Prognosis of Upper Eyelid Epiblepharon Repair in Down Syndrome

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• OBJECTIVE: To evaluate the recurrence rate after upper eyelid epiblepharon repair in patients with Down syndrome.

• DESIGN: Retrospective, observational study.

• METHODS: Total of 578 Korean children (21 with Down syndrome patients, 557 with non-Down syndrome patients), who had undergone epiblepharon repair and were followed up for more than 2 months, were included in this study. The recurrence rate was compared between two groups at 2, 6 months after surgery. Recurrence was defined as the re-appearance of cilia touching to cornea. The recurrence rate was also analyzed according to whether patients had undergone concomitant z-medial epicanthoplasty or not.

• RESULTS: Lower eyelid epiblepharon repair was performed on 22 eyelids of Down syndrome patients, and 1072 eyelids of non-Down syndrome patients. At 3 months after surgery, the recurrence rate was not significantly different between two groups ($P = 1.00$). Upper eyelid epiblepharon was repaired on 40 eyelids of Down syndrome patients, and 204 eyelids in non-Down syndrome patients. At 2 and 6 months after surgery, the recurrence rate was significantly higher in Down syndrome patients (27.5% and 29.4%) than non-Down syndrome patients (3.4% and 4.6%) ($P = 0.000$, $P = 0.004$, respectively). The recurrence rate of upper eyelid epiblepharon repair was not affected in both groups whether Z-epicanthoplasty was combined or not ($P = 1.00$ in both groups).

• CONCLUSIONS: In Down syndrome patients, the recurrence rate after upper eyelid epiblepharon repair was higher than non-Down syndrome patients. The effect of combined Z-medial epicanthoplasty was limited in both groups. (Am J Ophthalmol 2010;150:476–480. © 2010 by Elsevier Inc. All rights reserved.)

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EPIBLEPHARON IS RATHER COMMON CONDITION AMONG Asian children[1,2]; in this condition, a fold of skin and the underlying orbicularis muscle press the lashes against the eyeball. In general, the lower eyelids are commonly involved as compared to the upper eyelids.[1,2] Recurrence rates after surgical correction are considerably low,[2–5] and recent techniques of correction include cilia rotating tarsal fixation suture[6] and Z-medial epicanthoplasty.[7] However, the incidence and clinical manifestations of epiblepharon that occurs in Down syndrome patients remarkably differ from those of epiblepharon that occurs in non-Down syndrome Asian children.[8] The incidence of epiblepharon is much higher in Down syndrome patients than in non-Down syndrome patients, and characteristically, the upper eyelids are predominantly involved in the case of Down syndrome patients. Moreover, natural remission of the upper and lower eyelid epiblepharon rarely occurs in the case of Down syndrome patients; this is in contrast to that observed in the case of non-Down syndrome patients.[8] Therefore, almost all Down syndrome patients with epiblepharon need surgical correction. To our best knowledge, surgical success rate of epiblepharon repair in the case of Down syndrome patients has not been reported. In this study, we evaluated and compared the surgical results of epiblepharon repair in the case of Asian Down syndrome patients and in that of non-Down syndrome patients.

METHODS

THIS STUDY WAS A RETROSPECTIVE OBSERVATIONAL study. The following patients were included: those who had undergone upper eyelid epiblepharon repair performed by 2 of the authors (H.K.C & N.J.K) at Seoul National University Hospital, Seoul Metropolitan Government Seoul National University Boramae Medical Center, Seoul National University Bundang Hospital, between June 2003 and July 2008. Epiblepharon was diagnosed when cilia touched cornea with horizontal fold of redundant skin and the underlying pretarsal orbicularis oculi muscle[2] without inward rotation of eyelid margin. Surgical indications were severe corneal erosion, i.e., horizontal erosion, in more than one-third of the cornea or irritative symptoms such as foreign body sensation, ocular pain, or tearing that the parents thought were remarkable even after the

