Incidence and clinical characteristics of pediatric eyelid retraction

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PURPOSE To describe the incidence and clinical characteristics of upper and lower eyelid retraction in children.

METHODS The medical records of all pediatric patients (<19 years of age) diagnosed with upper and/or lower eyelid retraction from January 1, 1976, through December 31, 2010, were retrospectively reviewed.

RESULTS A total of 85 patients were diagnosed during the 35-year period, of whom 15 were residents of Olmsted County, Minnesota, yielding an annual age- and sex-adjusted annual incidence of 1.38 per 100,000 persons (95% CI, 0.70-2.05), or 1 in 72,463. Upper eyelid retraction was documented in 38 patients (45%; 24 unilateral and 14 bilateral); lower, in 25 (29%; 12 unilateral and 13 bilateral); and both upper and lower in 18 (21%; 3 unilateral and 15 bilateral). It was not recorded in 4 patients. The most common causes of eyelid retraction were thyroid eye disease (48 [56%]), primary congenital eyelid retraction (11 [13%]), and trauma (9 [11%]). Although there were no cases of visual impairment secondary to eyelid retraction, tearing, ocular surface irritation, and photophobia were noted in 38 patients (45%). Nineteen patients (22%) underwent surgical eyelid correction.

CONCLUSIONS Pediatric eyelid retraction is relatively rare, occurring in approximately 1 in 72,000. The leading causes of childhood eyelid retraction in this cohort were thyroid eye disease, primary congenital eyelid retraction, and trauma. No visual disturbances due to eyelid retraction were noted, and approximately 1 in 5 patients required corrective eyelid surgery. (J AAPOS 2019;23:213.e1-4)

Eyelid retraction, a vertically larger-than-normal opening of the eyelid, is relatively rare in children compared to adults. Etiologies of pediatric eyelid retraction include congenital aberrant innervation of the oculomotor nerve, trauma, thyroid eye disease (TED), hydrocephalus (the setting sun sign), dorsal midbrain syndrome (Collier sign), shallow orbits, and other rare causes.[1-3] The differential diagnosis and classification of eyelid retraction for both adults and children was reported by Bartley in 1995.[1] To our knowledge, however, there are no population-based studies on the clinical characteristics and etiology of childhood eyelid retraction. The purpose of the present study is to describe the incidence, clinical features, and management of childhood eyelid retraction diagnosed over a 35-year period in patients <19 years of age.

Subjects and Methods

This study was approved by the Institutional Review Board at Mayo Clinic and Olmsted Medical Group, complied with the US Health Insurance Portability and Accessibility Act of 1996, and adhered to the tenants of the Declaration of Helsinki. The medical records of all patients <19 years of age diagnosed as having eyelid retraction from January 1, 1976, through December 31, 2010, were retrospectively reviewed. Potential cases were identified using the Rochester Epidemiology Project, a medical records linkage database designed to facilitate population-based studies in Olmsted County, Minnesota.[4,5] The population of Olmsted County (124,277 in 2000) is relatively isolated from other urban regions, and nearly all medical care is provided to residents by Mayo Clinic or Olmsted Medical Group and affiliated hospitals.

The medical records of all potential patients were diagnosed and reviewed by an ophthalmologist (BGM). Eyelid retraction was considered present if the sclera was visible above or below the superior corneal limbus or inferior corneal limbus, respectively. Patients were considered to have a congenital form of eyelid retraction if treatment was sought from a physician within

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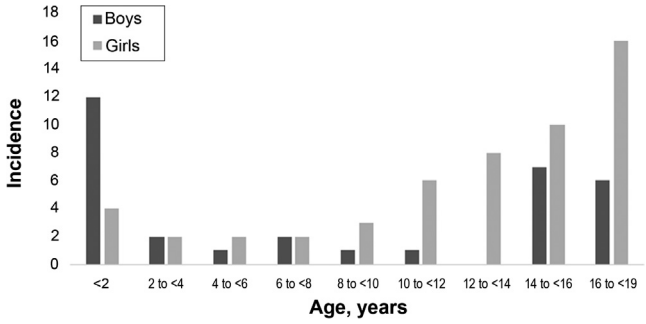

FIG 1. Age at diagnosis of pediatric eyelid retraction by sex (January 1, 1976, to December 31, 2010).

the first few months of life unless an acquired cause was specifically noted in the record. Late-presenting cases were deemed congenital if symptoms were observed within the first few months of life and were verified by a photograph or parental history.

To determine the incidence of eyelid retraction in children in Olmsted County, annual age- and sex-adjusted incidence rates were derived using the age- and sex-specific population figures for Olmsted County from the United States census. The 95% confidence intervals were calculated using assumptions based on the Poisson distribution.

