Ophthalmic Considerations in the Management of Tessier Cleft 5/9

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Purpose: To describe a patient with Tessier cleft number 5 and 9 and review the literature on the ocular impairment and management of this extremely rare anomaly.

Methods: Interventional case report and literature review.

Results: The literature review showed that the present patient is the second case with clefts 5/9. The ophthalmic consequences of this rare association are virtually unreported. Our case demonstrates that the presence of cleft number 9 adds a cicatricial component on the upper eyelid that severely impairs the dynamics of this lid. The corneal status of the patient was successfully managed with simultaneous upper eyelid lengthening and facial reconstruction.

Conclusion: In order to avoid corneal perforation, simultaneous upper and lower eyelid reconstruction is mandatory in cases of cleft 5/9. The affected patients should be continuously followed in order to prevent amblyopia.

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Craniofacial clefts are extremely disfiguring and complex malformations that may involve several structures such as nose, mouth, cheeks, eyelids, orbits, and forehead. In 1976, Paul Tessier proposed an anatomic classification of the craniofacial clefts using the orbit as the primary structure of reference. Tessier[1] distinguished 15 types of defects. Clefts number 0 to 7 are found below the horizontal line that passes through the midorbit. Cleft number 8 is located at the lateral canthus, and clefts 9 to 14 radiate around and above the orbit. Some clefts are in the same vertical position and appear in pairs crossing the orbit and affecting both face and cranium. Typical examples are 0/14, 1/13, 2/12, 3/11, 4/10, and 5/9.

Facial clefts number 3, 4, and 5, also known as oblique or oro-ocular clefts,[2] involve the mouth and the inferior aspect of the orbit. Cleft 5 is the most rare in this group, and its association with a defect that extends above the orbital midline (cleft 9) is extremely rare. We are aware of just one description of the 5/9 association.[3] We report here a second patient who presented with Tessier cleft 5/9, review the literature on this

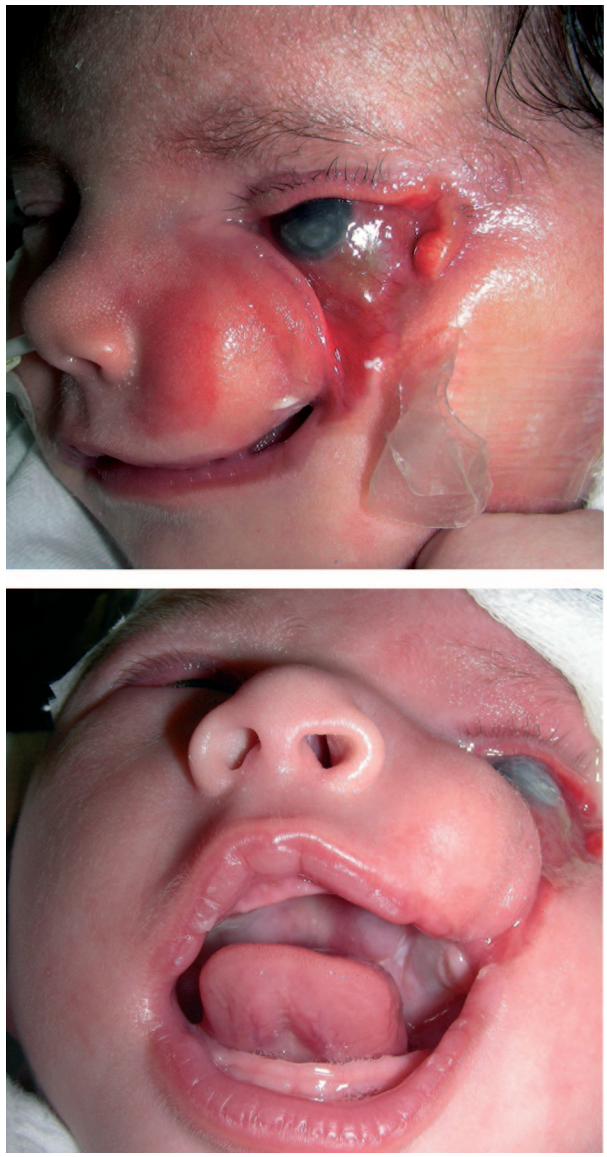

FIG. 1. Above, oblique view of the left side of the patient's face. Note the typical aspect of cleft 5, severe exposure keratitis. The upper eyelid contour is flat, and a furrow is present laterally interrupting the tarsal border near the lateral canthal tendon. Below, extension of the facial defect in the premolar area.