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TABLE. Types of Epicanthal Fold[10]

Type	Definition
Type I	No epicanthal fold.
Type II	The upper eyelid skin margin covers the tarsal border as it approaches the medial canthal ligament, and the lacrimal lake is covered only partially.
Type III	The upper eyelid skin margin curves over the lacrimal lake, covering the entire medial angle of the palpebral fissure.
Type IV	The lower eyelid skin crosses over the lacrimal lake, forming a reverse epicanthal fold.

affected children were treated with artificial tears. The following patients were excluded: those who were not followed up for more than 2 months; those who had any other disease, in which affected the eyelid position is affected, such as Stevens-Johnson syndrome; and those with ocular cicatricial pemphigoid or facial trauma. For cosmetic reasons, epiblepharon repair was performed on both the right and left sides for all the patients including those with epiblepharon on only 1 side. Lower eyelid epiblepharon was repaired using cilia rotating tarsal fixation suture and excision of the redundant skin and the pretarsal orbicularis muscle.[9] Upper eyelid epiblepharon was repaired by an incisional double-eyelid surgery with tarsal fixation suture. We performed a combined Z-medial epicanthoplasty on patients with a prominent epicanthal fold by using modified Park's Z-epicanthoplasty.[10] We used the classification system described by Park[10] to evaluate the severity of the epicanthal fold; further, this procedure was indicated in patients with epicanthal fold type II and type III (Table). At follow-up, the patients were evaluated for direction of the lashes and status of the cornea; recurrence was defined when the cilia touched the cornea again at each time for each eyelid. We compared the recurrence rate between the Down syndrome and non-Down syndrome patients. The recurrence rate between the 2 groups was also analyzed in both groups according to whether the patients had undergone concomitant Z-medial epicanthoplasty. In the case of simultaneous upper and lower epiblepharon repair, each eyelid was independently analyzed, because we think that the upper eyelid epiblepharon repair does not affect the lower eyelid epiblepharon repair and vice versa. Statistical analyses were performed using Statistical Package for Social Sciences (SPSS) software v. 13.0 (SPSS Inc, Chicago, IL), and two-sided $P < 0.05$ was considered statistically significant.

RESULTS

THIS STUDY INCLUDED 811 KOREAN PATIENTS WHO UNDERwent epiblepharon repair during the study period; of these, 233 patients who were not followed up for more than 2 months were excluded. Thus, 578 patients fulfilled the inclusion criteria; of these, 21 were Down syndrome patients, and 557, non-Down syndrome patients. The clinical characteristics of the patients were as follows: male versus female ratio was 10:11 among the Down syndrome patients, and 274:283, among the non-Down syndrome patients ($P = 0.887$, chi-square test); the mean age of the Down syndrome patients at operation was 7.33 ± 2.65 years and that of the non-Down syndrome patients was 6.19 ± 3.87 years ($P = 0.179$, independent T-test); further, the mean follow-up period for the Down syndrome patients was 12.5 ± 12.9 months and that for the non-Down syndrome patients was 8.6 ± 9.7 months ($P = 0.17$, independent T-test). The follow-up period of the non-Down syndrome patients was shorter than that of the Down syndrome patients although the difference was not significant.

