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# Craniofacial Cleft -Tessier 1-13: A unique enigmatic challenge

**Shweta Patil**[1]✉, **Anendd Jadhav**[2], **Apoorva Mishra**[3], **Nitin Bhola**[4]

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**Author Affiliation:**
[1]Postgraduate (PG) student, Department of Oral and Maxillofacial Surgery, Sharad Pawar Dental Colleg, Datta Meghe Institute of Medical Sciences (DMIMS) , Sawangi (M), Wardha, India; Email: shweta.patik06@gmail.com
[2]Associate Professor, Department of Oral and Maxillofacial Surgery, Sharad Pawar Dental College, DMIMS, Sawangi (M), Wardha, India; Email: anendd.j@gmail.com
[3]Assistant Professor, Department of Oral and Maxillofacial Surgery, Sharad Pawar Dental College, DMIMS, Sawangi (M), Wardha, India; Email: drmishraapoorva@gmail.com
[4]Professor and Head, Department of Oral and Maxillofacial Surgery, Sharad Pawar Dental College, DMIMS, Sawangi (M), Wardha, India; Email: drnitinbhola@gmail.com

✉**Corresponding author**
Postgraduate (PG) student, Department of Oral and Maxillofacial Surgery, Sharad Pawar Dental Colleg,
Datta Meghe Institute of Medical Sciences (DMIMS), Sawangi (M), Wardha, India;
Email: shweta.patik06@gmail.com

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## ABSTRACT

The exceptional cases of facial clefts are undoubtedly a surgical dogma to the surgeons. These cases bring the surgeon into a state of surgical muddle. The uniqueness of these defects demands a systematic multidisciplinary approach over a period of time. The identification of the type of cleft is critical in order to sketch out the treatment algorithm for that individual case. Therefore these extremely uncommon cases ought to be documented for further reference and enhancement of the literature available. To the best of our knowledge, this would be the sixth case of Tessier Cleft 1-13 to be documented.

**Keywords:** Tessier cleft 1-13, facial cleft, hypertelorism, encephalocele

## 1. INTRODUCTION

Craniofacial clefts are far less common than the most commonly treated cleft lip/palate deformities. Although considering its rarity, estimating a proper incidence or prevalence has not been possible, it is believed that the incidence lies in the range of 0.24% of all facial clefts (Gawrych et al., 2010; Eppley et al., 2005). Compared to conventional cleft lip and palate, the rate of occurrence of unusual craniofacial clefts can range from 9.5 to 34 per 1000 (Srinivas Reddy & Avni Acharya, 2021). According to WHO in 2002, proportionately clefts occur more commonly in Asian population. Cleft disorders are caused by a variety of circumstances, including inheritance, prenatal nutrition, drug exposure, and other environmental variables (Mohammad et al., 2021). There are three causative mechanisms of craniofacial clefts commonly mentioned in literature. The first is the disruption in the movement of cells of the neural crest. Second could be due the unsuccessful fusion of processes and the inability of mesoderm to develop inwardly and the third could be due to amniotic bands or genetics (Balaji, 2008; Carsten, 2017).

Craniofacial clefts project a complex anatomy with varying degrees of composite deficits. Hence, reconstruction is a challenge requiring multiple surgeries over a protracted period of time. Tessier originally brought attention to the link between hard and soft tissues in 1976, claiming that "a fissure of the soft tissue correlates, in general, with a cleft of the bony structure." He assigned a numerical numbering system from 0 to 14 to the orbit, based on its correlation to the 0 line (the midline cleft with vertical orientation).

The current case report aims to highlight the deformities associated with Type 1-13 of Tessier's classification of facial cleft and desired treatment

planning along with surgical intervention which will aid in overall development of the child.

## 2. CASE REPORT

The parents of a 2 year male child presented with a complaint of congenital nasal deformity. The mother gave history of deformity of external part of the nose since birth. Patient was conscious, co-operative and well-oriented. The general systemic and clinical examination was unremarkable. The examination of the face revealed an elongated facial structure with orbital hypertelorism (Figure 1).

