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Exotropia in children with high hyperopia

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Abstract

Purpose—To describe the clinical characteristics, treatment, and subsequent clinical course of children with exotropia and high hyperopia.

Methods—The medical records of 26 patients seen between 1990 and 2009 who had an exotropia and 4.00 D of hyperopia were retrospectively reviewed. We analyzed the clinical characteristics, treatments and subsequent alignment outcomes.

Results—A total of 26 patients between the ages of 2.5 months and 9 years were included. Of these, 15 had associated medical conditions or developmental delay. Of 22 patients with measured visual acuities, 19 had amblyopia (10 unilateral, 9 bilateral). None of the patients demonstrated fine stereoacuity. Twenty-three exotropic children were treated with spectacles; 15 were fully corrected, 10 of whose exotropia improved; 8 received partial correction of their hyperopia, 3 of whose exotropia improved. Six patients who presented with large, poorly controlled exotropia and did not improve with spectacle correction required strabismus surgery.

Conclusions—Children with high hyperopia and exotropia are likely to have developmental delay or other systemic diseases, amblyopia, and poor stereopsis. Treatment of high hyperopia in exotropic children with their full cycloplegic refraction can result in excellent alignment.

Children with hyperopia >3.50 D of spherical equivalent are at a risk for refractive amblyopia.¹ Glasses are usually prescribed to allow for normal visual development. Most children who have hyperopia >4 D are either orthotropic or have an esotropia.¹ Rarely, do children with 4 D of hyperopia present with an exotropia.²

Spectacle correction in children with hyperopia and exotropia can decrease accommodative demand, which can potentially worsen the exotropia. Low symmetric amounts of hyperopia in association with an exotropia usually do not require spectacle correction. With amblyogenic degrees of hyperopia, however, spectacle correction is necessary to permit normal visual development. Partial or full spectacle correction may be prescribed, depending on the ophthalmologist's preference. One study has shown that full spectacle correction of

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Study conducted at the North Shore–Long Island Jewish Health System.

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high hyperopia in exotropic patients can improve the exotropia.² The purpose of this study was to better understand the benefit of fully treating the high hyperopia in patients with an exotropia.

Subjects and Methods

The medical records of all patients seen by two of the authors (SK and SR) at the North Shore–Long Island Jewish Health System Department of Ophthalmology between January 1990 and December 2009 who had at least 10^Δ of exotropia and at least 4 D of hyperopia (spherical equivalent) in one or both eyes by cycloplegic retinoscopy were retrospectively reviewed. The exotropia was measured in prism diopters and could be either intermittent or constant. If the exotropia was intermittent, the control was classified as good, fair, or poor. Control was recorded as “good” if the patient became exotropic only after cover testing and rapidly returned to orthotropia with a blink; “fair” if the patient became exotropic with cover testing and returned to orthotropia with multiple blinks; “poor” if the patient had a spontaneous exotropia or did not become orthotropic with multiple blinks. Patients could also have coexisting vertical deviations.

Children who underwent previous strabismus surgery were excluded. All patients had complete ophthalmologic evaluations, including cycloplegic refractions with 1% tropicamide, 2.5% phenylephrine, and 1% cyclopentolate. Visual acuity was measured using fixation preference in preverbal children or by age-appropriate recognition optotypes, such as Snellen letters, Allen pictures, HOTV letters, or the E game. A patient was considered to have amblyopia if their best-corrected visual acuity was <20/40 (below the age of 5 years) or <20/30 (if 5 years or older) or if there was more than a one line difference in acuity at any age. If only fixation preference was used to determine vision, a patient was considered to have amblyopia if the patient was unable to maintain fixation through a blink. When possible, patients were also tested for stereoacuity using the Titmus test (Stereo Optical, Chicago, IL). Patients were divided into three treatment categories: full hyperopic correction (within 1 D of cycloplegic refraction), partial correction (cycloplegic refraction minus more than 1 D of hyperopia), and no correction (observation or patching). Resolution of exotropia was defined as orthotropia with or without an exophoria. Improvement was defined as a minimum of 5^Δ of improvement in the exotropia or an improvement in the control of an intermittent exotropia. Conversely, worsening of exotropia was defined as an increase of 5^Δ or more or a decreased control of an intermittent exotropia.

Approval from the Institutional Review Board from the North Shore–Long Island Jewish Health System was obtained. This study also complied with Health Insurance Portability and Accountability Act regulations. Informed consent was not required by the Institutional Review Board.

Results

A total of 26 patients (11 males) met inclusion criteria. Patient age at the first ophthalmological examination ranged from 2.5 months to 9 years (average, 4 years). This does not reflect the age of onset of symptoms. Patients with follow-up were followed for an average of 4.2 years (range, 6 months to 13.25 years). The average spherical equivalent of hyperopia was 6.33 D (range, 3.50–10.00 D).