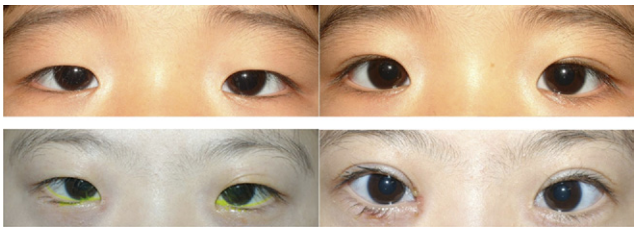

FIGURE 1. Preoperative and postoperative photographs of Down syndrome and non-Down syndrome patients. (Top) Upper and lower eyelid epiblepharon repair in non-Down syndrome patient. (Top left) In this patient, vertical cilia touching the cornea can be seen before repair. (Top right) The eyelid crease is present and no cilia can be seen touching the cornea more than 7 months after the surgery. (Bottom) Upper and lower eyelid epiblepharon repair with Z-medial epicanthoplasty in Down syndrome patient. (Bottom left) In this patient, severe epiblepharon in both the upper and lower eyelids along with the epicanthal fold can be seen before repair. (Bottom right) Photograph obtained 22 months after surgery showing well-everted cilia and cosmetic enhancement.

In lower eyelid epiblepharon repair, 1 of 22 eyelids recurred (4.8%) in Down syndrome patients, and 59 of 1072 eyelids recurred (5.5%) in non-Down syndrome patients by the two month postoperative visit. ($P = 1.000$, Fisher's exact test). However, the recurrence rate did not significantly differ between the 2 groups.

Upper eyelid epiblepharon repair was performed on 20 Down syndrome patients (40 upper eyelids) and 102 non-Down syndrome patients (204 eyelids; Figure 1). In upper eyelid epiblepharon repair, 11 of 40 eyelids recurred (27.5%) in Down syndrome patients, whereas 7 of 206 eyelids recurred (3.4%) in non-Down syndrome patients by the two month postoperative visit ($P = 0.000$, chi-square) and 5 of 24 eyelids recurred (20.8%) versus 7 of 146 eyelids recurred (4.8%) by the six month postoperative visit ($P = 0.004$, chi-square test; Figure 2). Kaplan–Meier survival curves for the recurrence of epiblepharon

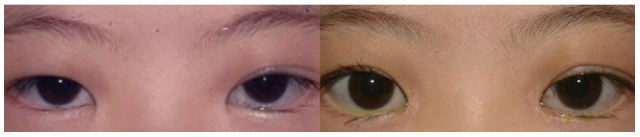

FIGURE 2. Photographs showing a recurrent case of epiblepharon in a Down syndrome patient. (Left) At the sixth postoperative month, a recurrence of epiblepharon involving the medial side of the left upper eyelid epiblepharon was observed. (Right) Compare the corrected right upper eyelid with the left upper eyelid.

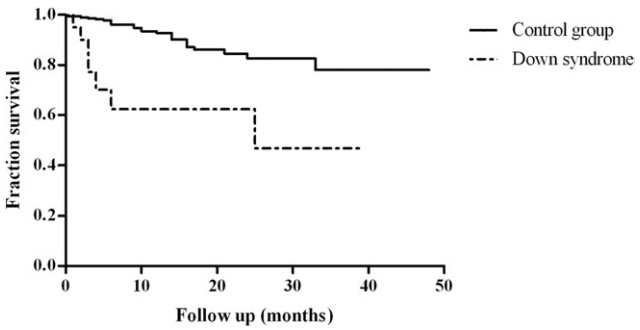

FIGURE 3. Graph showing an epiblepharon repair survival curve. Cumulative survival plot (Kaplan–Meier) for the Down syndrome and non-Down syndrome patients who underwent upper eyelid epiblepharon repair. Event was defined as epiblepharon recurrence. A large number of recurrences occurred in the early postoperative period in the case of Down syndrome patients, in contrast to the fewer number of recurrences in non-Down syndrome patients.

are shown in Figure 3. The cumulative probability of survival was significantly lower among the Down syndrome patients than among the non-Down syndrome patients (Log Rank $P = 0.000$, Breslow $P = 0.000$).