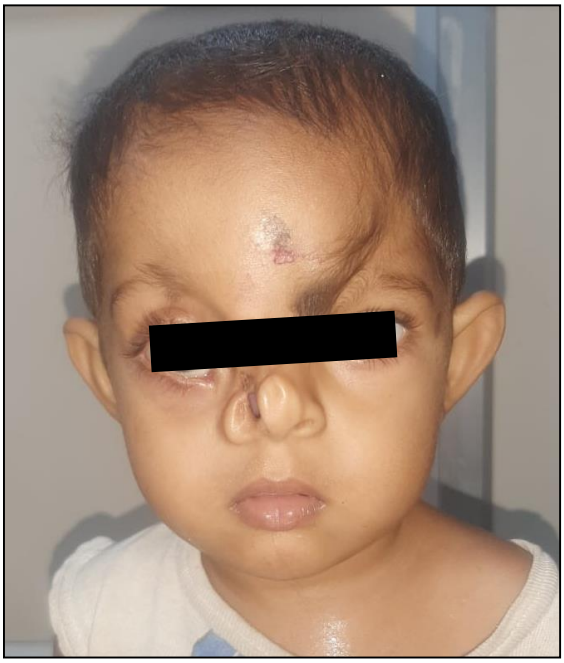

**Figure 1**Elongated face form with hypertelorism

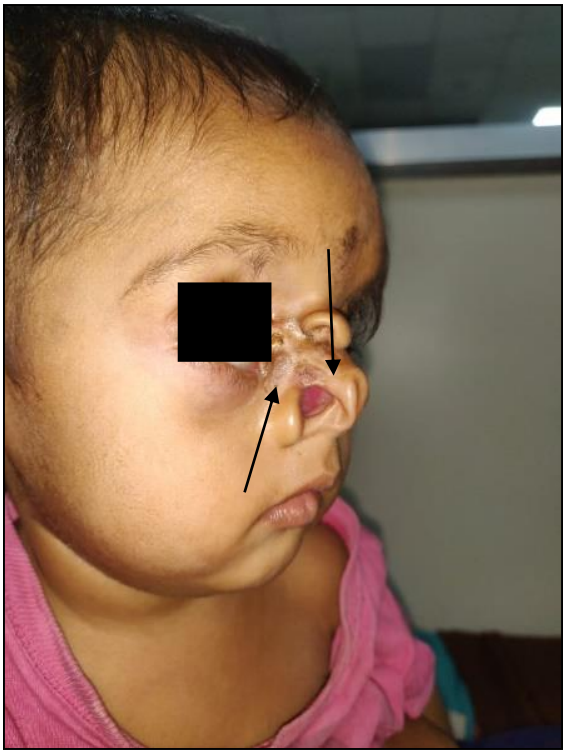

**Figure 2** Clefting of ala and coloboma of lower eyelid on right side

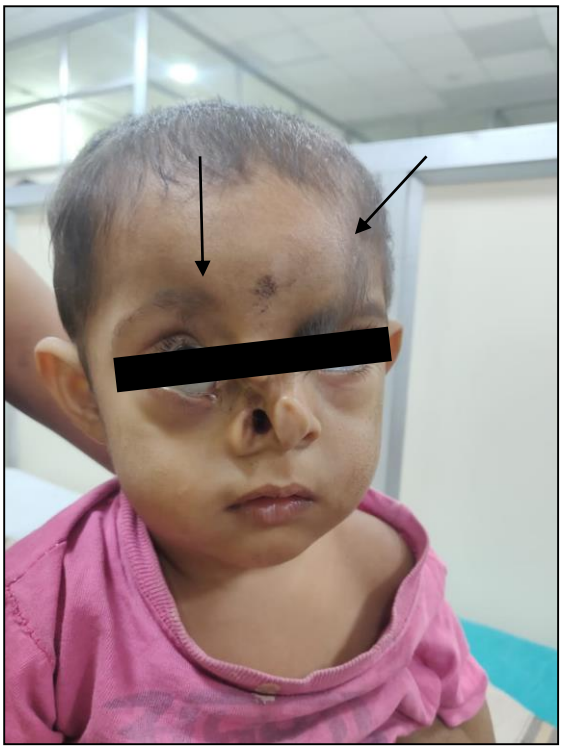

**Figure 3** Displacement of the uninterrupted medial brow on right side and para-median widow's peak on left side

The fronto-nasal angle was obtuse and clefting of ala of the right side was witnessed. It was associated with flattening of the nose's lateral aspect with a broad nasal bridge with coloboma over the lower eyelid of right side ( Figure 2). Medial canthus of the right eye was laterally displaced. A displaced uninterrupted eyebrow of right side was observed. The hairline presented with a paramedian widow's peak on the left side (Figure 3).

