Surgical treatment of upper eyelid coloboma
Our experience

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AIM: *The purpose of this paper is to evaluate the results obtained in the surgical treatment of upper eyelid coloboma with methods that do not involve a prolonged occlusion of the eye.*
MATERIAL OF STUDY: *We treated five patients aged between 7 months and 21 years; the surgical techniques adopted were the direct closure (2 patients), the full-thickness graft from the contralateral upper eyelid (2 patients) and the nasal chondromucosal flap (1 patient).*
RESULTS: *The results were good in all patients with vitality of all grafts and flaps; also absent were hematomas, seromas and infections. None of the patients developed amblyopia and lagophthalmos.*
DISCUSSION*: The use of these methods prevents the development of some complications, such as amblyopia, occurring with the use of other techniques, as shown in the literature. In addition, allowing the reconstruction with similar tissues or very flexible and thin tissues provides greater functionality to the new eyelid.*
CONCLUSIONS: *When possible, early intervention with the use of the described techniques will provide good results in the short and long term, allowing to avoid minimal complications which may arise from a prolonged occlusion of one eye.*

KEY WORDS: Coloboma, Eyelid, Surgical procedures

Introduction

Coloboma is a congenital malformation of the eyelids described for the first time in 1585 by Jacques Guillemeau. The etiology is unknown, characterized by the arrested development of a more or less extended segment of the eyelid. Coloboma appears as a full-thickness defect, single or multiple, unilateral or bilateral, upper or lower, isolated or associated with other ocular, facial or systemic malformations (Treacher Collins Franceschetti syndrome, Goldenhar syndrome). Isolated colobomas occur as a quadrangular defect, without corneopalpebral adhesions (cryptophthalmos), and leaves an intact eyelid margin (with eyelashes and tarsus). The lesions associated with other facial deformities often appear triangular with cryptophthalmos and the absence of lid margins to its sides; the cryptophthalmos is associated especially with colobomas of the upper eyelid, giving rise to specific syndromes such as Fraser syndrome or MOTA syndrome (Manitoba Oculotrichoanal syndrome) [1-6].
Several classifications have been proposed; Mustardé described three types of upper eyelid colobomas: isolated coloboma, coloboma associated with facial abnormalities and coloboma associated with epibulbar tumors. A more recent classification was suggested by Nouby, who

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divided coloboma into 5 degrees using an analysis of 26 patients:
Grade 1: coloboma without cryptophthalmos;
Grade 2: coloboma with abortive cryptophthalmos;
Grade 3: coloboma with complete cryptophthalmos;
Grade 4: classic cryptophthalmos (absence of all eyelid structures and complete coverage of eye by skin);
Grade 5: severe cryptophthalmos (with severe deformity of the nose and ectropion of the upper lip) [4].
Management of coloboma depends on the extent of the defect and associated malformations, but it is surgical in most cases. Several techniques have been proposed ranging from direct closure, use of the opposite eyelid or the contralateral one, to the use of nearby or distant tissues [2,6].
The aim of this paper is to evaluate the results obtained in the treatment of upper eyelid coloboma at the Department of Plastic and Reconstructive Surgery, University of Perugia, using techniques that do not require prolonged occlusion of the eye, such as direct closure, cutaneous-tarsal-conjunctival graft and the chondromucousal flap of the nose.

