Title

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Journal

The Cleft Palate-Craniofacial Journal, 58(10)

ISSN

1055-6656

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Publication Date

2021-10-01

DOI

10.1177/1055665621989129

Clinical/Case Report

American Cleft Palate–Craniofacial Association

The Cleft Palate-Craniofacial Journal
1-5

DOI: 10.1177/1055665621989129
journals.sagepub.com/home/cpc

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Bilateral Craniofacial Cleft (Tessier Type 4): Case Report and Oculoplastic Reconstruction

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Abstract

Bilateral Tessier type 4 craniofacial clefts are extremely rare and disfiguring malformations with vision-threatening ramifications. To date, there is no consensus in the literature with respect to the ideal surgical technique and management of these patients. Emergent eyelid reconstruction and additional procedures may be required to protect the cornea and avoid further ophthalmic and surgical complications. We present our experience and challenges of managing a case of bilateral Tessier type 4 clefting with an emphasis on oculoplastic considerations.

Keywords

midfacial growth, palatal development, craniofacial morphology, surgical technique, surgical complications

Introduction

Craniofacial clefts are among the most rare facial clefts described in the literature with an incidence estimated to range from 1.43 to 4.85 per 100 000 births (Kawamoto, 1976). Bilateral cases are even rarer representing a small fraction of these cases. Tessier's nomenclature, introduced in 1976, uses the orbit as a point of reference and is the most widely accepted classification of craniofacial clefts. A Tessier type 4 cleft is an oculofacial cleft extending vertically from the lower lid lateral to the punctum, through the infraorbital rim and orbital floor (medial to the infraorbital nerve), and through the maxillary sinus and cheek. The cleft continues through the lip and through the alveolus to form a complete cleft palate (Tessier, 1976). The medial canthal tendon is usually intact, but with inferolateral displacement, and the nasolacrimal duct may also be malformed (Portier-Marret et al., 2008). Variable involvement of the globe may result in a functional eye, microphthalmia, or anophthalmia (Tsur et al., 1991; Tokioka et al., 2005). In patients with visual potential, exposure of the globe from frequently associated eyelid colobomas may result in blinding keratopathy.

These rare craniofacial clefts present a formidable challenge given the lack of clearly defined guidelines for surgical management and for the timing of procedures that would result in optimal functional and aesthetic outcomes (Mishra and Purwar, 2009). We present a case of bilateral Tessier type 4 clefting with severe lower eyelid deformity and sight-threatening keratopathy.

Case Report

A female infant, product of a normal pregnancy and karyotype, was born with incomplete bilateral Tessier type 4 oculofacial clefts and systemic anomalies including a left clubfoot, right renal agenesis, and cerebellar hypoplasia. The patient has no pertinent family or genetic history that predisposes her to this condition. Initial facial examination revealed bilateral cleft lip and palate, microcephaly, and soft tissue hypoplasia with shortened oculo-oral and oculo-alar distances. Ophthalmic examination demonstrated severe bilateral lower eyelid colobomas, involving approximately 60% of the right lower eyelid, and 40% of the left lower eyelid. There was near-total lagophthalmos and a poor Bell's phenomenon bilaterally. Fluorescein staining of both corneas revealed extensive epithelial breakdown consistent with severe exposure-related keratopathy.

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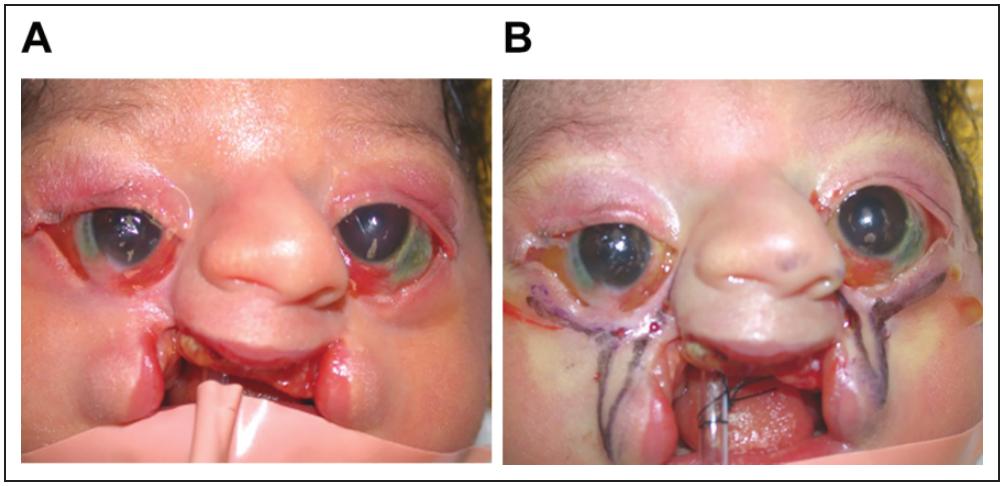