Results

A total of 85 new cases of pediatric eyelid retraction were diagnosed during the 35-year period. The median age of diagnosis was 13.5 years (range, 1 day to 18.9 years), and 53 patients (62%) were female (Figure 1). Fifteen of the 85 patients resided in Olmsted County at the time of their diagnosis, yielding an annual age- and sex-adjusted annual incidence of 1.38 per 100,000 persons (95% CI, 0.70-2.05), or 1 in 72,463 patients <19 years of age. Upper eyelid retraction was documented in 38 patients (45%); lower, in 25 (29%); and both upper and lower, in 18 (21%). Of the 38 cases of upper eyelid retraction, 24 (63%) were unilateral; 14 (37%), bilateral. Thirteen patients (52%) with lower eyelid retraction were bilateral. Of the 18 cases with both upper and lower eyelid retraction, 15 (83%) had bilateral involvement (Table 1).

The most prevalent etiologies included TED in 48 patients (56%), primary congenital eyelid retraction in 11 patients (13%), and facial trauma in 9 patients (11%), as shown in Table 1. Of the 15 patients residing in Olmsted County, 6 had TED, 5 had primary congenital eyelid retraction, 2 had a history of trauma, and 1 each had craniosynostosis and cicatricial eyelid retraction following congenital ptosis repair. Six (40%) of the 15 patients had bilateral involvement, 7 involved the left eye only, and the remaining 2 involved the right eye only.

Of the 85 patients, 38 (45%) presented with clinical signs, including ocular surface irritation in 22, tearing in 19, photophobia in 12, and diplopia in 3 at the time of their diagnosis. Documented ocular abnormalities such as lid lag, enophthalmos, proptosis, ectropion, and exposure keratitis were noted in 48 patients (56%); 21 (25%) were also noted to have clinically significant lagophthalmos. Three patients were reported to have amblyopia due to congenital ptosis (2) or refractive error (1). Other than the 11 cases of primary congenital eyelid retraction, 1 of the 3 patients with craniosynostosis had congenital eyelid retraction within 2 months of life. The remaining 73 cases were acquired eyelid retraction. There were no congenital cases of Graves orbitopathy in this study.

During a mean follow-up of 5.72 years (range, 1 month to 31 years), 63 of the 85 patients were managed conservatively, with topical lubrication when warranted, and 19 patients (22%) were managed surgically, through a variety of eyelid corrective procedures. The remaining 3 patients (3.5%) experienced improvement of eyelid position after orbital decompression for TED. All patients who underwent eyelid surgery experienced clinical improvement with the exceptions of 2 patients, whose condition remained unchanged. One of these patients had retraction caused by trauma; the other had linear scleroderma with hemifacial atrophy. None of the 85 patients were documented to have visual impairment secondary to eyelid retraction during the follow-up period.

Discussion

This study describes the incidence and clinical features of eyelid retraction from a large cohort of pediatric patients. Childhood eyelid retraction occurred in 1 in 72,463 individuals younger than 19 years in Olmsted County. The majority of cases were bilateral upper eyelid retraction in females occurring in the second decade of life. Four out of 5 children in this study had one of three diagnoses: TED, primary congenital eyelid retraction, or trauma. Rare forms of eyelid retraction comprised the remaining 20% of patients. Surgical intervention was required in a minority (22%) of patients.

TED was the most prevalent subtype of eyelid retraction among children in this study, comprising 56% of all cases, of which 3 of 4 were females. This predominance of females with TED has been reported to range from 75% to 79%,[6-8] although prior studies did not specifically refer to eyelid retraction. Figure 1 shows the study patients' sex by age at diagnosis, including a predominance of females presenting with TED in the second decade of life.

Traumatic eyelid retraction most commonly occurred among boys during the teenage years. The preponderance of both endocrinological (41/48 [85%]) and traumatic (7/9 [66%]) causes of eyelid retraction in the second decade of life is likely due to the onset of pubescence with hormonal changes and increased risk-taking behaviors, respectively.

Primary congenital eyelid retraction occurred more commonly among boys, usually in the first several months of life. Stout[2] also reported a majority of males (4 of 5 children) with congenital eyelid retraction after excluding underlying causes. It is unknown whether this apparent male predominance is real or not, given the relatively

Table 1. Etiology and demographics of 85 patients <19 years of age diagnosed with pediatric eyelid retraction in Olmsted County, Minnesota, 1976-2010

