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EXPERIENCE WITH RARE OBLIQUE FACIAL CLEFTS: A CASE SERIES

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ABSTRACT

Tessier facial clefts involve mouth, maxilla, eyes, nose, and forehead. They extend variably to viserocranium and neurocranium. These clefts can be described as oro-occular cleft and fronto-nasal dysplasia. These clefts are numbered from 0 to 14 which represent the extension of the cleft. Tessier clefts have well defined positions and have definite axes. These rare facial clefts impose a major reconstructive challenge and often present with serious asymmetry involving multiple areas of the face. There is a clear three-dimensional underdevelopment of hard and soft tissues of the orbit, maxilla, zygoma, nose and malar region. Further due to deficient growth of all affected tissues in the cleft area, the deformities at birth can become more obvious over the years. We are presenting a series of Tessier 3, 4 and a combination of 3 & 5.

INTRODUCTION

Oblique facial clefts are extremely rare congenital deformities. Their precise incidence in the population is unknown with reported incidence of 0.24% of all facial clefts [1]. Tessier described an anatomical classification system, in which number is assigned to each craniofacial cleft on the basis of its position relative to sagittal midline & the orbit [2-4]. Herein we are describing our experience with these rare clefts.

Case 1

A 3 year old girl presented with a craniofacial cleft. Physical examination of the parents did not show any abnormalities. Questioning of the family revealed that there was no family history of congenital anomalies. There was no family history of craniofacial deformities. The pregnancy had been uneventful. The cleft lip began lateral at the left Cupid's bow, skirting the nose, across the cheek, lateral to the lacrimal punctum undermined the nasal alar base and continued cephalad to end just into the medial third of the lower eyelid. Intraoral examination showed a right cleft of the alveolus and palate. The left orbit was

hypoplastic and displaced on its inferior and lateral axis. A diagnosis of Tessier 4 was made. [Fig 1]

Case 2

A 2 year old boy presented with a craniofacial cleft. The child had an older brother with cleft lip. Parents had a normal physical examination. The mother was on some kind of medicines, nature of which is not known, prescribed by a local practitioner fro anaemia during both pregnancies. The pregnancy had been uneventful. The cleft lip began at the outer third of lip near the angle of mouth, continued cephalad through cheeks to end just into the medial third of the lower eyelid. Intraoral examination showed a right cleft of the alveolus and palate. The right cheek was non-existent. Ventricular septal defect was diagnosed on echocardiography. A diagnosis of right Tessier 5 with congenital heart disease was made [Fig 2].

Case 3

A 4 year old boy presented with a bilateral craniofacial cleft. The child was depressed and blind completely. He had a younger sister who had died due to



congenital heart disease. Parents had a normal physical examination. He was born as preterm by spontaneous vaginal delivery and there was history suggestive of birth asphyxia. On the left side cleft lip began at the outer third of lip near the angle of mouth, continued cephalad through the alveolus and palate. A diagnosis of Right Tessier 3 with left Tessier 5 was made [Fig 3].

Discussion

The incidence of the rare facial clefts is between 1.43 and 4.85 per 100.000 births [3]. Tessier in 1976 described an anatomical classification system in which a number was assigned to each craniofacial cleft, The basis of division was its position relative to the sagittal midline and the orbit [4]. The no. 3 cleft was first described by Morian over a century ago and corresponds to the number 1 of the laterofacial cleft [5]. The known processes of craniofacial fusion fail to explain the embryology of the oblique facial clefts. The Tessier No.5 cleft corresponds to no known embryologic grooves or plane of mesenchymally supported epithelium. One proposed mechanism is the primary failure of development, a neurovascular insufficiency or necrosis, or tears in the

cheeks to end just into the medial third of the lower eyelid. On the right side cleft lip began at the cupid's bow, around the nostrils extending through the lacrimal punctum. Intraoral examination showed a bilateral cleft of

developing maxillary process [6]. Recent experiments suggest that these malformations are caused by a combination of directly tethered tissue migration (such as amniotic bands) and increased local pressure that produce cellular ischemia. The best technique for initial facial repair depends on the type of cleft. All our case are being managed in different stages. Tessier recommended one stage surgery in order to combine and organize the different cutaneous flaps, the extensive dissections, external cantholysis, internal canthopexy and the multiple osseous grafts. A number of different geometric tracings have been described including flap interdigitation techniques to close soft tissues [2]. These techniques use tissues from different facial areas leading to variation in skin colour and texture [7]. The visible scar is localized in the middle of the cheek and exerts traction on the lower eyelid.





Figure 3. Right Tessier 5 cleft with left Tessier 3 cleft



CONCLUSION

These complex clefts require early and staged repair. We recommended early repair using autogenously tissues and as minimal disposal of the healthy tissues as possible. Early rehabilitation with massage and physiotherapy are also recommended

Conflict of interest: None



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