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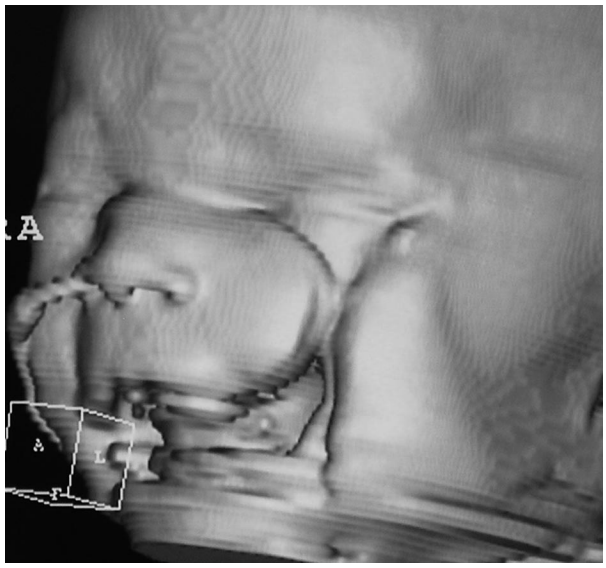


FIG. 2. Three-dimensional reconstruction of the clefts. The furrow involves the frontozygomatic suture.

rare association, and discuss the implications of lateral clefts that extend over the upper eyelid.

CASE REPORT

A 5-day-old female child, born by normal delivery at 37 weeks' gestation, presented with a disfiguring oblique oro-ocular cleft on the left side of her face. She was the product of an uneventful pregnancy with no exposure to any teratogenic agent. The parents were young, unrelated, and healthy. The defect extended from the left oral commissure, involved the middle and lateral thirds of the lower eyelid, and reached the upper eyelid and brow of the left eye. The upper crus of the lateral canthal tendon was probably not formed, and the upper eyelid was clearly disinserted and distorted upward. The upper eyelid contour was flat, and the amount of preseptal skin was reduced laterally. The upper eyelid margin was abnormally close to the eyebrow. Corneal perforation was imminent due to a large corneal ulcer (Fig. 1). CT of the face and orbit showed a skeleton cleft on the premolar region of the maxilla, extending to the orbital floor lateral to the infraorbital nerve and reaching the lateral orbital wall near the frontozygomatic suture (Fig. 2). The diagnosis of Tessier cleft 5/9 was made. No other physical anomalies were detected, and neurologic examination was according to age.

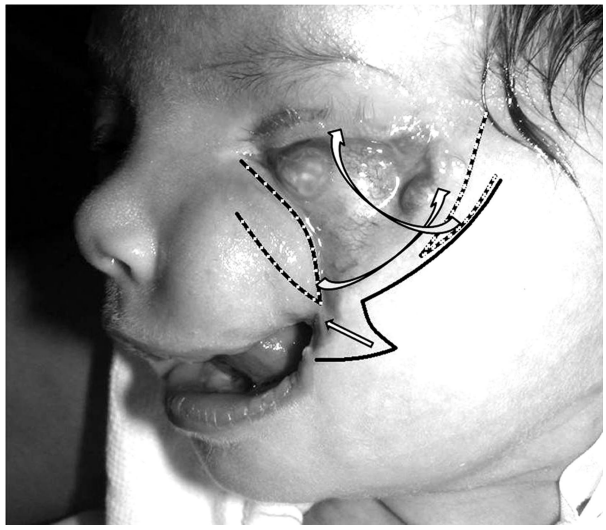
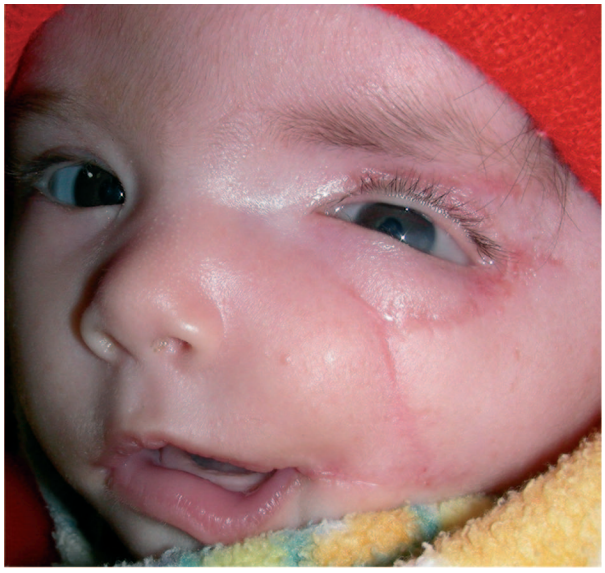


FIG. 3. Tissue mobilization for reconstruction. A superiorly based flap was brought to the lateral aspect of the upper eyelid.