Z-medial epicanthoplasty was combined with upper eyelid epiblepharon repair in 17 of 20 Down syndrome patients, and 6 of 102 non-Down syndrome patients (Figure 4). In both groups, the recurrence rate was not significantly different according to whether patients had undergone concomitant Z-medial epicanthoplasty or not; In the Down syndrome patients, recurrence was observed in 10 of 34 eyelids (29.4%) after upper eyelid epiblepharon repair combined with Z-medial epicanthoplasty and in 1 of 6 eyelids (16.7%) after upper eyelid epiblepharon repair without Z-medial epicanthoplasty ($P = 1.00$, Fisher's exact test). In the non-Down syndrome patients, recurrence was not observed in any of the eyelids when upper eyelid epiblepharon repair was combined with Z-medial epicanthoplasty; however, recurrence was observed in 7 of 192 eyelids (3.0%) when only upper eyelid epiblepharon repair was performed ($P = 1.00$, Fisher's exact test).

DISCUSSION

EPIBLEPHARON IS A COMMON ANOMALY OF THE EYELID among Asians and most commonly involves the medial side of the lower eyelids.[1] This condition is considered to be a congenital disorder in which a horizontal fold of redundant skin and the underlying pretarsal orbicularis oculi muscle tilt the eyelashes and press them against the globe. Entropion is characterized by an inward rotation of the entire eyelid and lashes; in contrast, the eyelid position is normal in the case of epiblepharon. Recurrence after epiblepharon repair has decreased from 23%[3] to 3.6%[6] with advancements in surgical techniques like cilia rotating tarsal fixation suture[6] and Z-medial epicanthoplasty.[7] But, these data were based on general population studies involving patients with mainly lower eyelid epiblepharon. Although we were unable to find the previous study that showed the protective effect of Z-medial epicanthoplasty, some authors believe that concomitant medial epicanthoplasty would enhance the success rate of epiblepharon repair,[7] because epicanthoplasty would enhance the rotational stability.

The incidence of epiblepharon is remarkably high among Korean Down syndrome patients. Previous studies focused on the presence of the epicanthal fold that presses the cilia of the upper eyelid to the cornea.[8] The unique characteristics of epiblepharon that occurs in Down syndrome patients, such as predominance in the upper eyelids and frequent association with an epicanthal fold, make the situation different. Rare spontaneous regression in Down syndrome patients with epiblepharon could be partially explained by these facts.[8]

In this study, we separately evaluated the upper and lower eyelids and upper eyelid predominance was also detected. In the control group, the end recurrence rate was 3.4–5.3% in the upper and lower eyelids throughout the follow up period. These rates were similar to those previously reported.[6] In addition, in the Down syndrome patients, the recurrence rate in the lower eyelids was 4.8%, however it was 20.8–28.1% in the upper eyelids. The recurrence rate at both 2 and 6 months after operation was higher in the Down syndrome patients than in the non-Down syndrome patients. The survival curves showed differences between the 2 groups for the entire follow-up period (Figure 3). However, in contrast to non-Down syndrome patients who showed low recurrence rate, a larger portion of recurrences occurred early (less than 6 months) in Down syndrome patients. Thus, the early postoperative recurrence in the Down syndrome patients suggests that the conventional incisional double-eyelid techniques with tarsal fixation might not be suitable for a large subgroup of Down syndrome patients. Hopefully, advancements in surgical technique will improve results for treatment of upper eyelid epiblepharon in Down syndrome patients. However, there have been no reports of

FIGURE 4. Photographs showing the detailed surgical procedure of upper eyelid epiblepharon repair with medial Z-epicanthoplasty. (Top left) Markings for skin incision. The position of the eyelid crease usually was indicated at 5 to 6 mm from the cilia line, and markings for Z-epicanthoplasty also were traced. (Top right) After skin incision, an epicanthal flap was developed. The flap was freed cautiously from the surrounding fibrous soft tissues and the medial canthal tendon and was rotated. (Bottom left) Dissection was performed between the orbicularis muscle and the tarsus to expose the anterior surface of the tarsal plate, and the hypertrophic pretarsal orbicularis muscle beneath the lower edge of the skin incision was excised. (Bottom right) After fixation sutures between the tarsal plate and lower skin edge were applied, skin incisions were closed.

fascinating surgical technique for Down syndrome with epiblepharon so far.

