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Cutler Beard Procedure for Bilateral Upper Eyelid Coloboma in Charge's Syndrom: Surgical Outcome

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ABSTRACT
Ocular coloboma is one of the anomalies that commonly seen in CHARGE syndrome. This syndrome is an autosomal dominant malformation which includes six major features: coloboma, heart defect, atresia of the choanae, retarded growth and development delay, genital hypoplasia, ear anomalies. Cutler-Beard procedure is one of the major technics when a total or near total upper eyelid is missing. To report the surgical outcome in bilateral upper eyelid coloboma in CHARGE syndrome patient after Cutler Beard's procedure. A 8 month-old boy presented to the hospital with bilateral upper eyelid coloboma since birth. Other clinical manifestations including bilateral optic nerve coloboma, external ear anomalies, bilateral undecensus testis, growth and developmental delay, and CHARGE facies which were consistent with 2 and 4 major and minor diagnostic criteria. The patient underwent eyelid reconstruction by using Cutler Beard procedure in both of eyes. Cutler Beard procedure may be used as the primary reconstruction method in bilateral upper eyelid coloboma in CHARGE syndrome.
Keywords. CHARGE syndrome; upper eyelid coloboma; Cutler Beard

INTRODUCTION

Ocular coloboma is one of the anomalies that commonly seen in CHARGE syndrome but also in can be appear in a variety of congenital syndromes (George et al., 2020). The mesodermal lid folds fusion failure become the cause of the upper eyelid coloboma (Jacobs et al., 2019). The patient commonly may present to ophthalmologists due to ocular coloboma (Lingam et al., 2021). It is important for the ophthalmologist to be familiar with CHARGE syndrome, because early management of associated defects is necessary (Thomas et al., 2021).

Eyelid reconstruction is essential. The technique will depend on the size of the defect and on the presence of corneal exposure (Alghoul et al., 2019). Direct tissue apposition was performed if the defect in the uppereyelid include less than 25% or one-third of the margin (Stewart & Burkat, 2022). If the small defects involving the upper eyelid margin and does not place too much tension on the wound, it can be repaired by primary closure (Yan et al., 2022).

Surgery can be delayed until 3-4 year-old, if the defect is small and there is no exposure of the cornea (Trief & Colby, 2017). A tenzel semicircular rotational flap can be used for moderately sized defects (25%-50%) including approximately one-third of the eyelid margin (Trigaux et al., 2023).

Hughes and Cutler Beard surgical procedures frequently used for bilateral upper eyelid colobomas with large defects (more than 50% of the eyelid) (Tenland et al., 2021). This reconstruction technique gives an excellent functional and cosmetic result (Wallace & Ashraf, 2021). Early treatment to prevent the complication that might be happen: corneal leukoma, symblepharon and amblyopia (Sharma et al., 2023).

Cutler beard procedure is a two-stage technique and an option frequently used for large upper eyelid defects. An incision is made below the lower eyelid tarsus, a full-thickness lower eyelid flap is moved into the upper eyelid defect by rise the flap behind the remaining lower eyelid margin. It results a temporary occlusion of the palpebral fissure so if the technique is used during early age, it may become a deprivation amblyopia (Eton et al., 2021).

In this case we report our experience in the surgical management and surgical outcome after cutler beard procedure in congenital upper eyelid coloboma in CHARGE syndrome. [3]

CASE REPORT

A 8 month-old boy was referred to the outpatient clinic with bilateral congenital upper eyelid coloboma. He has normal gestational age and delivery. Complete ophthalmology examination showed visual acuity of light perception in both of the eyes, large upper eyelid coloboma and symblepharon bilaterally (figure 1). The size of the right upper eyelid was 7 x 4 mm and in the lef upper eyelid was 8x 6 mm for the width and height. The medial part of the upper eyelid fused to more than half superior portion of the corneal surface in the right eye and slightly better in the left eye in which fusion only in perilimbal area of the corneal surface. Other anterior and posterior segment examination was difficult to perform in both eyes due to the palpebral-cornea attachment. Systemic investigation exhibit various congenital abnormalities such as hypospadias, square face with board prominent forehead, broad nasal bridge (figure 2), and external ear abnormalities.

The patient was consulted to pediatric department and diagnosed with multiple congenital anomalies with microcephalic, growth retardation and developmental delay without neurological deficit and cardiovascular malformation. Otolaryngology consultation found normal conduction hearing pathway, unfortunately sensory hearing pathway test was not performed due to poor compliance of the family.

