Congenital eyelid retraction

J R O Collin, L Allen, S Castronuovo

Abstract

Twenty two patients with primary congenital lid retraction affecting either the upper or lower eyelids or both are presented. The clinical features and management are discussed in the hope that recognition of this clinical entity will prevent unnecessary investigation.

Eyelid retraction is usually secondary to thyroid disorders, trauma, proptosis, seventh nerve palsy, or neurological abnormalities affecting the third nerve. Primary congenital lid retraction has been described only relatively rarely as individual case reports.¹⁻⁶ It is not well recognised as a clinical entity, and so individual patients may be investigated unnecessarily and the diagnosis not considered.⁷ This report reviews the findings in 22 patients with congenital lid retraction affecting either the upper or lower eyelid.

Patients and methods

Twenty two patients with congenital lid retraction were examined over a 10-year period. To be included in this series as a case of upper lid retraction the upper lid had to be above the limbus on the affected side with a difference of at least 1½ mm between the upper eyelid levels on the two sides. The contralateral upper lid had to cover the limbus by not more than 1½-2 mm with the eyes in the primary position of gaze to avoid a potential diagnosis of contralateral ptosis. For lower lid retraction the affected lower lid had to be more than 1½ mm below the lower limbus with the contralateral lower lid at the limbus in the primary position of gaze. There was no history of thyroid problems in any of the patients or their mothers. Pregnancy and childbirth had been normal so far as could be ascertained in 18 cases. There were four cases with a history of forceps delivery. There was no other history of trauma. There was no proptosis. Seventh nerve function was normal. The pupils were equal and there was no ocular motor palsy. In all cases only one side was affected. The findings on full ocular examination were normal, and there was no significant refractive error. No other systemic abnormalities were detected in 19 cases. One child had a congenital cleft palate. Two children had asthma.

In all cases the condition was present at birth and this was verified with photographs if the patient presented after infancy. Sixteen patients presented as children before the age of 10 and four between 10 and 25, the range being nine weeks to 50 years of age. The presenting symptom was cosmetic in 18 patients, the condition often being thought to be either a large eye or a contralateral ptosis. Four patients had symptoms of redness or soreness in the mornings when they

Figure 1: Congenital left upper lid retraction.

woke up which required lubricants. There was a positive family history in only one case, with a father being said to have had a similar condition to his daughter, who was the patient. Only one patient had both upper and lower lid retraction.

There were 19 cases with congenital upper lid retraction (Fig 1). The degree of lid retraction varied from 1½ to 5 mm average 3 mm. The levator function varied from 12 to 18 mm and was not significantly different from that on the contralateral side. The affected eyelid was definitely higher on extreme downgaze than the unaffected lid in at least 14 cases, and the rest were equivocal. The skin crease was the same height or lower than on the unaffected side in the majority of cases and was usually deeper, though this was difficult to measure quantitatively. Fourteen patients had normal binocular single vision with normal acuities and a full range of ocular movements. Three patients had a concomitant squint. This was an ipsilateral hyperopia with limitation of depression in two patients and an ipsilateral esotropia in one. In the first two cases the vision in the affected eye was normal, and in the third it was hand movements. Three patients had limitation of elevation on the involved side. Two patients had anisometropic amblyopia in the contralateral eye.

There were four cases with a congenital lower lid retraction (Fig 2). This varied from 2 to 4 mm, average 2.8 mm. The lower lid retractor function appeared normal, but it was not easy to get accurate measurements. There was limitation of elevation of the lower eyelid in full upgaze. The lower lid skin creases were marked. All cases had full binocular single vision with normal visual acuities and normal ocular movements.

Figure 2: Congenital left lower lid retraction.

Moorfields Eye Hospital,
City Road, London
EC1V 2PD
J R O Collin
L Allen
S Castronuovo

Correspondence to:
J R O Collin, FRSC.
Accepted for publication
22 March 1990

Figure 3: Same patient as in Figure 1 postoperatively.

All cases were observed for a minimum of one month (range one to 18 months). There was no change in the physical signs in any of the patients, and when they had not been seen early in life photographs and the subjective opinion of the patients and their families suggested that the condition had not changed. The indication for surgery in 15 patients was the cosmetic appearance of the upper or lower eyelid, and in only four cases was surgery performed because of symptoms of corneal exposure. If the patient presented as a child and was asymptomatic, the preferred age for surgery was around 4 years, when the child was aware of the defect and postoperative care was simplified.

At operation there was no gross limitation of movement of either the affected upper or lower eyelids on lid traction. An anterior approach was used initially to lower the upper lid in 11 patients and a posterior approach in five. Three patients did not undergo surgery. At operation the lid retractor complex appeared relatively normal, with a normal or thicker than normal levator muscle and aponeurosis but no obvious abnormality of the other eyelid structures. Routine light microscopy similarly did not show any gross abnormality, but one report did suggest that striated levator muscle fibres extended more anteriorly than usual. The lid was lowered with a z-myotomy or recession of the upper lid retractors, including Müller's muscle and the aponeurosis, with lysis of Whitnall's superior suspensory ligament of the globe as required. No scleral or other grafts were used. The lid was put on traction for 24-48 hours, and massage was used postoperatively if necessary to try to prevent the lid from rising too much.

