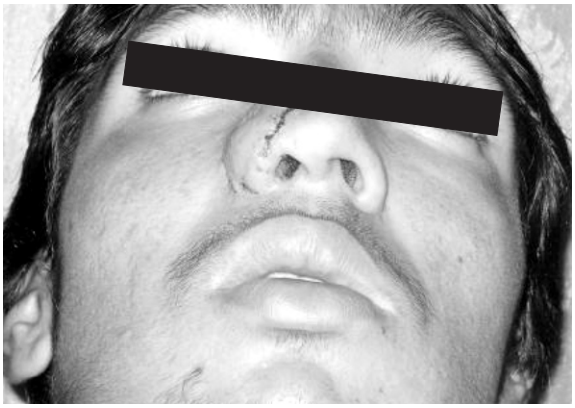


Figure 9 b: Post-operative probosis lateralis



Surgical Technique

Various methods have been suggested for the treatment of proboscis lateralis, ranging from the extirpation of the proboscis, to the tunneling method. We corrected the proboscis by making incision around the proboscis. We lift up the proboscis as a flap. Nasal mucosa of proboscis was separated and excised. Defect in nasal mucosa was stitched and the proboscis skin was replaced. Flap debulking will be done in future (Figure 9 a & b).

A summary of salient features of the eight cases is given in Table 1.

DISCUSSION

Craniofacial clefts are of particular relevance to plastic surgeons, Maxillofacial surgeons & ENT surgeons. While providing surgical treatment to patients with complex craniofacial deformities the plastic surgeons encounter certain inadequacies. Firstly the health status of the patient varies greatly and most of the times it is difficult to verify the fitness of the

Table 1: Clinical features of Craniofacial Clefts in Our series

Case No.	Clinical Features
Case 1	Bilateral Tessier 4 cleft The cleft passing through the cheeks into the lower eyelid. Left microphthalmia
Case 2 & 3	Unilateral Tessier 7 cleft Both the cleft were on left side. No hemifial microsomia. No external ear deformities
Case 4 & 5	Bilateral Tessier 7 cleft Right sided hemifial microsoma observed in one patient Preauricular skin tags in one patient
Case 6 & 7	Tessier 30 cleft The obicularis Oris muslae was affected in the both cases. No bone involvement Probosis Lateralis
Case 8	Left side Probosis Lateralis Probosis connected to the nasal canting

patient for surgical treatment. Secondly the lack of proper equipment may restrict the anesthetist and the surgeon to achieve their task up to the mark. Also the quality of postoperative care requires high levels of expertise and trained staff. To the proper management of such patients can only be accomplished in designated craniofacial centers where they can be operated according to the standardized protocol. It must be realized that the optimal treatment for craniofacial clefts require multiple, staged reconstructive procedures at different ages of the children concerned.

This series of 08 patients with rare craniofacial clefts represents our early experience in soft tissue reconstruction of these deformities. Apart from the surgical and anesthesiologic limitation we also find certain limitations in the proper diagnosis of such cases. The lack of 3D-CT scan confines us to diagnose the deformity on clinical examination and surgical exploration. Due to all these limitations we focused our surgical treatment on soft tissue reconstruction only.

Tessier bilateral cleft 4 was reconstructed with local flaps by cheek advancement and lip repair by rotation advancement technique. No peri-operative complication occurred in this patient.

The unilateral and bilateral cleft 7 benefited from a single stage pure soft tissue repair. We did not find deformities of external ear and occurrence of preauricular tags in any unilateral cleft # 7 patients. This is in contrast to several reports where external ear deformities occur in approximately two thirds of patients cleft 7[7]. However David et al described Tessier no 7 cleft never affecting the external ear[8,9].

Median clefts of the lower lip are rare and just 68 cases have been reported till date[10-13]. Soft tissue repair of cleft 30 as single stage procedure gave good result because there was no bony involvement.

Proboscis lateralis is a rare anomaly among the nasal malformations. Many surgical options described for its correction in multiple steps[14,17,18]. In our case we also planned multistage procedure for its correction. In the first stage we have removed the nasal mucosa from the proboscis and close the nasal defect and in the second stage we will bulk the proboscis that was used for reconstruction of right nostril.

Summarizing our results, we would like to mention that we achieved acceptable results in all the cases where single stage soft tissue reconstruction was the appropriate surgical treatment. Regular follow-up of these patient is also very important for the successful out come of surgical treatment.

CONCLUSION

While summarizing our early experience of the reconstruction of rare craniofacial clefts we would like to state that while operating with limited experience and equipments we have achieved acceptable results in all cases where one stage pure soft tissue reconstruction was the appropriate method of surgical treatment. We hope that in years to come we would be able to provide good care to patients with complex craniofacial deformities.

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