Ocular examination revealed visual acquity of perception of light with accurate projection in both the eyes. Mild vertical orbital dystopia was evident. Right eye showed mild divergent squint with right hypertropia and restricted ocular movements in the dextro-positions of gaze. The right nasal bone was separated from the external nasal dorsal surface and septal mucosa visible externally on the right side. In order to rule out brain herniation associated with the facial cleft, CECT was done which was suggestive of a Focal defect in the left basi-frontal bone with modest herniation of the surrounding brain parenchyma spreading to the fronto-ethmoidal recess on the right side till the level of soft tissue at the nose's root, suggesting Fronto-ethmoidal encephalocele. The axial cuts revealed a normal nasal septum and nasal cavity.

## 3. DISCUSSION

Incidence of craniofacial clefting approximates from 1.4 to 4.9 per 1,00,000 births (Hormozi et al., 2017). Infection and prenatal radiation, maternal drug intake, nutritional insufficiency, and metabolic derangement are all examples of environmental influences. Drugs like anticonvulsants, chemotherapeutics, corticosteroids and tranquilizers may act as etiological factors. The commonly used system for classification of facial clefts was propagated by Tessier in 1976. The clefts are enumerated from 0 to 14 with the orbital centre used as a reference point. A full evaluation of the patient must begin with a complete detailed history and physical examination, which must include an eye examination. The identification of deficient tissues and the tissues, use of which can be done for reconstruction of the defects is crucial. If reconstruction is to be considered, reconstruction of soft as well as bony tissues ought to be done. Soft tissue procedures are carried out during infancy while, hard structures shall be dealt and propelled until childhood.

The present case belongs to Tessier Cleft No. 1 continuing into the cranium as a Tessier No. 13 cleft. The Tessier cleft 1 is a cleft of the para-median region with vertical orientation along with soft-tissue notching that stretches from the nose's dome to the medial canthus as well as the medial eyebrow. Deformity of half of the nose is termed as Nasoschizis. A cleft of the lip can appear in the Cupid's bow area, which is where the "typical" cleft lip appears. The nasal defect might be anything from entirely absent upper and lower lateral cartilages to para-median soft-tissue fissures or nasal dorsum contour anomalies. The pyriform aperture, which is located lateral to the anterior nasal spine, may have a bone fissure between the central and lateral incisors. Hypertelorism is caused

by a fissure between the nasal bone (which may be notched or missing) and the frontal process of the maxilla. In extreme cases, hemi-nasal atrophy or a proboscis may be visible.

In this facial cleft, the nasolacrimal system is spared. Displacement of an uninterrupted medial brow is pathognomonic of Tessier cleft 13 (Raposo, 2021) (Figure 2). An encephalocele is a protrusive part emerging through a defect in the skull comprising of components of the cranial cavity. In a fronto-ethmoid encephalocele as present in this case, the cranial defect is located between the frontal bone and the ethmoid process in the anterior midline. This cranio-facial deformity leads to hypertelorism, orbital dystopia, elongation of face, and malocclusion (Hunt & Hobar, 2003). Tessier cleft 13 causes inferior displacement of the cribriform plate, passing through it, widens the olfactory groove. Increase in the transverse dimensions of the ethmoid may be seen. As mentioned in literature the primary goal is protection of the cornea/vision.