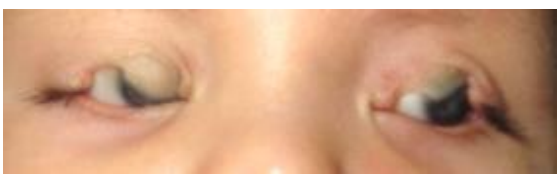

Figure 1: Bilateral upper eyelid coloboma with fused to more 50% corneal surface in the right eye and fusion in perilimbal area of the corneal surface in the left eye.

Figure 2: Distinctive facial feauters, square face with board prominent forehead, broad nasal bridge

Modified Cutler Beard procedure was decided to become the technique for the upper eyelid reconstruction, because the defect was large and bilaterally. From july 2018 until March 2019, The patient had four times surgical reconstruction, which was the first procedure in July on the upper eyelid coloboma in the left eye, the cutler beard sharing procedure in September 2018. The second reconstruction was in October 2018 in the right eye and sharing procedure was planned in December 2018, unfortunately because of the parents economic problems, the right eye sharing procedure delayed until March 2019 (figure 3). Routine post-operative follow up was made in two until three times after the surgery.

Figure 3: before reconstruction (1); after release cutler beard procedure in left eye (2); after release cutler beard in both eyes (3)

Currently, one month after the cutler beard procedure, the function of the both eyelid was examined and found 0,5 mm of lagophthalmos in the right eye and 1 mm in the left eye (figure 4). Half of the eyelash was lost in both of the eyes. Corneal surface hazy due to the upper eyelid fused. For the visual acuity only limited to fixation to the light with a right esotropia of 15^{o} was present in the right eye. In the left eye by using CSM (central –steady –maintain) we found the light reflex was seen in the center of the pupil, no nystagmus, holds steady fixation on the moving target and continues to stay fixated on the target. With the retinoscope value for the left eye was spherical +3.50 cylinder -1.50 dioptri Axis $100^{0.}$ Retinoscopy evaluation was not performed in the right eye due to the keratopathy was more than 50% of the corneal surface (figure 5).

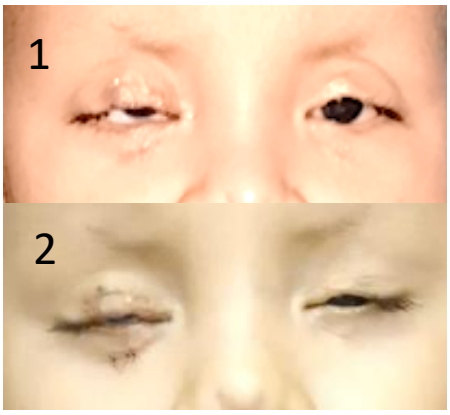

Figure 4. one month after complete cutler beard procedure in both of the eyes; front view with eyes open (1); front view when botheyes were closed while sleeping (2)

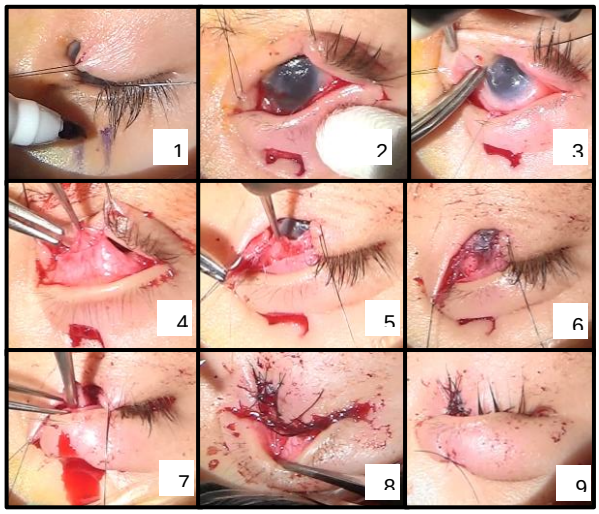

Figure 5. More than 1 half of the corneal surface has keratopathy in the right eye (1). Slightly hazy on the superolimbal of the corneal surface in the left eye (2)

DISCUSSION

Bilateral upper eyelid coloboma are uncommon congenital malformation caused by failure fused of the mesodermal lid folds in embryogenesis phase. It has become challenges for the surgeon because of the procedure can result in amblyopia. Some study reported the reconstruction may be delayed until age of 3 or 4 years (Perez et al., 2023). In these cases, the large defect was the reason for the early procedure due to prevent exposure of the corneal surface (Ahmed et al., 2015).

