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Prevalence of dissociated strabismus in children with ocular misalignment: a population-based study

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Abstract

Purpose—The purpose of this study was to report the prevalence of dissociated strabismus among a population-based cohort of children diagnosed with all forms of ocular misalignment.

Methods—The medical records of all children (<19 years of age) identified by the resources of the Rochester Epidemiology Project who were diagnosed with strabismus while residing in Olmsted County, Minnesota, from January 1, 1985, through December 31, 1994, were retrospectively reviewed for the prevalence of dissociated vertical (DVD) or horizontal (DHD) deviations.

Results—Of 627 children with strabismus, 12 (1.9%) were found to have DVD at their initial diagnosis at a mean age of 52 months (range, 3.5–206 months). During a mean follow-up of 10.4 years (range, 1–20 years), an additional 44 children were diagnosed with DVD, yielding a total of 50 children (7.9%). The common strabismus subtypes with the highest prevalence of DVD were, in declining order, congenital esotropia (53%), esotropia in the setting of a developmental deficit (25%), and accommodative esotropia (3.4%). None of the subjects were recorded as having DHD.

Conclusions—In this patient cohort, DVD was relatively uncommon occurred most frequently among those with early-onset strabismus and strabismus in the setting of a developmental deficit.

Dissociated vertical deviation (DVD) is characterized by an upward extorsional deviation of either eye that occurs when the patient is fatigued or daydreaming, or when fusion is disrupted on occlusion followed by a downward movement of the eye when a refixation stimulus occurs.^{1–3} The condition is almost always bilateral (often asymmetrical) and can be differentiated from a hypertropia by a lack of hypotropia in the contralateral eye.⁴ DVD is reported to be associated with early disruption of normal binocular function; however, its exact etiology remains unknown.^{1,2,5} Theories explaining the mechanism of DVD have been proposed by Brodsky,⁶ Guyton,⁷ and Ten Tusscher and Van Rijn.⁸

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Although DVD has been reported to be associated with various forms of congenital and acquired strabismus, the literature describes its association primarily with infantile esotropia.^{4,5,9} There are, to our knowledge, no population-based studies on the prevalence of DVD among a cohort of children with strabismus. The purpose of this study was to assess the prevalence of dissociated strabismus among a population-based cohort of patients younger than 19 years of age diagnosed with any form of ocular misalignment diagnosed during a 10-year period using a population-based medical record retrieval system.

Subjects and Methods

Mayo Clinic Institutional Review Board approval was obtained for this study. The medical records of all patients <19 years of age who were residing in Olmsted County, Minnesota, when diagnosed with childhood strabismus from January 1, 1985, through December 31, 1994, were retrospectively reviewed. Potential cases of childhood strabismus were identified using the resources of the Rochester Epidemiology Project, a medical records linkage system designed to capture data on any patient–physician encounter in Olmsted County, Minnesota.^{10,11} The racial distribution of Olmsted County residents in 1990 was 95.7% white, 3.0% Asian-American, 0.7% African American, and 0.3% each for Native American and other. The population of this county (106,470 in 1990) is relatively isolated from other urban areas and virtually all medical care is provided to residents by Mayo Clinic and their affiliated hospitals. Unaffiliated area optometrists were rarely the sole provider of eye care to children with strabismus during the study years of this investigation.

Patients newly diagnosed by an ophthalmologist as having any form of horizontal or vertical strabismus during the study period were identified. The surgical records of patients who underwent inferior oblique (myectomy and anterior transposition) or vertical rectus surgery were also reviewed to avoid missing any potential cases of DVD. All diagnoses were entered into the Rochester Epidemiology Project database by trained personnel who reviewed the entire medical record. Trained residency checkers verified the patients' residency status at the time of birth and at diagnosis using information from city and county directories. Children not residing in Olmsted County at the time of their diagnosis were excluded. The medical records of all patients were reviewed for the occurrence of dissociated strabismus at the initial and final follow up examination.

Results

A total of 627 new cases of childhood strabismus were identified during the study period. The specific types of strabismus, their definition, and proportion of each, has previously been reported.¹²⁻¹⁴ Central nervous system (CNS) strabismus was defined as strabismus in the setting of a developmental deficit and both terms are used interchangeably.

At the initial examination, 12 children (1.9%) were found to have DVD at a mean age of 52 months (range, 3.5-206 months), as shown in Table 1. DVD was diagnosed in 2 (6.6%) of 30 children with infantile esotropia at a mean age of 0.4 years, 2 (1.1%) of 175 children with accommodative esotropia at a mean age of 5.5 years, and in 1 each of children with CNS esotropia, intermittent exotropia, CNS exotropia, sensory exotropia, CNS hypertropia,

and other forms of hypertropia. Two of the 627 had isolated DVD without horizontal strabismus. The 2 cases of isolated DVD without associated horizontal strabismus were found during review of the surgical records of patients who, during the study interval, underwent inferior oblique or vertical rectus muscle surgery. There was no known family history of strabismus for these 2 patients and neither underwent horizontal strabismus surgery.