Figure 1. Initial examination photo demonstrating bilateral lower lid colobomas with globe exposure and bilateral cleft palate. External photo demonstrating appearance after modified Hughes procedure with superiorly based nasolabial transposition flaps. Note residual right lower lid retraction (A). Superiorly based nasolabial transpositional flaps were marked at the time of initial repair but abandoned in lieu of Tenzel semicircular rotational flaps (B).

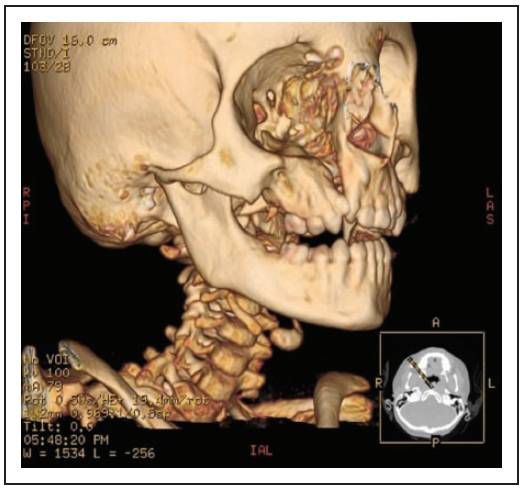

Figure 2. 3D reconstruction demonstrating extent of the orbitofacial cleft on the right. A similar appearance was noted on the left (not shown). Cleft begins between the lateral incisor and the canine and extends to the inferior orbital rim medial to the infraorbital foramen.

She was also noted to have right inferior conjunctival-cutaneous adhesions, proptotic globes, and bilateral scleral thinning (Figure 1). Imaging studies demonstrated bilateral clefting extending from the palate through the inferior orbit without inferior prolapse of the orbital contents. The visual axis, including both globes, the optic nerves, chiasm, and visual pathways, were unremarkable and suggested normal visual potential (Figure 2).

Procedures

Emergent surgery was performed to reduce corneal exposure at 3 days of age. The lower lids were reconstructed with Tenzel semicircular rotational flaps to recruit additional tissue from the lateral canthal region. Medial canthopexies were also performed to ensure appropriate eyelid-globe apposition by attaching the medial edge of the lower lids to the intact medial canthal tendons. Care was taken to protect the canthopexies from tractional forces by reinforcing the junction with horizontal mattress sutures placed over silicone bolsters (Figure 3). Despite initial improvement, within several weeks, she developed bilateral medial canthal dehiscence and subsequent corneal decompensation. She was treated conservatively with aggressive ocular lubrication for several months to allow for further facial growth before another repair was attempted. At age 5 months, the posterior lamella of both lower eyelids was reconstructed using Hughes tarsoconjunctival flaps, with concomitant secondary obstruction of the visual axis. The anterior lamella was augmented directly over the Hughes flaps using superiorly based nasolabial flaps transposed 90° laterally to increase the oculo-oral distance (Longaker et al., 1997) (Figure 4). Cleft lip repair, initially deferred to facilitate weight gain, was also performed at this time (cleft palate repair was performed uneventfully at 12 months). The Hughes flap was divided one week later to minimize the risk of occlusion amblyopia. Her lagophthalmos and exposure keratopathy improved postoperatively, despite persistent eyelid retraction, and she was maintained on frequent topical lubricants (Figure 5).

