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Adult-onset nonparalytic, small-angle hypertropia

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Abstract

Purpose—To describe the prevalence and clinical features of a common but underrecognized disorder of adult vertical strabismus.

Methods—The medical records of all adult (≥ 19 years of age) residents of Olmsted County, Minnesota, diagnosed with nonparalytic, small-angle hypertropia (NPSAH) from January 1, 1985, through December 31, 2004, were retrospectively reviewed for demographic and clinical features.

Results—Of 753 patients diagnosed with adult-onset strabismus, 99 (13.1%) were found to have NPSAH, yielding an annual incidence of 7.50 per 100,000 patients >18 years of age and a cumulative incidence of 1.28%. Diplopia was reported at the initial diagnosis in 91 patients (93.8%), with 90 (92.8%) having the diplopia in primary or reading position. The median initial angle of hypertropia was 2 (range, 1–22) at near and 2 (range, 0–12) at distance. After a median follow-up of 10.8 years (range, 6.2 months to 23.7 years), the final median angle of vertical deviation was 4 (range, 0–20) at near and 4 (range, 0–16) at distance for all 99 patients.

Conclusions—NPSAH is a relatively common but infrequently recognized disorder among adults. More prevalent among elderly and female patients in this study cohort, the vast majority presented with diplopia and a hypertropia of $\leq 10^\circ$ that progressed over time.

Adult strabismus is a relatively common disorder.^{1,2} Although the demographics and clinical characteristics of childhood strabismus have been well-described,^{3–7} less is known concerning adult-onset strabismus. Existing studies of ocular misalignment among adults are primarily retrospective reports of patients who have undergone surgery and do not include medically managed cases or the more rare or unusual forms of strabismus.^{2,8–11} Only recently have reports^{1,12} provided a more accurate representation of the various forms of

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adult strabismus. Although paralytic strabismus has been shown to be the most prevalent form among adults, we recently reported that nonparalytic, small-angle hypertropia comprised 13.1% of all forms of adult-onset strabismus in a population-based cohort.¹² The purpose of the current study was to describe the clinical characteristics and early natural history of adults diagnosed with this form of hypertropia over a 20-year period.

Subjects and Methods

This study was approved by the Mayo Clinic and Olmsted Medical Group Institutional Review Board. The medical records of all patients at least 19 years of age who were newly diagnosed with a nonparalytic, small-angle hypertropia from January 1, 1985, through December 31, 2004, while residing in Olmsted County, Minnesota, were retrospectively reviewed. Cases of adult hypertropia were identified using the Rochester Epidemiology Project (REP) database, a medical record linkage system that combines all patient-physician encounters in Olmsted County, Minnesota.¹³ Due to the relatively isolated nature of this county from other metropolitan areas, the Mayo Clinic and Olmsted Medical Group and their affiliated hospitals are responsible for providing virtually all of the medical care to its residents.

Potential cases of new-onset adult strabismus were verified by searching the REP database for International Classification of Diseases, 9th edition, codes for strabismus and other disorders of binocular eye movements. The various forms of adult strabismus observed in this population have previously been reported.¹² Nonparalytic, small-angle hypertropia was defined in this study as either a comitant hypertropia or vertical diplopia in patients who had no signs of trochlear nerve palsy and were managed using prisms of $\leq 3^\circ$. The medical records of all patients were reviewed carefully for oblique muscle dysfunction and any description of fundus torsion to specifically exclude a small- or long-standing trochlear nerve palsy. Other forms of vertical strabismus, including those associated with myasthenia gravis, thyroid eye disease, childhood onset, or due to a neurologic or sensory etiology, were also excluded from the study. Ocular alignment was measured by a number of methods, including Maddox rod, alternate prism and cover test, and Lancaster red-green test. Patients also routinely underwent a measurement of visual acuity, ocular rotations, and assessment of the anterior and posterior segments.

The incidence rate of adult-onset nonparalytic, small-angle hypertropia was estimated using the age- and gender-specific population figures for Olmsted County, Minnesota, census data for 1985 through 2004. The 95% confidence interval for the incidence rate was calculated assuming a Poisson error distribution. The rate of progression of hypertropia was calculated using the Kaplan-Meier method. All of the statistical tests performed were two-sided and an α of 0.05 was set as the threshold of significance.

Results

Nonparalytic, small-angle hypertropia was diagnosed in 99 (13.1%) of the 753 strabismic adults over the 20-year period, yielding an annual age- and gender-adjusted incidence of 7.50 (95% CI, 6.01–8.99) per 100,000 residents >19 years of age, and a cumulative

incidence of 1.28 per 100 for the adults included in this study. The median age at diagnosis was 71 years (range, 27–98 years), with a significant increase in incidence with increasing age ($P < 0.05$). There were 63 (64%) women in the study population.

The presenting ophthalmic findings of the 99 patients are shown in Table 1. Of 97 queried patients, 91 (94%) were found to have diplopia on their initial examination: 77 (79%) in primary gaze, 13 (13%) in reading gaze, and 1 (1%) in peripheral or side gaze. The median angle of hypertropia at vertical distance fixation was 2° at near (range, 1°–22°) and 2° (range, 0°–12°) at distance. Only 3 patients had an initial vertical deviation of at least 11°. Fifty-one patients (51%) also had a mean horizontal phoria of 5.4° (range, 2°–15°). Torticollis was present in 4 patients (4%), and the mean stereoacuity was 40 arcsec (range, 0–400 arcsec). The most prevalent associated systemic condition was hypertension, in approximately one-third of patients.