Patients with grotesquely malformed soft tissue might be treated in the 1st month of birth to regain a more typical look. Each part of the facial structure shall be evaluated and treated individually. Giventhat the majority of individuals with such abnormalities have lower-than-average facial growth potential, it is critical to wait until the entire development of the defect has occurred before beginning surgical treatment. Local flaps built following aesthetic lines are preferable in the restoration of such deformities. Multiple treatments performed over a long period of time are required to get maximum cosmetic outcomes. Osteotomies and bone transplants are used in skeletal reconstruction (autogenous bone grafts extracted from the calvaria rib or iliac crest are primarily preferred). Bone grafts should be cautiously used as they are susceptible to have effects on the growth potential eventually leading to resorption.

Significant hypoplasia has lesser pitfalls since their growth potential is antecedently hampered. Excess bone tissue between the orbits is decreased using paramedian wedge osteotomies to allow corrective rotation of the orbit, box and circular osteotomies are combined to release the orbits independently from the rest of the craniofacial skeleton in the treatment of hypertelorism. These units are rotated and medialized into the proper position, and plates, absorbable and non-absorbable, are used to attach them. An early and complete surgical treatment of the fronto-ethmoid encephalocele is critical to allow the developing brain and eyes to remodel the facial deformity (David, 1993). Based on surgical experience the goals of treatment of a fronto-ethmoid encephalocele have been laid in literature (Holmes et al., 2001).

## 4. CONCLUSION

The management of these defects surgically is challenging and requires a step by step repair. A proper sequence of the surgical procedures needs to be established for the desired outcomes before the intervention is initiated. There is no denial that a multidisciplinary approach is a key to success for such cases. Therefore it is evident that no specific treatment algorithm can be devised for such defects. Each case poses differently and desires an individualised and customised treatment approach.

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**Conflict of interests**

The authors declare that there are no conflicts of interests.

**Data and materials availability**

All data associated with this study are present in the paper.

## REFERENCES AND NOTES

1. Balaji SM. Modified Canthopexy. J Oral Maxil Surg 2008; 7:414-19.
2. Carstens MH. Pathologic anatomy of the soft palate, part 1: Embryology, the hard tissue platform, and evolution. J Cleft Lip Palate Craniofac Anomal 2017; 4(1):37. doi: 10.4103/jclpca.jclpca_9_17
3. David DJ. Cephaloceles: classification, pathology, and management--a review. J craniofac surg 1993; 4(4):192-202.
4. Eppley BL, van Aalst JA, Robey A, Havlik RJ, Sadove AM The spectrum of orofacial clefting. Plast Reconstr Surg 2005; 115:101-9 doi: 10.1097/01.PRS.0000164494.45986.91
5. Gawrych E, Janiszewska-Olszowska J, Chojnacka H. Tessier type 3 oblique facial cleft with a contralateral complete cleft lip and palate. Int J Oral Max Surg 2010; 39:1133-6. doi: 10.1016/j.ijom.2010.05.012.

6. Gosla Reddy S, Acharya AP. Rare Facial Clefts. Oral Maxillofac Surg Clinic 2021 (pp. 1735-1768). Springer, Singapore. doi: 10.1007/978-981-15-1346-6_77
7. Holmes AD, Meara JG, Kolker AR, Rosenfeld JV, Klug GL. Frontoethmoidal encephaloceles: reconstruction and refinements. J Craniofac Surg 2001; 12(1):6-18. doi: 10.1097/00001665-200101000-00003
8. Hunt JA, Hobar PC. Common craniofacial anomalies: facial clefts and encephaloceles. Plast Reconstr Surg 2003; 112(2):606-15 doi: 10.1097/01.PRS.0000070971.95846.9C
9. Kalantar-Hormozi A, Abbaszadeh-Kasbi A, Goravanchi F, Davai NR. Prevalence of rare craniofacial clefts. J Craniofac Surg 2017; 28(5):e467-70. doi: 10.1097/SCS.0000000000003771
10. Mohammadi M, Alavi A, Hovakhti A. The size of palatal cleft and surgical outcome in palatal cleft patients treated with and without feeding plate. Medical Science 2021;25(112):1381-1390
11. Raposo-Amaral CE, Oliveira YM, Denadai R, Ghizoni E, Raposo-Amaral CA. An Extended 45-year Long-term Follow-up on a 1–13 Rare Facial Cleft Patient. J Craniofac Surg 2021; 32(7):e638-42. doi: 10.1097/SCS.0000000000007648