Despite close ophthalmic follow-up, at 9 months of age she presented with a 2-mm full thickness perforation of her left cornea. A left lamellar corneal patch graft and bilateral lateral tarsorrhaphies were emergently performed. Her corneas remained stable with chronic low-grade exposure after this intervention. At 4 years of age, a full thickness retroauricular skin graft was placed in her right lower lid to correct progressive lid retraction secondary to insufficient bony support and soft tissues adhesions to the orbital rim. Autogenous bony on-lay grafts to augment the orbitomalar region were discussed, but ultimately declined by the patient's family. The patient is currently 8 years old and continues to have a stable eye

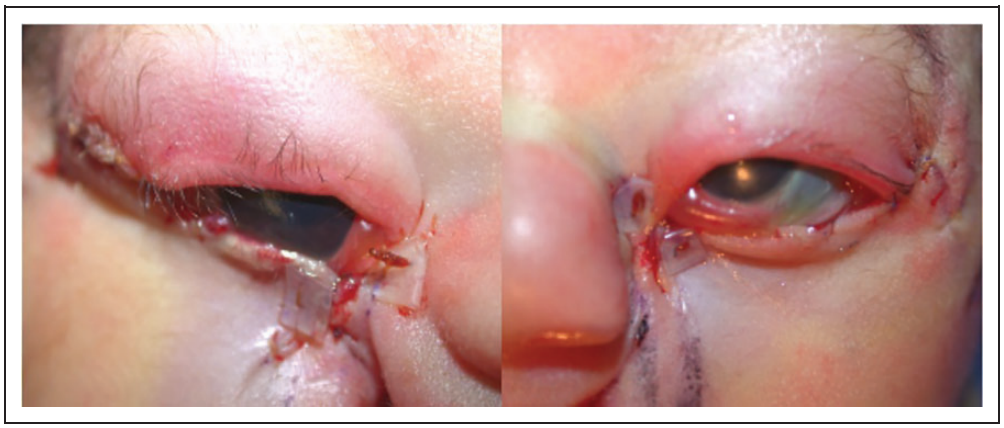

Figure 3. Immediate postoperative appearance after completion of bilateral Tenzel rotational flaps and medial canthopexies. Silicone bolsters are visible at the medial canthus bilaterally.

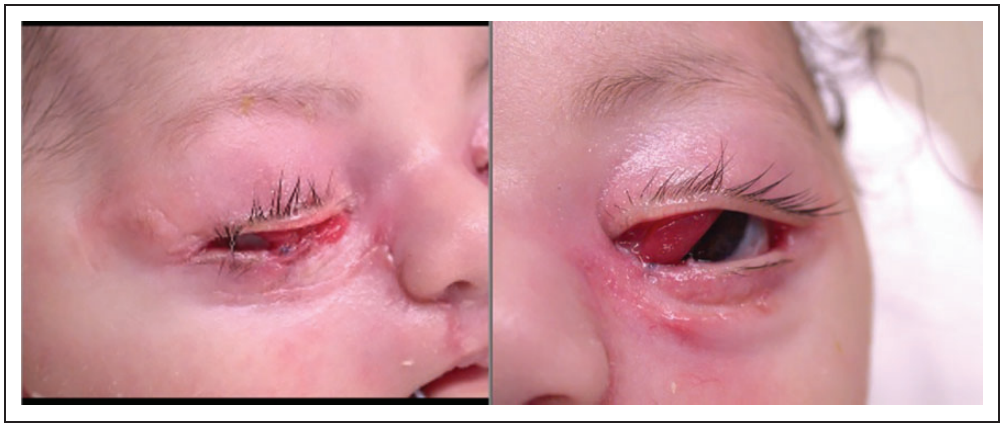

Figure 4. Postoperative appearance 1 week after Hughes tarsoconjunctival flaps and nasolabial transposition flaps.

examination, with manageable corneal exposure, and can fix and follow with each eye. A more comprehensive assessment of visual function cannot be performed given comorbid epilepsy, cerebral palsy, global developmental delay, and profound sensorineural hearing loss.

Discussion

Bilateral Tessier type 4 craniofacial clefts are an extremely rare and disfiguring malformation with few cases described in the literature. The etiology of this malformation remains unknown, though evidence suggests that amniotic band constriction with resultant disruption in tissue migration, ischemia, and necrosis may partially explain the development of orbitofacial clefts (Alonso et al., 2008; Abdollahifakhim et al., 2013).