During a mean follow-up of 10.4 years (range, 1-20 years), an additional 44 patients were diagnosed with DVD, yielding a total of 50 children (7.9%) with a final diagnosis of DVD. Of the 12 patients diagnosed with DVD at the initial examination, only 6 had a final diagnosis of DVD. Of these 6 patients, 2 belonged to the congenital esotropia subgroup and 1 each belonged to the CNS esotropia, exotropia, hypertropia, and isolated DVD subgroups. An additional 44 children developed DVD over a mean follow-up of 10.4 years. DVD was ultimately diagnosed in 16 of 30 children (53.3%) with infantile esotropia at a mean age at final visit of 11.2 years, 2 of 5 children (40%) with CNS hypertropia at a mean age at final visit of 16.7 years, 11 of 44 children (25%) with CNS esotropia at a mean age at final visit of 15.8 years, and 2 of 17 children (11.7%) with sensory exotropia at a mean age at final visit of 9.4 years. DVD occurred in 1 child in the subgroups of acquired nonaccommodative esotropia, sensory esotropia, other forms of esotropia, congenital exotropia, paralytic hypertropia, and with other forms of hypertropia. DVD was associated with intermittent exotropia and accommodative esotropia in less than 5% of patients (Table 1). The mean age at final visit of children with DVD in association with various forms of esotropia, exotropia, and hypertropia was 15.3 years. There were no cases of dissociated horizontal deviations recorded in this cohort.

Discussion

This study provides population-based data on the prevalence of DVD in childhood strabismus from an upper Midwest population of the United States. We found a prevalence of 1.9% on initial presentation at a mean age of 52 months (range, 3.5-206 months), increasing to 7.9% after a mean follow-up of approximately 10 years. Dissociated vertical deviation was found to be most prevalent among children with early onset strabismus and strabismus in the setting of a developmental deficit.

Although there are no known studies on the prevalence of DVD, some reports have discussed the prevalence in relation to the timing of surgical intervention. Arslan and colleagues⁴ studied the development of DVD among children with infantile esotropia who underwent surgical treatment and that, overall, 47.9% of children with infantile esotropia developed DVD at an average age of 3.2 years, similar to the results of the present study (53.3%). DVD had occurred in 24.1% of the cases who underwent surgery between the ages of 6 months and 2 years and in 52% of the cases that were surgically treated after age 2 years; however, their study did not include other forms of strabismus.⁴ In contrast, Neely and colleagues⁹ reported that DVD occurred in 92% of patients treated for infantile esotropia and hypothesized that its development occurs regardless of the timing of surgical intervention in the first 24 months of life. The higher prevalence in their series may be due

to referral bias of a tertiary treatment center or to a later age at DVD diagnosis. Neither group, however, evaluated the occurrence of DVD in other forms of strabismus.

The prevalence of DVD in esotropic children with an abnormal CNS has not, to our knowledge, been reported in the literature. The findings from the present study demonstrate a prevalence of 25% in CNS esotropia, the strabismus subtype with the second highest prevalence of DVD following infantile esotropia among common subtypes. This study also found an association between DVD and other CNS strabismus subtypes, including CNS hypertropia (40%) and CNS exotropia (10%).

The occurrence of DVD in more common forms of strabismus, such as accommodative esotropia and intermittent exotropia, is poorly documented in the literature. DVD occurred in 3.4% of children with accommodative esotropia in this cohort; however, there are no known reports available for comparison. The prevalence of DVD among patients with intermittent exotropia has been reported by Lim and colleagues,⁵ who found that patients with intermittent exotropia and DVD developed strabismus at a younger age than those with intermittent exotropia without DVD. It was further demonstrated that, when compared to the infantile esotropia group with DVD, the amount of DVD measured in patients with intermittent exotropia was smaller.⁵ However, no prevalence rates were reported, whereas the present study found a rate of 1.8% in children with intermittent exotropia. Among patients with sensory strabismus, Kutluk and colleagues² found that DVD occurred more frequently in sensory esotropia than exotropia (18.3% vs 8.1%); however, they found a higher prevalence of DVD in sensory exotropia (11.7%) compared to sensory esotropia (4%).

DVD has been reported to occur, although less commonly, in association with vertical deviations.¹⁵ Cases of DVD in monocular elevation deficiency have been well recognized and develop more commonly in the younger age groups of patients being observed for long periods of time and who have an associated horizontal strabismus.¹⁵ The present study found DVD in 1 of 5 children (20%) with CNS hypertropia and 1 of 16 (6.3%) with other forms of hypertropia.

