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Strabismus in Childhood Eyelid Ptosis

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Abstract

Purpose—To report the prevalence and causes of strabismus among children with eyelid ptosis diagnosed in a well-defined population over a 40-year period.

Design—Retrospective, population-based cohort study.

Methods—We retrospectively reviewed the charts of one hundred and seven patients (< 19 years) for the prevalence and causes of strabismus who were diagnosed with childhood eyelid ptosis as residents of Olmsted County, Minnesota, from January 1, 1965, through December 31, 2004.

Results—Strabismus was diagnosed in 20 (18.7%) of the 107 patients with childhood ptosis. Eight (9.9%) of the 81 patients diagnosed with simple congenital ptosis had strabismus, of which there were 4 (4.9%) cases of exotropia and 4 (4.9%) cases of esotropia. There were no cases of isolated vertical deviation.

Conclusions—Strabismus occurred in 1 in 5 children diagnosed with any form of childhood ptosis in this population-based cohort. Strabismus affected approximately 1 in 10 patients diagnosed with simple congenital ptosis, with a predominance of isolated horizontal deviations equally divided between esotropia and exotropia.

Strabismus has been reported to occur in 10.3 to 32% of patients with childhood ptosis.[1-3] The purpose of this study is to describe the rate and types of strabismus among a population-based cohort of 107 children diagnosed with ptosis during a 40-year period.

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Methods

The medical records of all patients younger than 19 years of age with childhood ptosis, while residing in Olmsted County, Minnesota, diagnosed between January 1, 1965, and December 31, 2004, were retrospectively reviewed after obtaining institutional review board approval. The medical records of 107 patients (81 patients with simple congenital ptosis) met inclusion criteria[4] and were reviewed for the presence of strabismus defined as an intermittent or constant horizontal deviation of 10 or more prism diopters, a vertical deviation of 2 or more prism diopters, or other eye movement disorders. The incidence and demographics of this population of children with eyelid ptosis and the prevalence of amblyopia have been previously reported.[4,5]

Results

Twenty (18.7%) of the 107 study patients were diagnosed with strabismus in Olmsted County, Minnesota, during the 40-year period. Further information concerning the forms of ptosis and strabismus are shown in the Table. There were 11 (55%) females compared to 9 (45%) males. All cases of strabismus occurred in patients with unilateral ptosis with the exception of one patient each with congenital fibrosis of the extraocular muscles (CFEOM), Noonan syndrome, and childhood myasthenia gravis.

Eight (9.9%) of the 81 patients with simple congenital ptosis were diagnosed with strabismus, of which there were 4 (4.9%) cases of exotropia and 4 (4.9%) cases of esotropia. There were no cases of isolated vertical deviation. However, two patients with exotropia had associated vertical deviations; overactive inferior oblique muscles in one and dissociated vertical deviation in a second. The median age of diagnosis in those simple congenital ptosis patients with esotropia was 5.02 (range, 1.59 – 10.6) years, and with exotropia was 1.63 (range, 1.35 – 9.37) years. Two of the eight patients with simple congenital ptosis and strabismus had amblyopia. Both cases were in children with exotropia and in each case the amblyopia was secondary to strabismus.[5]

Discussion

Strabismus occurred in nearly 1 in 5 patients with any form of childhood ptosis in this population-based cohort diagnosed over a 40-year period. Within the group of 96 patients with any congenital form of ptosis, 81 patients were diagnosed with simple congenital ptosis, of which, 8 (9.9%) patients were diagnosed with strabismus. This rate falls at the low end of the range of previous non population-based estimates (10.3-32%) of strabismus associated with congenital ptosis.[1-3]

Strabismus has an estimated prevalence of 1% to 5% in the general population.[3] There is no definitive explanation as to why the incidence of strabismus is higher among patients with simple congenital ptosis, although genetic predisposition or an intrauterine insult (e.g. fibrin emboli and focal hypoperfusion) to overlapping regions of the oculomotor nuclear complex or the third cranial nerve may play a leading role.[3,6] Furthermore, it has been suggested that some cases of strabismus may be secondary to visual occlusion and disruption of binocularity by the ptotic eyelid.[2]

While an insult to the superior division of the third cranial nerve would clearly result in a vertical deviation along with ptosis due to disruption of superior rectus and levator superioris palpebrae muscle function, prior reports have instead found a strong predominance of isolated horizontal deviations in patients with simple congenital ptosis.[1,3] Indeed, while vertical deviations were noted in two simple congental ptosis patients with horizontal strabismus in this cohort, there were no cases of isolated vertical deviation noted in the study. Horizontal strabismus occurred in 9.9% of the 81 patients with simple congenital ptosis.

There are imitations to the findings in this study including the relatively homogeneous semi-urban white population demographics of Olmsted County, along with the potential that some patients sought care outside the region, thus underestimating incidence data. Further limitations for this cohort of patients has been previously published.[4,5]

The findings of this study provide population-based prevalence rates for strabismus in childhood ptosis diagnosed over a 40-year period. Strabismus affected approximately 1 in 10 patients diagnosed with simple congenital ptosis, with a predominance of isolated horizontal deviations equally divided between esotropia and exotropia.

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Biography

Vitae: Greg Griepentrog is an assistant professor of Oculofacial and Orbital Surgery in the Department of Ophthalmology at the Medical College of Wisconsin, Milwaukee, Wisconsin. His research interests include ophthalmic epidemiology, facial anatomy, and periocular surgical techniques.

Table 1

Strabismus in 107 patients <19 years diagnosed with childhood ptosis in Olmstead County, Minnesota from 1965 to 2004

Ptosis Etiology	Number (%)	Strabismus Cases (%)		
		Exotropia (XT)	Esotropia (ET)	Other Forms of Straismus
Congenital				
Simple Congenital Ptosis	81 (75.7)	4 (4.9)[a]	4 (4.9)	0
Blepharophimosis	3 (2.8)	1 (33)	0	0
Congenital CNIII Palsy	3 (2.8)	0	0	3 (100)
Marcus Gunn Jaw Wink	3 (3.8)	0	1 (33)	0
Congenital Horner's Syndrome	2 (1.9)	0	0	0
Central Core Myopathy	1 (0.93)	0	0	0
CFEOM	1 (0.93)	0	0	1 (100)
Myotonic Dystrophy	1 (0.93)	0	0	0
Noonan Syndrome	1 (0.93)	0	1 (100)[b]	0
Acquired				
Aponeurotic Dehiscence	4 (3.7)	0	1 (25)	0
Acquired CN III Palsy	2 (1.9)	0	0	2 (100)
Acquired Horner's Syndrome	2 (1.9)	0	0	0
Traumatic Structural Ptosis	2 (1.9)	0	0	1 (50)
Childhood Myasthenia Gravis	1 (1.9)	1 (100)	0	0
Total	**107**	6 (5.6)	7 (6.5)	7 (6.5)

CFEOM = congenital fibrosis of the extraocular muscles; CN = cranial nerve.

[a] One XT patient with additional overactive inferior oblique muscles, and another XT patient with dissociated vertical deviation.

[b] Child with both ET and left hypertropia.