Tessier type 4 clefts have been mostly reported to occur sporadically without any accompanying defects (Tsur et al., 1991; Alonso et al., 2008; Portier-Marret et al., 2008; Laure et al., 2010). Our patient, however, presented with multiple anomalies including right renal agenesis, a hypoplastic inferior cerebellum and vermis, and carried a diagnosis of cerebral palsy, epilepsy, and global developmental delay. Few cases in the literature have highlighted the co-occurrence of anomalies, such as central nervous system and renal defects in patients with unilateral or bilateral Tessier type 4 clefts (Tokioka et al., 2005).

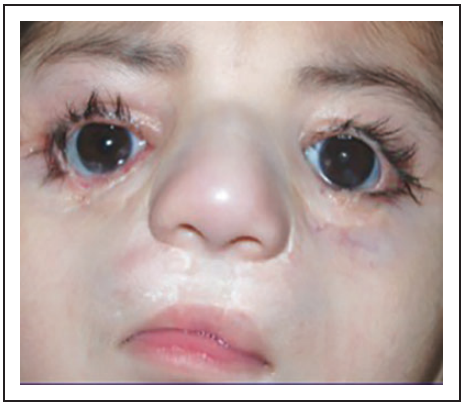

Figure 5. External photo demonstrating appearance 2 years after Hughes procedure with superiorly based nasolabial transposition flaps. Also note the presence of bilateral lateral tarsorrhaphies and residual eyelid retraction.

Due to its rarity and significant anatomic disruption, the surgical management of bilateral Tessier type 4 clefts remains challenging. To date, there are no clearly defined guidelines for

managing these cases beyond maximizing functional and aesthetic outcomes (Mishra and Purwar, 2009). The literature also lacks morphologic and anthropometric data that could provide useful reconstructive guidelines (Omodan et al., 2019). The advantages and shortcomings of various surgical methods have been extensively discussed in the literature. All parties agree that any associated eyelid colobomas resulting in corneal exposure require repair early in the neonatal period to prevent keratopathy and blindness (Sano et al., 1983; Longaker et al., 1997; Coruh and Gunay, 2005; Tokioka et al., 2005; Mishra and Purwar, 2009; Chen et al., 2012). According to Resnick and Kawamoto, the comprehensive surgical treatment of these complex cases includes medial canthopexy and lower eyelid reconstruction, cleft lip repair, bone grafting for orbital bony discontinuity and maxillary deformities, soft tissue reconstruction of cheek defects, tissue expansion of anophthalmic or microphthalmic orbits, and subsequent surgical revisions for bone and soft tissue deformities as needed (Resnick and Kawamoto, 1990; Coruh and Gunay, 2005; Chen et al., 2012). Numerous approaches to anterior lamellar repair in these complex cases have been described including the use of lip flaps, cheek flaps, z-plasty, and other techniques (Horoz et al., 2016). However, especially in cases with eyelid coloboma, posterior lamellar reconstruction is a critical and underemphasized step.

Even with aggressive early intervention, the visual consequences of orbitofacial clefting with comorbid eyelid deformities can be devastating. In our case, the need for urgent surgical repair to protect the eyes was recognized at birth. Our initial reconstructive approach prioritized, the minimization of general anesthesia exposure and the avoidance of any obstruction of the visual axis that could result in amblyopia. Bilateral Tenzel semicircular flaps with medial canthopexies were therefore chosen to reconstruct the lower eyelids at 3 days of age. The successful utilization of these flaps to repair lower eyelid defects involving up to 60% of the lid by recruiting lateral canthal soft tissue is well established (Tenzel and Stewart, 1978). Unlike Hughes tarsoconjunctival flaps, this approach offers adequate corneal protection in a single-staged procedure and without obstruction of the visual axis. Despite careful surgical technique, our patient unfortunately developed bilateral medial canthal dehiscence weeks after surgery, resulting in partial recurrence of the lower lid defects and corneal decompensation. We attribute this late dehiscence to increasing tractional forces on the canthus from rapid facial growth in infancy and overall soft tissue deficiency in the oculo-alar region. A second attempt at reconstruction was delayed until age 5 months to allow for further growth and expansion of the soft tissue envelope. It was performed using bilateral Hughes tarsoconjunctival flaps to recreate the posterior lamella, and superiorly hinged nasolabial transposition flaps to augment the anterior lamella. This composite reconstruction consisting of a tarsoconjunctival advancement flap and overlying nasolabial transposition flap, along with permanent lateral tarsorrhaphies, ultimately stabilized the ocular surface. To the best of our knowledge, this technique has not been described in the literature.