Materials and methods

This study evaluates the treatment of five patients with congenital coloboma of the upper eyelid who were treated from January 2006 to May 2011 at the Department of Plastic and Reconstructive Surgery, University of Perugia, complex structure of Perugia and Terni.
The patients were three males (60%) and two females (40%) aged between 7 months and 21 years, with an average of 6.1 years.
All five patients had a complete unilateral upper eyelid coloboma, four on the left and one on the right, localized in two cases in the lateral third and the other three in the middle third. All cases had an isolated coloboma, without further ocular or craniofacial deformities.
A careful evaluation of the coloboma was performed in all patients. Whenever possible, they underwent an eye examination that included an evaluation of visual acuity, presence of strabismus and state of the cornea. Finally, a thorough physical examination to assess the presence of associated systemic anomalies was performed.
The chosen treatment was surgical in all patients, immediately if severe corneal exposure or chronic exposure keratopathy were present or delayed if corneal lesions were absent. In case of small lesions and to prevent corneal damage, a conservative medical therapy was implemented in order to minimize any possible surgical or anesthesiological risks in younger patients.
The choice of surgical technique was made on the basis of the location and, especially, the extent of the coloboma. In small lesions (up to one quarter of the eyelid) a direct closure was preferred and in larger colobomas a cutaneous-tarsal-conjunctival graft from the contralateral eyelid or chondromucous flap from the lateral region of the nose (Scuderi flap) were used. All of the techniques that were used have been described in the literature.
During the follow-up, both functional (mobility and effectiveness in the coverage of the new eyelid) and aesthetical outcomes were evaluated.

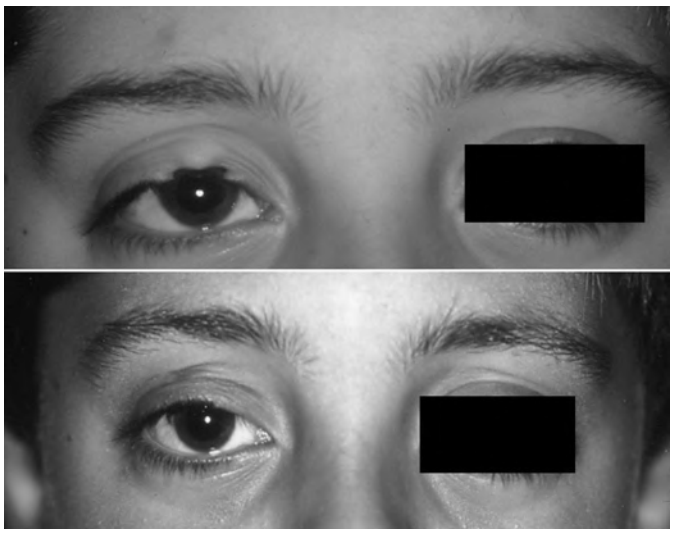

Fig. 1: Direct closure of the coloboma, before and after surgery.

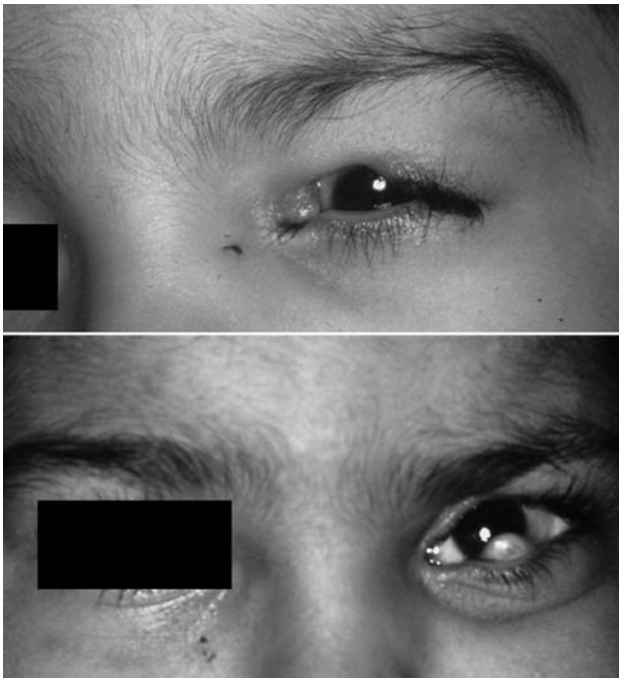

Fig. 2: Use of full-thickness graft from the contralateral eyelid, appearance before and after surgery.