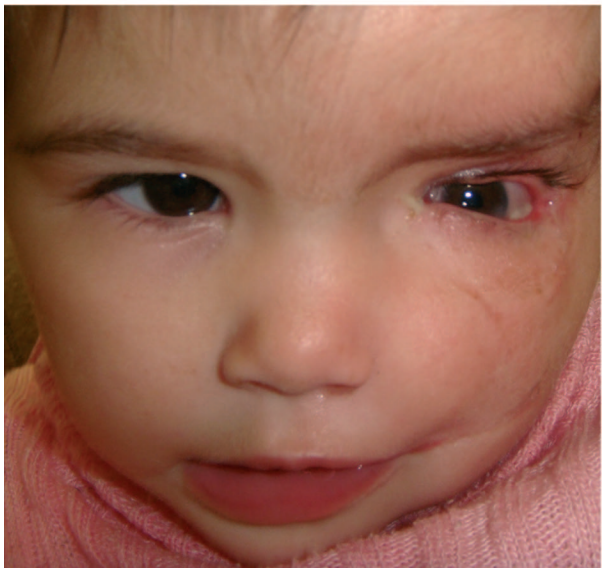


FIG. 4. Above, immediate postoperative result. The left cornea ulcer was healed, leaving a small area of corneal opacity. The left upper eyelid contour is normal. Note the eye motility limitation. The right eye is in adduction, but the left eye is fixed in primary position of gaze. Below, 6 months after surgery, the cornea is still clear, but there is a lateral upper eyelid flare, indicating a retraction that should be corrected. There is conjunctival discharge and the left eye is enophthalmic with motility limitation.

The soft tissue component of the craniofacial cleft was repaired at 11 days of age, with a combination of Z plasty and 2 rotational flaps. The lower eyelid was reconstructed

Demographic and clinical data of patients with clefts number 5, 9, and 5/9

Study	Case	Sex	Cleft	Side	Associated clefts/side	Ophthalmic findings associated to the clefts		
						Globe	Eyelid	Orbit
Tessier[1]	1	NS	5	Bilat	—	OD, microphthalmia; OS, anophthalmia	Lower eyelid coloboma OU	OD-Floor defect.
Kawamoto[5]	2	NS	5	R	7/Bilat	NS	NS	NS
David et al.[3]	3	F	5 + 9	R/9, L/9 + 5	4/R	OD, phitisis bulbi; OS, exposure keratitis	Soft tissue distortion: lateral third of upper eyelid, lateral canthus, lateral eyebrow	Absence of lateral wall OU
Tessier[16]	4	NS	9	Bilat	—	NS	Upper eyelid defect OU	Roof defect OD Bone formation upper lateral angle OS
David[7]	5	M	5	L	4/R	—	Lower eyelid defect	Floor defect
	6	M	5	Bilat	—	—	Lower eyelid defect OS	Floor defect OS
Galante & Dado[8]	7	NS	5	R	—	OD, hyperopia + astigmatism	Lower eyelid defect	Inferior rim defect
	8	F	5	L	—	OS, hyperopia + astigmatism	Lower eyelid defect	Inferior rim defect
Darzi & Chowdri[9]	9	F	9	L	3/L	OS, microphthalmia	Lateral upper eyelid and brow defect	Supero-lateral rim defect
	10	M	5	Bilat	—	—	Lower eyelid defect OU	Floor and rim defect OU
Menard et al.[10]	11	M	5	Bilateral	4/Bilat	NS	NS	NS
Dumortier et al.[17]	12	M	9	Bilat	—	Strabismus	Small superolateral defect OU	Hypoplasia of the superolateral orbital rim
Bilkay et al.[11]	13	F	5	Bilat	6, 1(R)	OS, anophthalmia	Lower eyelid defect OD	Inferior rim defect OU
Kara & Öçsel[12]	14	F	5	R	—	NS	Lower eyelid defect	Inferior rim defect
Sieg et al.[13]	15	M	5	Bilat	—	NS	Lower eyelid defect OU	NS
	16	M	5	Bilat	—	OS, severe keratitis loss of vision	Lower eyelid defect OU	NS
Morovic et al.[14]	17	F	5	Bilat	—	NS	OD, external canthal anomaly ″canthoschizis″	NS
Olasoji et al.[15]	18	M	5	L	—	OS, anophthalmia	Lower eyelid defect	NS
Al-Ani et al.[18]	19	M	9	R	—	OD, microphthalmia “coloboma” of iris	Distortion of the lateral brow	Right plagiocephaly, hypoplasia of the lateral orbital wall
Da Silva et al.[6]	20	F	5	R	4/L	NS	Lower eyelid defect	NS
	21	F	5	R	4/L	NS	NS	NS
	22	M	5	Bilat	3/L	NS	Lower position of lateral cathus OD lower eyelid defect OS	NS
	23	NS	5	L	—	NS	Coloboma of inferior eyelid with eyelashes extending over the cornea	NS
	24	M	5	Bilat	—	OD, anophthalmia	Lower eyelid defect OD	NS
	25	M	5	L	4/R	NS	Coloboma of lower eyelid	NS

