Lash Ptosis in Congenital and Acquired Blepharoptosis

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Objective: To determine the prevalence of lash ptosis (LP) in eyes with congenital and acquired blepharoptosis.

Methods: We retrospectively graded photographs of 228 eyes from 174 patients with congenital or acquired blepharoptosis for LP. We used a 4-point rating scale for LP, in which 0 indicates no LP; 1, minimal; 2, moderate; and 3, severe. A prospective evaluation of LP in 30 eyes from 15 patients without blepharoptosis (control eyes) was also performed.

Results: A total of 107 eyes (in 87 patients) demonstrated congenital blepharoptosis and 121 eyes (in 87 patients) had acquired blepharoptosis. A moderate to severe rating of LP (rating, ≥ 2) occurred in 60.7% of eyes with congenital blepharoptosis, 28.9% of eyes with acquired blepharoptosis, and 6.7% of control eyes. Lash ptosis (rating, ≥ 1) was present in 91.6% of eyes with congenital blepharoptosis, 83.5% of eyes with acquired blepharoptosis, and 33.3% of control eyes. The mean LP rating was 2.1 for eyes with congenital blepharoptosis, 1.3 for eyes with acquired blepharoptosis, and 0.6 for control eyes.

Conclusions: Lash ptosis was common in the patients with blepharoptosis. Moderate to severe LP occurred more commonly in all forms of blepharoptosis compared with normal eyes, with more frequent and more severe LP demonstrated in eyes with congenital blepharoptosis.

Lash ptosis (LP) has received minimal attention in the ophthalmic literature and is often an overlooked sign that may coexist with congenital and acquired blepharoptosis. Also known as eyelash ptosis, this term refers to a global declination of lash follicles of the upper eyelid (Figure 1 and Figure 2).1,2 Although LP is a well-recognized feature of floppy eyelid syndrome (FES), its significance in congenital and acquired blepharoptosis is not well reported.1 To our knowledge, the prevalence of LP in patients with blepharoptosis has not been reported.

Lash ptosis has been associated with FES, congenital lamellar ichthyosis, long-standing ocular leprosy, bilateral acoustic neuroma, and latanoprost-induced LP.2-8 Culbertson and Ostler8 noted that LP is almost universally present in FES. Guimaraes and Cruz2 found LP in 48 of 74 patients with long-standing ocular leprosy (65%). Others3-5 have noted single cases of LP in congenital lamellar ichthyosis and after latanoprost use. Mulhern et al6 studied 62 patients with facial palsy and found that 26 had LP as a long-term consequence (42%).

Hypotheses on the etiology of LP are largely based on anatomical changes within the upper eyelid. The eyelid margin is separated by the gray line into the anterior skin-muscle lamella and a posterior tarsococonjunctival layer. The eyelash bulbs of the upper eyelid lie in a space between the Riolan muscle and the pretarsal orbicularis oculi.9 The eyelash follicles may extend posteriorly to embed in the tarsus.10 The eyelashes emerge through this space and exit through the eyelid margin. The natural contour of the eyelashes projects downward initially with a curvilinear superior and anterior projection away from the globe (Figure 2A and B).9

To our knowledge, the presence of LP in congenital or acquired blepharoptosis has not been studied systematically. A simple, semiquantitative grading scale to categorize LP by severity would aid in stratification of eyes with LP into those that may benefit from surgical correction at the time of blepharoptosis repair and those that may be observed. In addition, a more thorough understanding of the factors involved in the etiology of LP is a prerequisite to proper correction at the time of surgery. We conducted a comparative observational study in which the prevalence and severity of LP in individuals with congenital and acquired blepharoptosis were compared with the prevalence and severity of LP in normal control subjects.