Results

All five patients presented a complete coloboma of the upper eyelid, sizes ranged from less than one quarter of the eyelid (2 patients, 40%), to one third (2 patients, 40%) and up to a maximum of 2 / 3 of the eyelid (1 patient, 20%).
All colobomas were classified according to the grade 1 of Nouby's classification (coloboma without cryptophthalmos). Four patients had an adequate upper eyelid fornix with good eyelid closing and an absence of ocular abnormalities (strabismus and corneal lesions). Only one patient had a small fornix with partial corneal exposure, which resulted, due to unsuccessful treatment at a young age, in the onset of a progressive photophobia with initial keratopathy. None of the patients had concomitant systemic malformations.
In the two patients with mild coloboma (less than 25% of the eyelid length), direct suturing was performed; the edge of the defect was trimmed with the transformation of coloboma in a pentagonal loss of substance. The tarso-conjunctival layer was separated from the skin layer proceeding to the next 2-layer suture (Fig. 1).
Where the presence of coloboma extended up to 1/3 of the eyelid, the therapeutic choice was the use of a cutaneous-tarsal-conjunctival graft (full thickness graft) from the contralateral upper eyelid. The graft size was determined by the length of the coloboma under tension, with particular attention to allow the direct closure of the donor area. After trimming the edge of defect, the graft was transferred to the area lacking substance and sutured; a compression bandage was added to facilitate engraftment (Fig. 2).
In the adult patient with coloboma extending to two-thirds of the eyelid, reconstruction was performed with a chondromucosal flap harvested from the lateral side of the nose (Scuderi flap), associating a skin graft for cutaneous coverage.
The results were good in all patients; both the grafts and flaps were viable without signs of partial or total necrosis. There were no cases of hematoma, seroma or infection.
Normal length and good mobility of the reconstructed eyelid was achieved; during the follow-up, there were no cases of development of lagophthalmos. Time of follow-up was between 6 months and 24 months, with an average of 16 months.

Discussion

The management of congenital eyelid coloboma depends on the severity of the eyelid defect and the health of the corneal surface, which often reflects the location and the size of the defect itself. If the eye is fully exposed due to a large coloboma, it is essential that the reconstruction occur early to prevent corneal injury and subsequent vision loss. If there is exposure, surgery may be delayed until three to four years of age but not beyond. However, during this time period it is necessary to implement some measures to protect the eye itself, like ample lubrication or the use of protective lenses [7].
The surgical reconstruction techniques are determined by the size of the defect. For small defects, involving less than 20-25% of the eyelid, a primary closure can usually be performed with good results (Fig. 1). This technique is based on the Mustardè criteria that provide the possibility of a direct suture for loss of substance involving up to a maximum of one quarter of the length of the eyelid [8,9]. In this approach, it is necessary to transform the deformity in a pentagonal loss of substance and subsequent 2-layer suture. A lateral canthotomy and cantholysis of the lateral tendon may be necessary to minimize horizontal tension on the wound.
If the coloboma exceeds 25% of the eyelid, it is necessary to resort to more creative and technically difficult reconstructive techniques that are utilized in plastic surgery, ranging from the use of grafts to the use of flaps. In our experience, where loss of tissue extended up to about half of the eyelid surface, we preferred the use of full thickness grafts from the contralateral eyelid. This technique has proven very useful in giving good results; its use allows all structures of the eyelid to be replaced by a similar tissue, which present the same characteristics. Additionally, this technique does not cause a prolonged occlusion of the eye and requires only a one-stage reconstruction. The lack of an eye occlusion reduces the risk of developing iatrogenic sensory deprivation amblyopia. Consequently, these risks are present in other techniques such as the Cutler-Beard method, which requires tarsorrhaphy for at least three to four weeks as well as a two-stage reconstruction [10,11].
The main disadvantages to the use of this graft is the possibility of intervention only in unilateral form, forcing in the case of bilaterality to use more complex techniques such as Cutler-Beard or the muscle-cutaneous Tenzel flap. A further disadvantage, pointed out by other authors such as Hoyama et al., is the need to operate on an unaffected contralateral eyelid, as it is the donor site [10].
For large colobomas, more than 50% of the eyelid or bilateral lesions, other surgical procedures should be used: techniques that exploit the remaining eyelid, such as the modified Hughes procedure, the lower eyelid, such as the Cutler-Beard, the surrounding tissues, such as the Tenzel semicircular flap, or distant tissues [12].
Several authors have proposed using a graft of oral or nasal mucosa to reconstruct the conjunctival layer and a musculocutaneous flap for the reconstruction of the anterior lamella in order to minimize the trauma on other oculo-palpebral components [11]. This flap is usually taken from nearby areas like the eyebrow region, taking the skin and the superficial part of the orbicularis oculi muscle; this technique has been proven effective by providing a good opening of the eyelid, even though there