Several theories regarding the etiology of DVD have been advanced. Brodsky⁶ proposed that it results from asymmetrical visual input after disrupted binocular development, as in the case of early-onset strabismus, inciting a dorsal light reflex; in humans, new vertical vergence movements of the eyes produce an inappropriate vertical divergence, with torsional characteristics. According to Guyton,⁷ patients with an early disruption of binocular fusion produce an imbalanced input to the vestibular system; this triggers latent nystagmus with a cyclovertical component, eliciting a normal but exaggerated oblique-muscle-produced cycloverversion/vertical vergence that appears to partially damp the cyclovertical nystagmus, thereby improving vision in the fixing eye while simultaneously producing an unavoidable elevation and extorsion of the fellow eye. Ten Tusscher and Van Rijn⁸ argued that DVD is the result of a supranuclear disorder of the vertical vergence system; their “corticotectal lateralization” hypothesis links the origin of DVD to an altered interaction of cortical and subcortical pathways. In infantile strabismus the disruption of

binocular fusion results in dissociated activation of subcortical pathways. Inhibition of one corticotectal pathway leads to elevation and cycloverversion of the ipsilateral eye.⁸

Dissociated horizontal deviations (DHD) is an uncommon spontaneous exodeviation assumed to have a similar pathophysiology as DVD and dissociated torsional deviation. There are no known prior reports on the prevalence of DHD. Brodsky reported that it is present in 50% of patients who develop consecutive exotropia following surgical correction of infantile esotropia.¹⁶ Postoperative patients were not included in the current study which may explain the complete absence of its occurrence in this cohort.¹⁶

The present study has several limitations. Individuals with a small angle of deviation may not have been noticed by the caretaker and thus not evaluated by the study ophthalmologists. Likewise, small or imperceptible amounts of DVD may not have been noticed by the ophthalmologist, further underestimating the true prevalence of DVD. Additionally, not all patients were evaluated 10 years after their initial diagnosis of strabismus, increasing the possibility that DVD was underdiagnosed in this cohort. Although it is true that DVD was not specifically noted within the medical records and therefore potentially not recorded; nevertheless, the majority of children in this study had multiple examinations, often by pediatric/strabismus specialists, and this study includes both the number found on initial examination and on the final examination for all patients. This study is further limited by the region in which it was performed. Because the ethnic composition of Olmsted County was 95% white during the years of the study, our results are best extrapolated to the semiurban white populations of the United States.

Literature Search

PubMed was searched, without date restriction, for English-language articles using the following search terms and combinations: *dissociated vertical deviation (DVD)*, *prevalence of DVD*, *dissociated horizontal deviation (DHD)*, *prevalence of DHD*; *DVD AND heterotropia* OR *esotropia* OR *exotropia* OR *Duane syndrome* OR *inferior oblique overaction*.

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Table 1
Prevalence of dissociated strabismus among children with ocular misalignment in Olmsted County, Minnesota, 1985-1994

Childhood strabismus subtype	Number	Number with DVD (%) at initial presentation	Mean age, years, of children with DVD at initial diagnosis	Mean age, years, of children w/o DVD at initial diagnosis	Number with DVD at initial and final	Number with DVD at final but not initial	Number with DVD (%) at final	Mean age, years, in children with DVD at final	Mean age, years, at final in children w/o DVD at final
Esotropia									
Accommodative	175	2 (1.1)	5.5	4.2		6	6 (3.4)	13.4	12.2
Acquired nonaccommodative	64			4.5		1	1 (1.6)	13.3	11.4
CNS	44	1 (2.2)	17.2	2.4	1	10	11 (25)	15.8	10.9
Congenital	30	2 (6.6)	0.4	0.6	2	14	16 (53.3)	11.2	7.4
Paralytic	25			3.8					10.6
Sensory	25			4.0		1	1 (4)	15.7	14.5
Other	4			13.1		1	1 (25)	24.8	18.2
Unknown	13			7.8		1	1 (7.7)	11.0	12.2
Exotropia									
CNS	30	1 (3.3)	2.2	6.4	1	2	3 (10)	12.6	12.9
Congenital	1			0.3		1	1 (100)	8.1	
Convergence insufficiency	40			11.5					15.9
Intermittent	106	1 (1.0)	9.6	6.4		2	2 (1.8)	12.9	12.2
Paralytic	8			7.6					15.4
Sensory	17	1 (5.8)	2.7	2.7		2	2 (11.7)	9.4	13.7
Other	3			14.7					15.3
Hypertropia									
CNS	5	1 (20.0)	1.0	5.9	1	1	2 (40)	16.7	16.8
Paralytic	13			7.3		1	1 (7.7)	11.3	13.8
Sensory	1			2.2					16.8
Other	16	1 (6.3)	2.1	8.0		1	1 (6.3)	7.5	12.7
Unknown	2			12.9					18.1
Isolated DVD	3	2 (66.7)	3.1	2.0	1		1 (33.3)	6.9	8.5
Comitant vertical deviation	2			4.3					11.7
Total	627	12			6	44	50		