Of the four patients with lower lid retraction all underwent surgery by the posterior approach. In two cases the lid was elevated by a recession of the lower lid retractors only, and two had a scleral graft interposed between the lower lid retractors and the lower border of the tarsus. The sclera was covered by mobilised conjunctiva.

Figure 4: Congenital right upper lid retraction.

Figure 5: Same patient as in Figure 4 postoperatively with ptosis.

Postoperatively the lower lid was put on traction for 48 hours. No gross abnormality of any of the lower lid structures was detected at surgery.

Results

Sixteen of the 19 patients with upper lid retraction underwent upper lid lowering. Twelve achieved a successful result (Fig 3), but in one patient the lid was too low and in three the lid was too high, with a difference of more than 1 mm between the upper lids. The patient whose lid was lowered excessively underwent subsequent ptosis surgery with a successful result (Figs 4, 5, 6). The three patients whose lids were not lowered adequately underwent a further upper lid lowering procedure with a successful result in two. One is waiting for further surgery when older.

All four of the patients with lower lid retraction underwent lower lid raising and achieved a satisfactory result, with lid levels which were approximately equal (Fig 7). The follow-up time for the results of both upper and lower lid surgery varied from one month to 2.3 years, average 11.2 months.

Discussion

Primary congenital lid retraction is a distinct entity and does not appear to change with time. It can involve the upper or lower eyelids or both and can only be diagnosed after excluding the other causes of lid retraction. The majority of cases present because of abnormal appearance, and in this series only 20% had symptoms of corneal exposure. It seems reasonable to manage these patients initially with observation to confirm the diagnosis by ensuring that the condition remains stable. Extensive investigations are not required. Lubricants can be prescribed if there are symptoms of corneal exposure. Observation

Figure 6: Same patient as in Figures 4 and 5 postoperatively after ptosis correction.

Figure 7: Same patient as in Figure 2 postoperatively.

includes orthoptic assessment, since three cases in this series did have squints, two of which were associated with amblyopia.

Eyelid retractor lengthening procedures are effective but the results are unpredictable (Figs 4-6). It is wise to delay surgery until the child is old enough to co-operate and to facilitate post-operative massage and so on which may be required to try to control the eyelid height. This usually means operating at about 4 years of age, but early surgery may be indicated if the child has symptoms of corneal exposure unrelieved by lubricants. The choice of an anterior or posterior approach to lower the lid depends on the height of the skin crease and the individual surgeon's preference. The skin crease will rise with a posterior approach retractor recession but theoretically can be controlled with an anterior approach procedure. Those patients with the greatest lid retraction all underwent an anterior approach upper lid retractor recession in an attempt to keep the skin crease equal in both upper eyelids. The posterior approach was used to correct the more minor degrees of lid retraction. The control of lid height and contour appeared better with the posterior approach, but this could have been because a lesser degree of change in eyelid level was being attempted. The predictability of eyelid lowering in this series would probably have been improved by delaying surgery until all the patients were old enough for local anaesthesia, but there was one adult whose result was unsatisfactory even after surgery under local anaesthesia. The results of upper lid

retractor lengthening are variable in cases with other causes of lid retraction, such as thyroid eye disease, and there does not seem to be any particular way of reducing this unpredictability.⁸

The aetiology of the condition remains obscure. Examination of the eyelid tissues at operation did not show any obvious gross abnormality, though it did seem that the levator muscle fibres perhaps extended more anteriorly than usual and that the aponeurosis and orbital septum were particularly well formed as distinct layers. Routine histological examination confirmed the presence of normal looking, striated levator muscle fibres. The levator function in all these cases was good, but there was lagophthalmos on downgaze, which suggests that either the levator muscle or Müller's muscle did not relax properly. This might be due to an element of dystrophy, hypertrophy, or aberrant innervation of the muscle, or because the muscle, aponeurosis, or orbital septum was congenitally shorter and thicker than normal. It was not possible in this series to come to any more definitive conclusions about the aetiology, but our findings do show that the condition exists as an idiopathic, primary, benign entity, recognition of which may save patients unnecessary investigation.

This paper was presented in part to the American Society of Ophthalmic Plastic and Reconstructive Surgery in New Orleans on 28 October 1989.

- 1 Putterman AM, Urist M. Surgical treatment of upper eyelid retraction. *Arch Ophthalmol* 1972; 87: 401-5.
- 2 Yee RD, Odeh-Nasralla N. Congenital, paradoxical retraction of the lower eyelid. *Am J Ophthalmol* 1974; 78: 1027-9.
- 3 Baylis HI, Kamin DF. Correction of upper eyelid retraction. *Am J Ophthalmol* 1976; 82: 790-4.
- 4 Leone CR Jr, Lewis R. Congenital upper eyelid retraction. *J Pediatr Ophthalmol Strabismus* 1976; 13: 350-2.
- 5 Ballen PH, Rochkopf L. Congenital retraction of the upper lid. *Ophthalmic Surg* 1987; 18: 689-90.
- 6 Rovit AJ, Deupree DM, Zang Y, Biglan AW. Treatment of congenital unilateral upper eyelid retraction with a marginal myotomy procedure. *Ophthalmic Surg* 1988; 19: 872-5.
- 7 Shields CL, Nelson LB, Carpenter GC, Shields JA. Neonatal Graves' disease. *Br J Ophthalmol* 1988; 72: 424-7.
- 8 Collin JRO. *A manual of systematic eyelid surgery* 2nd ed. London: Churchill Livingstone, 1989: 144-6.