Hughes tarsoconjunctival flaps are flaps consisting of upper eyelid tarsus and a vascularized pedicle of upper eyelid conjunctiva that are transposed inferiorly across the surface of the globe to recreate the posterior lamella of the lower eyelid. The recruitment of upper eyelid tissue helps address the underlying periocular soft tissue deficiency seen in Tessier type 4 clefts. These flaps are traditionally left in place for 4 or more weeks to allow for vascular integration of the transposed tarsus and to counteract any downward cicatricial forces on the lower eyelid during healing (Rohrich and Zbar, 1999). Although it is unknown how long the visual axis of an infant can be occluded without developing amblyopia, 4 weeks was felt to be excessive in our case. In addition, several authors have reported successful division of Hughes flaps as early as 7 days without compromising flap viability or functional outcomes (McNab et al., 2001; Leibovitch and Selva, 2004). This was true in our case as our patient's Hughes flaps were divided after one week without incident. We submit, however, that maintaining a longer connection between the upper and lower lids may have further improved her eyelid retraction. Numerous authors have described modifying Hughes flaps to keep the visual axis clear, including "button-holing" the flap centrally. However, in our limited experience, the palpebral fissure remains too narrow to preserve visual function with these techniques (Hargiss, 1989; Leibsohn et al., 1993).

As described by Longaker et al., the use of nasolabial transpositional flaps in anterior lamellar eyelid reconstruction provides better support for the lid than free skin grafts and increases the oculo-alar distance. This results in improved cosmesis over other procedures (Longaker et al., 1997). This technique involves elevating an appropriately sized mycocutaneous flap along the nasolabial fold with a superior-based hinge and transposing the flap 90° laterally. In their original description, a subciliary incision was made along the horizontal length of the eyelid, followed by undermining of the incision. The nasolabial transposition flap was then used to "fill the gap," thereby lengthening the anterior lamella of the eyelid. In our case, the nasolabial transposition flap was placed directly on top of the Hughes tarsoconjunctival flap, creating a composite reconstruction that lengthened both the anterior and posterior lamella of the eyelid. Despite a dramatic improvement in eyelid position after this procedure, months of severe corneal exposure resulted in corneal thinning and ultimately perforation on the left side. After the placement of a corneal patch graft, along with permanent lateral tarsorrhaphies, the ocular surface stabilized and she continues to have functional vision in both eyes to date.

Thus far, our approach has achieved an acceptable functional and aesthetic outcome without addressing her midfacial skeletal insufficiency. Although we believe autogenous bony onlay grafting to her inferior orbital rims and malar regions may have improved her lower eyelid position, there is disagreement in the literature about the optimal timing for bone grafting in these cases (Longaker et al., 1997). Since bone resorption may occur over time and early manipulation of bony skeleton may disrupt normal midfacial development, some authors advocate for waiting until growth is near complete before performing bony augmentation (Coruh and Gunay, 2005; Tokioka

et al., 2005). Although she could still potentially benefit from bony augmentation, given the stability of her ocular surface and the overall complexity of her care, her family remains reluctant to pursue further intervention at this time. Additionally, her nasolacrimal dysgenesis remains unrepaired as epiphora may be protective in cases with chronic corneal exposure. Surgical repair could be considered in higher functioning patients bothered by epiphora, or those with chronic dacryocystitis.

As demonstrated by our case, the surgical management of these complex facial clefts with eyelid colobomas remains challenging and requires a multidisciplinary approach with staged procedures. Appropriate protection of the eyes is paramount in children with clear visual potential and requires early and aggressive intervention. Early eyelid reconstruction in these cases with the use of Hughes tarsoconjunctival flaps and overlying nasolabial transposition flaps adequately reconstructs the eyelid in a bilamellar fashion and addresses underlying oculo-alar soft tissue insufficiency. Repair of the nasolacrimal system may not be necessary in all cases. Rapid division of the Hughes flaps, along with the use of permanent lateral tarsorrhaphies, may further optimize this reconstructive approach. This technique may be used in conjunction with bone grafts and full thickness skin grafts and should be added to the reconstructive surgeon's repertoire.

Declaration of Conflicting Interests

The author(s) declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

Funding

The author(s) received no financial support for the research, authorship, and/or publication of this article.

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