There are several procedures for bilateral upper eyelid coloboma with large defect. A study by Witmer et al. has reported a 2-month-old with bilateral upper eyelid colobomas and lateral modified Hughes procedure was selected to become the technique. Other case reported by Zhang et al. with a 5 day-old-newborn with a large upper eyelid coloboma were using internal cantholysis followed by direct closure (Zhang et al., 2017). Tenzel's semicircular flap, mustarde rotational flap technique for large upper eyelid coloboma were also reported in some cases (Francis et al., 2022).

In this case, cutler beard procedure was selected to become the technique (figure 6). The procedure was started with measure the size of the defect and make the lower eyelid flap. It was made 4 mm below eyelid margin. Seperating the anterior and posterior lamellae which were sutured separately to the levator muscle. Pull the superior side pass through the tarsus in upper eyelid until reached the defect. Release the attachment between upper eyelid and corneal surface. Make a suture between lower eyelid conjungtiva edge to edge with upper eyelid conjungtiva by using 8.0 absorbable vycril. Then sutured the skin by using 6.0 nonabsorbable. The left eye sharring procedure was made 8 months. The right eye reconstruction was made a month after the second procedure and the sharring procedure was made 5 months after that.

In this patient, the location of the tarsus was 2 mm below the eyelid margin and the bridge flap incision was made 4 mm below the eyelid margin. So the lower eyelid bridge flap didn't contain tarsus. The defect was bilaterally so it could not get the tarsal graft from the other eye and the surgeon also did not make a tarsal graft from the auricular cartilage. The failure of tarsoconjungtival flap or cartilage grafts have been reported by Fang et.al (2016).

After the sharring procedure, triamcinolone was injected subcutan in the upper eyelid. The purpose this injection was to inhibit the cicatrix formation so reduce the occurrence of lagophthalmos. Some study has reported the function of the triamcinolone injection to reduce scarring for concerns in cosmetic and functional (Huang et al., 2021).

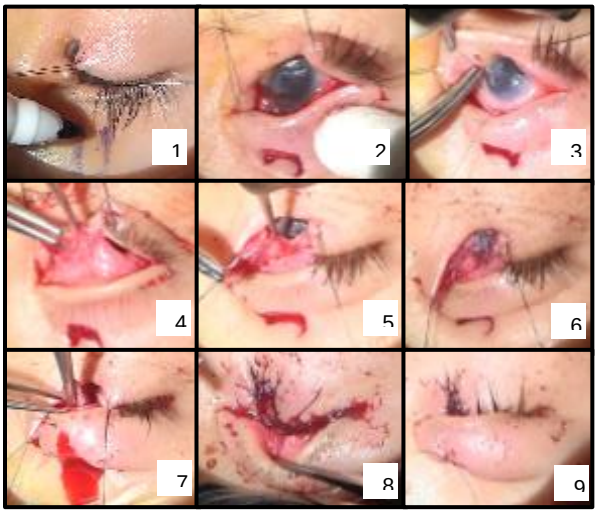

Figure. 6. The modified Cutler Beard procedure in the left eye. After measure the size of the upper eyelid defect, a mark was made in the lower eyelid to estimate the incision size (1). Full thickness lower eyelid flap was made (2). Release the coloboma with a fused corneal surface (3). A lower eyelid flap was divided into posterior and anterior lamina (4). Suture by edge to edge to the conjugngtival upper eyelid (5). Conjungtival flap (6). Closure of the anterior lamina (7). The bridge flap appearance from inside (8). The bridge flap appearance from outside (9).

Follow up after surgery carried out for one week to assess the stability and functional of the eyelids. High risk of amblyopia is the biggest problem for the cutler beard procedure in early age due to a long-term occlusion. In this case, there were significant delay sharring procedure time in the right eye which is 5 months and 8 weeks in the left eye. The normally sharing procedure is made 4 - 6 weeks after the first procedure. This delay can increase the risk of amblyopia due to the long term of occlusion of visual axis. We couldn't measure the visual acuity for the right eye due to the keratopathy, but for the left eye by using the CSM and retinoscope test we did not find any problem. Study by Lodhi et al. also had a same result by using the cutler procedure in congenital bilateral upper eyelid coloboma where they did not found such problem after sharring procedure (Beri et al., 2022).

Lagophthalmos was found in both of the eyes after the surgery. 0.5 mm in the right eye and 1 mm in the left eye. It was happened because of absence of the tarsal graft. No corneal exposure due to good bells phenomenon. This also the reason why the secondary tarsal implantation was postponed.

CONCLUSION

Modified Cutler Beard procedure can become the technique for bilateral uppereyelid coloboma in CHARGE syndrome. It gives a good result in anatomical, functional and cosmetic. Visual acuity examination showed no amblyopia in the left eye.

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