The 99 patients were followed for a median of 10.8 years (range, 6.2 months to 23.7 years), with only 1 patient having follow-up of <1 year. Eighty-four patients (84.8%) received prisms in their spectacles at some time during the follow-up period, and 1 patient underwent surgery for correction of a vertical deviation of 3°. The final median angle of hypertropia at vertical distance fixation for the 99 patients was 4° (range, 0°–20°) at near and 4° (range, 0°–16°) at distance, a mean increase of 2° from the initial evaluation.

Discussion

Nonparalytic, small-angle hypertropia (NPSAH) comprised 13.1% of patients in this population-based cohort of adult-onset strabismus. The mean age at diagnosis occurred in the seventh decade of life, and two-thirds of the cohort were female. Most patients were diplopic, with a small angle of deviation that showed a progressive increase over time. The use of prisms in spectacles was the most common form of therapy in this cohort of patients.¹⁴

There are few published studies with which to compare the findings of this report. The relatively common occurrence of the specific form of hypertropia observed in this cohort has not been previously reported. Prior studies of adult strabismus are comprised almost exclusively of postoperative outcomes, with most studies having no patients with hypertropia.^{2,9–11} In a systematic review of Medicare patients diagnosed with strabismus, Repka and colleagues¹ observed that 19% had some form of hypertropia. Similarly, El-Sahn and colleagues¹⁵ reported the prevalence of vertical deviation alone to be 18.9% in a cohort of strabismic patients >60 years of age. However, neither of these reports provide clinical characteristics or distinguish NPSAH from other forms of hypertropia. Several studies^{16–19} describing the evaluation of certain forms of adult-onset strabismus include vertical strabismus, but without extensive clinical description. A recent study on the etiology of hypertropia in 300 adults reported that none had NPSAH²⁰; however, 9 of the study patients (3%) were diagnosed with decompensated hypertropia. Although difficult to make this diagnosis without observing decompensation, it is possible that some or all of these patients had NPSAH. Regardless, their observed decompensated hypertropia prevalence of 3%

among all forms of adult hypertropias is significantly lower than the 13.1% prevalence of NPSAH diagnosed among all forms of new-onset adult strabismus in the present study.¹²

Although nonparalytic hypertropia occurs relatively commonly in elderly adults, it appears to be underrecognized and often overlooked. Because the deviation is initially small or intermittent and readily controlled, patients may not seek care immediately or may be unable to properly express their symptoms. When finally examined by an eye care professional, generally a nonstrabismus specialist, the patient may report having diplopia or, often, difficulty seeing. The unfamiliar examiner may not perform an alternate cover or similar test, further postponing the diagnosis. Frustrations and concerns about symptoms may persist until the patient is appropriately evaluated and diagnosed. The small angle of deviation and the delayed or unrecognized diagnosis are likely to have contributed to its absence in the scientific literature. An increased awareness and earlier identification of this form of adult strabismus would enhance patient satisfaction and outcomes.

Several theories have been proposed for the occurrence of nonparalytic hyperdeviations during adulthood. Chaudhari and Demer¹⁹ suggested that such a deviation may be a result of “sagging eye syndrome,” which is characterized by the asymmetric stretching of the lateral rectus–superior rectus band ligament and leads to inferior sag of the lateral rectus pulley, thus causing the acquired diplopia reported in their study’s hypertropic and esotropic patients.¹⁹ This uneven stretching and sag can be explained by aging-related degeneration of the orbital connective tissue.¹⁸ Others have proposed that some cases of adult hypertropia may be due to comitant skew deviations as a result of microvascular ischemia–related lesions in the cerebellum, brainstem, or the vestibular system.²¹ A third explanation for at least some of these patients is retinal diplopia in which epiretinal membrane wrinkling leads to displacement of one macula relative to the other, resulting in diplopia.²² The wrinkling appears to be due to traction at the vitreoretinal interface, caused by disorders such as ocular histoplasmosis, macular degeneration, and diabetic retinopathy. Among the various causes for epiretinal membrane–related diplopia (ie, retinal misregistration, refractive error, or strabismus), those with strabismic retinal diplopia may be identified by testing negative on the tests for retinal misregistration, or experiencing a resolution of diplopia with the use of prisms or surgery, similar to the patients in this study.²³ All three entities occur almost exclusively in older individuals, consistent with the observation that nonparalytic hypertropia increases in incidence with increasing age.¹²

There are a number of limitations to the findings of this study. Given the relatively small deviations and seemingly unrecognized nature of this form of strabismus, it is likely that the prevalence is underestimated in the study population. Moreover, some private optometrists working in Olmsted County are not involved in the REP database. Although many cases assessed by local optometrists are referred to one of the two medical institutions associated with the REP for higher-level care, the symptoms of some adults may not have been severe enough to seek such care. This study is also limited by the frequent use of the Maddox rod to assess ocular alignment. Although the Maddox Rod is inferior to the alternate cover and prism test for assessing strabismus, most patients were examined on multiple visits for their symptoms of vertical diplopia. Finally, given the limited racial and ethnic composition of

Olmsted County, our results are best extrapolated to the small, urban, white populations of the United States.

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Table 1.

Presenting ophthalmic features of 99 patients with nonparalytic hypertropia

Characteristic	Result
Median age at diagnosis, years (range)	71 (27–98)
Sex, no. (%)	
Male	63 (63.6)
Female	36 (36.4)
Patients with diplopia, no. (%)	
Primary gaze	77 (79.4)
Reading gaze	13 (13.4)
Peripheral gaze	1 (1.0)
Median vertical alignment at diagnosis, PD	
Near (range) [N = 92]	2 (1–22)
Distance (range) [N = 82]	2 (0–12)
Type of deviation at diagnosis, no. (%)	
Right hypertropia	51 (51.5)
Left hypertropia	48 (48.5)
Patients with reported systemic conditions, no. (%)	
Hypertension	35 (35.4)
Diabetes	3 (3.0)
Cerebrovascular disease	4 (4.0)

PD, prism diopter.