After obtaining institutional review board approval, patients diagnosed as having congenital or acquired blepharoptosis between Janu-
Lash ptosis with a declination below the horizontal plane (LPR, ≥ 1) was noted in 98 of the 107 eyes with congenital blepharoptosis (91.6%), 101 of the 121 eyes with acquired blepharoptosis (83.5%), and 10 of the 30 control eyes (33.3%). A moderate to severe degree of LP (LPR, ≥ 2) was observed in 65 of the eyes with congenital blepharoptosis (60.7%), 35 of the eyes with acquired blepharoptosis (28.9%), and 2 control eyes (6.7%). No LP (LPR, < 1) was observed in 9 eyes with congenital blepharoptosis (8.4%), 20 eyes with acquired blepharoptosis (16.5%), and 20 control eyes (66.7%).

### RESULTS

The congenital blepharoptosis group consisted of 50 males and 37 females with an mean (SD) age of 15.9 (15.1) years. The acquired blepharoptosis group consisted of 50 males and 37 females with a mean (SD) age of 56.7 (19.8) years. There were 107 eyes with congenital blepharoptosis, 121 eyes with acquired blepharoptosis, and 30 normal eyes (control group). The acquired blepharoptosis group included patients with levator dehiscence (63 patients), dermatochalasis (10), anophthalmia (5), thyroid eye disease (3), myasthenia (1), third nerve palsy (1), chronic progressive external ophthalmoplegia (1), phthisis bulb (1), and traumatic blepharoptosis (1) and 1 patient with acquired blepharoptosis due to ectropion repair.

The mean (SD) LPRs were 2.1 (0.8) (95% confidence interval [CI], 1.9-2.2) in eyes with congenital blepharoptosis, 1.3 (1.0) (95% CI, 1.0-1.6) in eyes with acquired blepharoptosis, and 0.6 (0.7) (95% CI, 0.4-0.1) in control eyes. For comparison, the LPRs of 67 fellow eyes with unilateral congenital blepharoptosis and 53 with unilateral acquired blepharoptosis were evaluated, and the results did not differ significantly from those of control eyes (mean LPR, 0.6 for both).

Lash ptosis with a declination below the horizontal meridian characterizes an LPR of 3. Three masked graders were provided an equal number of photographs of the eyes of patients with acquired blepharoptosis who were evaluated between September 5, 2003, and April 1, 2006, were gathered for comparison. Sixteen controls were identified who had had no obvious eyelid abnormality; 1 control subject was excluded secondary to congenital blepharoptosis.

Each eyelid was assessed using a 4-point rating scale for LP and was assigned an LP rating (LPR) (an LPR of 0 indicates no LP; 1, minimal; 2, moderate; and 3, severe) (Figures 1 and 2). An LPR of 0 represents the natural position of eyelashes relative to the eyelid margin (0° to ≥ 30° above the horizontal). Eyelashes oriented nearly parallel to the horizontal meridian relative to the eyelid margin (0° to 30° below the horizontal) are rated as having an LPR of 1. An LPR of 2 characterizes eyelashes with an orientation that is 31° to 45° below the horizontal meridian. Eyelashes oriented at more than 45° below the horizontal meridian characterize an LPR of 3. Three masked graders (A.R.H., K.J.M., and M.S.L.) separately determined the degree of LP in each eye in primary gaze. The LPRs were averaged. Any discrepancy in the rating by greater than 1 was adjudicated. Grading consistency and quality assurance (80%-90% agreement) were confirmed by intragrader and intergrader exercises on a random sample of photographs.

### COMMENT

Although LP has been described in association with several conditions, an objective method of grading LP does not exist, to our knowledge. In their analysis of eyelid changes in long-standing ocular leprosy, Guimarães and Cruz depicted variations of LP. A dichotomous relationship was defined for LP that distinguished LP according to corneal touch. Four variations of LP were pictured, but the authors did not use a scale or rating system. In our study, a simple 4-point grading scale was used to rate the severity of LP. The measurement of angular displacement of the anterior projection in primary gaze determines the LPR (Figure 1).