M, male; F, female; R, right; L, left; Bilat, bilateral; NS, not stated.

with a medially based flap from the inner aspect of the cheek. The lateral remnants of the tarsal plate were preserved to form the lower eyelid margin. No conjunctival grafts were necessary to reform the lower fornix. At the same time, the upper eyelid was lengthened with a flap brought from the lateral aspect of the cheek (Fig. 3). The immediate result of the procedure was excellent (Fig. 4). The corneal ulcer healed, leaving a residual superficial corneal opacity. The upper eyelid contour was corrected, and the vertical dimension of the upper eyelid was close to normal.

Postoperative ophthalmic examination showed that the patient had a high degree of anisometropia (right eye, −plano; left eye, +7.0 sph), and her left eye motility was severely restricted in all gaze directions. To prevent left eye amblyopia, the fixating right eye was continuously patched. Six months after surgery, a significant contraction of the lateral third of the upper eyelid was apparent (Fig. 3, right). Although the cornea was still well lubricated, additional surgery was considered to further expand the lateral part of the upper eyelid.

DISCUSSION

Although a precise estimate of the occurrence of craniofacial clefts is difficult to establish, there is a consensus that the oblique clefts are the most rare with a prevalence of about 0.2% among all clefts.[4,5] It is also agreed that cleft number 5 is the least common of the oblique clefts. Cleft number 9 is even more rare than number 5. Tessier, in his classic article,[1] stated that he had never seen a number 9 cleft in the 336 patients he had examined. Some reports[6] quoting other authors mention 26 cases of cleft number 5 in the literature. We were able to find only 23 undisputed cases of number 5 cleft (30 eyes),[1,3,5–15] 5 cases of number 9 cleft (7 eyes),[3,9,16–18] and just 1 eye with the number 5/9 association (case 3 in the Table).

The aim of soft tissue repair in cleft number 5 is to simultaneously reconstruct the face and lower eyelid. In most cases, local tissue mobilization is enough to close the defect.[13] Nevertheless, the ophthalmic management of patients with cleft 5 is not restricted to eyelid reconstruction. Consequences of an orbital cleft for visual development have not received appropriate attention. In fact, no ophthalmologic information was available for 17 (56.7%) of 30 eyes with number 5 cleft listed in the Table. Even with this lack of documentation, it seems reasonable to assume that the degree of eye involvement will be dependent on the extension of the fissure in the orbit. Data compiled in the Table demonstrate that 6 eyes (20%) were severely impaired or blind, 2 were described as having refractive errors, and only 5 eyes were reported to be normal. It seems clear that just reconstructing the lower eyelid is not enough for the majority of cases.

For 2 of 7 orbits with number 9 cleft, no information was given on the degree of eye involvement. It is obviously difficult to draw conclusions on the effect of this cleft on the ocular globe. However, 2 eyes were reported to have strabismus and 3 were blind. The only eye with the 9/5 association was described as having a severe exposure keratitis.

Our case demonstrates that the combination of number 5 and 9 clefts within the same orbit induces complex problems for eye rehabilitation. Extension of the facial cleft in the orbit as a furrow on the floor may lead to enophthalmos and eye motility limitation. In this context, patching will be necessary to prevent amblyopia. Even if the globe seems to be normal, refraction is mandatory. Our patient had 7 diopters of hyperopia, emphasizing the necessity of optical correction and continuous visual assessment.

Presence of a lateral upper eyelid defect adds an important component that threatens the vision. Ocular surface protection and lubrication are highly dependent on the upper eyelid dynamics. If the blinking mechanism is impaired, severe corneal exposure ensues. In this context, the surgeon must direct all efforts at normalizing the upper eyelid structure. Local flaps and/or skin grafts should be used to lengthen the lateral aspect of the upper eyelid. In our case, the upper eyelid was satisfactorily reconstructed with a lateral based flap, which allowed a quick corneal recovery.

In summary, if the eye is not destroyed by the association of clefts number 5 and 9, ophthalmic management of this rare condition is dictated by the need of reconstructing both lower and upper eyelids and preventing amblyopia, which includes optical correction and patching. The reconstruction of the orbital floor may be delayed. Long life strabismus may be an unavoidable complication.

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