Etiology	Sex M/F	Laterality			Eyelid involvement		
		Right	Left	Bilateral	Upper U/B	Lower U/B	Upper & lower U/B
Thyroid (n = 48)[a]	12/36	4	10	34	11/9	1/11	2/10
Primary congenital (n = 11)	8/3	5	3	3	7/2	—	1/1
Trauma (n = 9)	5/4	4	5	—	3/0	6/0	—
Orbital tumor (n = 4)	1/3	1	2	1	—	3/0	0/1
Cicatricial (n = 2)	0/2	1	1	—	2/0	—	—
Craniosynostosis (n = 3)	2/1	—	—	3	—	0/1	0/2
Chiari II with hydrocephalus (n = 3)	2/1	—	1	2	1/2	—	—
Linear scleroderma with hemifacial atrophy (n = 1)	0/1	—	1	—	—	—	0/1
CN 4, 6, 7 palsy (n = 1)	0/1	1	—	—	—	1/0	—
Hydrocephalus/intraventricular hemorrhage (n = 1)	1/0	—	—	1	0/1	—	—
Treacher collins syndrome (n = 1)	1/0	1	—	—	—	1/0	—
Idiopathic (n = 1)	0/1	—	—	1	—	0/1	—
Total = 85	32/53	17	23	45	24/14	12/13	3/15

B, bilateral; *CN*, cranial nerve; *U*, unilateral.
[a]Of 48 thyroid cases, 4 patients were reported to have bilateral eyelid retraction without upper versus lower eyelid involvement specified.

uncommon occurrence and infrequent systemic evaluation of newborns with eyelid retraction. The largest cohort to date was reported by Collins and collagues[9] and included 22 patients with primary congenital eyelid retraction; however, no information was provided regarding the gender of these children. The authors[9] described anterior extension of the levator muscle fibers during biopsy and histologic studies confirmed normal-appearing striated levator muscle fibers. Case reports of primary congenital eyelid retraction by Ballen and Rochkopf[10] and Leone and Lewis[11] describe pathologic thickening of the lateral and medial horns of the levator aponeurosis, but with grossly normal levator function. Mee and McNab[12] published 2 cases of congenital lower eyelid retraction due to a tight inferior rectus muscle, a potential variant of ocular fibrosis syndrome. Nevertheless, there is no known definite cause for congenital upper or lower eyelid retraction.

TED is rare in children compared to adults.[13,14] In adults, eyelid retraction is reported to be the most common clinical sign of Graves ophthalmopathy, observed in 38% to 75% of patients.[15,16] Similarly, eyelid retraction in children is also strongly linked to TED, as demonstrated in this study. The prevalence of eyelid retraction among children with TED has ranged from 83% to 91% in prior reports with relatively small populations.[7,17] The occurrence of eyelid retraction in children without aberrant regeneration, particularly in the second decade of life, should prompt an investigation for thyroid dysfunction.

Disease laterality is suggestive but not pathognomonic with respect to underlying etiology. TED, for example, generally involves both eyes but presented unilaterally in 29% of TED cases in this study. Eyelid retraction due to trauma occurred unilaterally and primary congenital eyelid retraction showed no predilection, equally occurring on the right side, the left side, or in both eyes. In acquired unilateral or bilateral eyelid retraction without a known etiology, further evaluation should be performed including thyroid studies. Additional features that warrant neuroimaging include levator synkinesis suggestive of an oculomotor nerve palsy, proptosis, ophthalmoplegia, and new-onset unilateral eyelid retraction.

Symptomatic medical therapy and surgical intervention were employed in less than half of the patients managed in this series. Symptoms of corneal exposure were observed in approximately 2 of 5 patients and were generally managed medically. Additional ocular abnormalities, such as proptosis and lid lag, primarily associated with TED, were thought to have contributed to the increased prevalence of symptoms. Surgical intervention, occurring in 1 in 5 children in this study, resulted in improvement of the eyelid malposition in nearly all patients. Indications for surgical management included severe exposure keratopathy, incomplete lid closure, visually threatening symptoms, chronically stable retraction, and undesirable lid height asymmetry.[2,18,19] Other investigators have detailed the advantages and disadvantages of the various surgical approaches to upper eyelid retraction, including mullerectomy, spacer grafts, levator recession, modified full-thickness anterior blepharotomy, and tarsorrhaphy.[2,19-22]

There are several limitations to the results of this study. Its retrospective design is limited by incomplete data and uneven follow-up. Additionally, although all patients were diagnosed or reviewed by an ophthalmologist, some cases may have been initially evaluated by a non-ophthalmologist and potentially excluded from the study, thereby underestimating the true incidence of the condition in this population. Other, more mild, forms of retraction may have gone unnoticed and avoided the study altogether. Although Olmsted County is relatively isolated, some residents with eyelid retraction may have sought care outside the region, potentially resulting in underestimation of the

incidence. However, this is the only known population-based report on pediatric eyelid retraction, finding that thyroid disease, primary congenital eyelid retraction, and trauma were the most prevalent etiologies.

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