Prominent epicanthal folds (more than type 2)[10] which indicate the need for a surgical correction, were found much more frequently observed among the Down syndrome than among the non-Down syndrome patients. Therefore, a significantly higher number of the Down syndrome patients (81% of the Down syndrome patients vs 2% of the non-Down syndrome patients) needed concomitant Z-medial epicanthoplasty with upper eyelid epiblepharon repair. In the case of epiblepharon, the prominent epicanthal fold may play an additional role by pressing the cilia of upper eyelid to the cornea.[7] Similarly, Z-medial epicanthoplasty might decrease the recurrence rate by enhancing the rotational stability.[6] Therefore, it is reasonable to state that higher Z-medial epicanthoplasty rates would mask the real difference between 2 groups of patients. In the case of the concomitant upper eyelid epiblepharon repair and Z-medial epicanthoplasty, the recurrence rate in the Down syndrome patients was 29.4–30.0%, whereas recurrence was not observed in the non-Down syndrome patients. While patients with Down syndrome seem to have a higher failure rate after Z-medial canthoplasty for epicanthal folds, patients with Down syndrome have a higher incidence of epicanthal folds in the setting of epiblepharon. Our results support that Down syndrome patients may have a poor surgical prognosis for success after epicanthoplasty. This may imply nuanced differences in the etiology of the eyelid malposition in patients with Down syndrome. Conventional Z-medial epicanthoplasty may fail to provide enough rotational stability in Down syndrome patients. Further studies and analysis according to the severity grading of the epicanthal fold[10] may define the effect of epicanthoplasty more precisely.

Recurrence was not observed in any of the non-Down syndrome patients who underwent concomitant Z-medial epicanthoplasty; however, the recurrence rates with or without concomitant epicanthoplasty in the case of the Down syndrome patients were similar. This difference suggests that factors other than the rotational instability, which seems to be corrected by epicanthoplasty, may be responsible for the higher failure rate among the Down syndrome patients. Patients with Down syndrome often exhibit a hypoplastic maxilla with a less prominent nose. Specifically the vertical (length of the nasal bridge, height of the nose) and antero-posterior (nasal tip protrusion) dimensions are reduced, and the horizontal dimensions (alar base width, superior and inferior widths of the nostrils) are increased.[11] We speculated that the flatter nose in Down syndrome patients predisposes the cilia of the upper eyelids to rotate medially and eventually touch the cornea.

Compared to non-Down syndrome patients, Down syndrome patients have a higher recurrence rate after upper epiblepharon repair, even in cases with concomitant Z-medial epicanthoplasty. Further advances in surgical techniques that address a possible tendency for increased rotational instability and other associated maxillofacial stigmata of Down syndrome that may contribute to the eyelid malposition may improve recurrence rates.

THE AUTHORS INDICATE NO FINANCIAL SUPPORT OR FINANCIAL CONFLICT OF INTEREST. INVOLVED IN DESIGN OF STUDY (N.J.K., H.K.C.); Conduct of study (K.M.L., N.J.K., H.K.C.); Collection and management of data (K.M.L., N.J.K., H.K.C.); Analysis and interpretation of data (K.M.L., M.J.L., K.-W.L., N.J.K., H.K.C., S.I.K.); Preparation of manuscript (K.M.L., M.J.L., H.K.C.); and Review or approval of manuscript (K.M.L., M.J.L., K.-W.L., N.J.K., H.K.C., S.I.K.). The protocol of this study was approved by the Institutional Review Board of Seoul National University (Boramae) Hospital.

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Biosketch

Kyoung Min Lee, MD, graduated from the Seoul National University Hospital, Korea, in 2007 and underwent residency training in the same hospital. Dr Lee is now working as a chief resident in the Seoul National University Hospital, Korea.