Three masked graders were provided an equal number of photographs of the eyes of patients with acquired
blepharoptosis and congenital blepharoptosis. The mean LPR in each group demonstrated a higher prevalence of LP in all forms of blepharoptosis relative to control eyes. Eyes with congenital blepharoptosis demonstrated a higher LPR and a greater prevalence of moderate to severe LP than did eyes with acquired blepharoptosis. Moderate to severe LP (LPR, ≥ 2) was more likely to occur in eyes with congenital blepharoptosis (60.7%) than in eyes with acquired blepharoptosis (28.9%). The LPR varied broadly among the eyes with acquired blepharoptosis, with a relatively equal frequency across all LPs. The numbers of each type of acquired blepharoptosis were too small to correlate with LPR.

Our evaluations included assessment of LP in the nonptotic fellow eyes of patients with unilateral congenital or acquired blepharoptosis. Within this subset of eyes, the LPR was similar to that in the control group. The average LPR for both patient groups was 0.6, and we found no significant difference between the groups. Among fellow eyes, severe LP occurred in 9.2% of eyes (11 of 120), moderate LP in 27.5% (33 of 120), and minimal to no LP in 63.3% (76 of 120). This suggests that LP uniquely occurs in eyes with eyelid blepharoptosis.

Lash ptosis has been found in association with several conditions. Review of these cases offers clues to the etiology of LP. Langford and Linberg suggest that LP may result from anatomical changes in the orbicularis oculi, Riolan muscle, and tarsal plate. Netland et al demonstrated a deficiency of elastin in the tarsus and pretarsal orbicularis in FES. Deficiency of elastin plays a role in eyelid laxity and similarly may contribute to a deficiency of eyelash support. Mulhern et al studied patients with facial palsy and found that 42% of them had LP as a long-term consequence. Loss of tone of the pretarsal orbicularis and Riolan muscle may compromise support to these muscle fibers and eyelash follicles. Excess skin laxity, as in dermatochalasis, could also alter the underlying eyelid muscle tension. Furthermore, LP has been reported in association with trichomegaly secondary to latanoprost use.

Relatively larger eyelashes may overwhelm the ability of the supportive apparatus to maintain proper projection of the follicle. Similar changes in eyelid anatomy may explain LP in congenital blepharoptosis. Changes to the tarsus may alter the direction of eyelashes because the hair follicles embed posteriorly within the tarsus. Terminal fibers of the levator aponeurosis then weave through the orbicularis oculi to insert into the subcutaneous tissue and skin. Disruption of these fibers by congenital dystrophy leads to a lack of an eyelid crease and perhaps to a lack of eyelash support. The close proximity of the aponeurosis and orbicularis suggests that loss of these aponeurotic fibers may alter the eyelid anatomy and mechanically cause laxity of the overlying eyelid skin and muscle, hence producing LP. Lash ptosis in congenital and acquired blepharoptosis may stem from a laxity of anterior lamellar structures and an underlying connective tissue deformation.

Surgery for LP requires attention to the primary underlying disease process as well as to the degree of LP. Surgical tightening procedures in FES have been shown to improve both overall eyelid laxity and LPR. However, these procedures would not be effective in cases of LP not resulting from eyelid laxity. Latanoprost-induced LP has been effectively corrected with anterior lamellar repositioning. Guimarães and Cruz recommend that, in cases of leprosy, LP be corrected via resection of the pretarsal anterior lamella and fixation of the muscle-skin layer to the levator aponeurosis. The authors have had success with double-armed 6-0 chromic mattress fashion through the orbicularis near the anterior eyelash line and secured to the superior edge of the tarsus to rotate the eyelashes. Multiple such sutures should be placed because the effect of each suture is limited to its immediate surrounding area.

Lash ptosis is best identified by direct examination of the patient in primary gaze. Evaluation with the chin parallel to the ground allows for the most accurate assessment of the eyelids and eyelashes in relation to normal anatomy. For the evaluation of LP, it is helpful to examine the patient in primary gaze from frontal and lateral perspectives.

In summary, the prevalence of LP has been underreported and is frequently seen in congenital and acquired blepharoptosis. Eyes with congenital blepharoptosis often demonstrate moderate to severe LP. In cases of severe LP in association with blepharoptosis, surgical correction may be warranted to correct both.

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