is an absence of tarsal (or cartilage) in the newly reconstructed eyelid [11,13].
In our experience, a Scuderi chondromucosal flap should be used. The procedure is derived from Micali's description of a full-thickness mucosal-chondrocutaneous flap harvested from the lateral side of the nose and then modified by Scuderi, who used only an ipsilateral axial chondromucousal flap associating a free skin graft for cutaneous coverage [14,15]. This technique provides a thin and pliable flap and does not require a long-term eye occlusion. Also, the presence of its own vascularization warrants needed support to the reconstructed eyelid. Finally, the use of a skin graft implies the absence of soft-tissue bulkiness, so that the eyelid can be easily lifted by the levator muscle of the upper eyelid. Scuderi et al. obtained similar results with the use of this flap for total or subtotal reconstruction of the eyelid in patients who underwent radical surgery for the presence of skin cancers [15].
The disadvantage of this technique is the possibility of its application only in the treatment of adult colobomas because the nasal donor area in the child is growing and important anatomical and functional deficits may result from its partial use.

Conclusions

The correct surgery choice should be directed by the severity of the malformation as well as the age at which to intervene. The larger defects should be treated early, while smaller ones may be delayed but to no later than four years of age.
In case of monolateral lesions, the surgical techniques preferred are either direct closure or the use of full-thickness grafts from the contralateral eyelid, as they provide good results with minimal complications, particularly those resulting from prolonged occlusion of the eye (such as amblyopia). In addition, reconstruction with similar tissue results in better functional and aesthetic outcomes. In the reconstruction of colobomas of the eyelid with large extended sections, the choice must also be conditioned on the age of the patient. In children, it becomes necessary that the recourse be to partially sacrifice the lower eyelid with a temporary occlusion of the eye; in adults, more results can be obtained with the nasal chondromucousal flap. The latter, like the previous grafts, avoids the total or partial sacrifice of the lower eyelid and the eye occlusion.
To conclude, according to our experience, we consider these techniques very valuable for the reconstruction of the eyelid colobomas in most of defects.

Riassunto

SCOPO: Lo scopo di questo articolo è quello di valutare i risultati che sono stati ottenuti nel trattamento chirurgico del coloboma palpebrale superiore con metodiche che non comportino occlusione prolungata dell'occhio.
MATERIALE E METODI: Sono stati trattati cinque pazienti con età compresa tra i 7 mesi ed i 21 anni; le tecniche chirurgiche adottate sono state la sutura diretta (2 pazienti), l'innesto cutaneo-tarso-congiuntivale (a tutto spessore) dalla palpebra controlaterale (2 pazienti) ed il lembo condro-mucoso della regione laterale del naso (1 paziente).
RISULTATI: I risultati ottenuti sono stati buoni in tutti i pazienti con vitalità di tutti gli innesti e lembi; assenti anche ematomi, sieromi ed infezioni. In nessun paziente si sono verificati casi di ambliopia e lagoftalmo.
DISCUSSIONE: Il ricorso a queste metodiche evita lo sviluppo di alcune complicanze, come l'ambliopia, che si manifestano con il ricorso ad altre tecniche, come si evince dalla letteratura. In aggiunta permettono la ricostruzione con tessuti analoghi o tessuti molto flessibili e sottili che garantiscono una maggior funzionalità alla neopalpebra.
CONCLUSIONI: L'intervento precoce, quando possibile, ed il ricorso alle tecniche descritte garantiscono buoni risultati sia a breve che a lungo termine, permettendo di ridurre al minimo possibile le complicanze che possono derivare dall'occlusione prolungata di un occhio.

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