Of the 26 children, 23 had an intermittent exotropia, and 3 had a constant exotropia. Of the children with a constant exotropia, 1 patient had a congenital exotropia diagnosed at 4 months of age. The average deviation was 23^Δ of exotropia (range, 10^Δ–45^Δ). Ten patients had a basic type of exotropia, with a difference of <10^Δ between distance and near. Six

patients had convergence insufficiency, with near deviation greater than distance of at least 10^{Δ} . Five patients had divergence excess, with distance deviation greater than near of at least 10^{Δ} . We were unable to quantify both distance and near deviations in 2 patients because of variability of the deviation in one and inability to fixate at distance (at 6 months of age) in the other. We did not differentiate between true and pseudodivergence excess because patch testing was not performed in all patients.

Fifteen patients in this study had developmental delay (11) or other associated medical conditions (7), including eyelid hemangioma, periventricular leukomalacia, VATER syndrome, Treacher-Collins Syndrome, Jacobsen syndrome, Down syndrome, and macrocephaly.

In 22 of our patients, we were able to assess visual acuity either by optotypes or fixation preference testing; for 4 patients we were unable to document a fixation preference because of difficulty with the examination. Of the 22 patients, 19 (86%) had amblyopia as defined previously. In 10 children, amblyopia was unilateral; in 9 bilateral, with amblyopia worse in one eye than the other in 3 children. Only 3 of 22 patients had vision measured with optotypes that was not defined as amblyopia.

Binocular function with near stereoacuity was ascertainable in 17 of 26 children. Of the 17 patients, 4 had no measurable stereoacuity, 3 had only 3000 arcsec, 5 had 400–800 arcsec, and 5 had 70–200 arcsec. No patients had fine stereoacuity (less than 70 seconds) at time of initial therapy.

Of the 26 patients, 6 children were already wearing spectacles at the initial visit; 18 were untreated though with known exotropia. Two patients presented with no known deviation but developed a new intermittent exotropia when prescribed their full hyperopic correction. Decreasing the power of the hyperopic prescription resolved the exotropia in 1 patient and improved the exotropia in the other. The patient with partial resolution of the exotropia eventually worsened and required strabismus surgery. Of the 18 children with no prior treatment and a known exotropia prior to spectacle correction, 15 children received and complied with spectacles. One child had part-time patching therapy without spectacles and 2 children were lost to follow-up. Of the 6 patients that presented to our service already wearing spectacles with an exotropia, 1 required surgery and 1 required surgery but was lost to follow-up.

Of the 26 children, 23 were treated with spectacles (Table 1). Of these, 15 received their full cycloplegic correction and 11 received partial correction. Of the 15, 10 (67%) with full hyperopic correction experienced improvement or resolution of the exotropia; 2 (13%) experienced no improvement. The average exotropic deviation in patients corrected with full hyperopic correction was 26^{Δ} before and 13.5^{Δ} after spectacle correction. Of the 8 children who received partial hyperopic correction, 3 (38%) had improvement or resolution in the exotropia; 5 patients (63%) had either no improvement or worsening of their exotropia. The average deviation of all patients with partial hyperopia correction was 21^{Δ} before and 16^{Δ} after spectacle correction. The 6 patients with complete resolution of exotropia with partial or full hyperopic correction had an average of 17^{Δ} of exotropia with good to excellent control of their exotropia prior to spectacle correction.

Of our 26 patients, 5 eventually underwent strabismus surgery, with the surgical planning based on preoperative measurements with correction. Of these 5 patients, 1 had a congenital exotropia; 4 had large poorly controlled exotropia that did not improve with spectacle correction or worsened over time. The patients who had strabismus surgery had an average exotropic deviation of 31^{Δ} (range, 20^{Δ} – 45^{Δ}) compared with the average exotropia of 21^{Δ} for the 21 patients who did not undergo surgery. All 5 patients that underwent strabismus

surgery developed an accommodative esotropia postoperatively that was controlled with their hyperopic spectacle correction.

Discussion

Our case series suggests that developmental delay or other associated medical conditions may occur frequently in children with exotropia and high hyperopia. According to Atkinson and Braddick,³ the incidence of hyperopia of at least 3.50 D in the general population is only 5% between the age of 6-9 months, with the incidence decreasing on follow-up. Nielsen and colleagues⁴ showed a higher incidence of high levels of hyperopia in children with developmental delay, with >3 D spherical equivalent in 15.3% of children with developmental delay and an even greater incidence of 21.8% in children with IQs <50. The higher prevalence of developmental delay in our patient series may be due in part to the fact that our study population is from a tertiary care center.

There was also a high incidence of amblyopia in our population compared to other children with high hyperopia. Colburn and colleagues¹ reported only 19% of patients with at least 3.75 D of hyperopia had amblyopia at presentation, and 38% of these patients without spectacle correction eventually developed amblyopia. By comparison, the incidence of amblyopia in our series was 86%. Given that the vast majority of our patients had intermittent exotropia (88%), one might expect the incidence of amblyopia to be similar to that of patients with intermittent exotropia. In fact, our patients also had a much higher incidence of amblyopia (85%) than seen in a retrospective review of patients with intermittent exotropia, where more than 98% of patients had a visual acuity of > 20/40.⁵ This may be related to the high incidence of developmental delay in our study population.

The level of stereoacuity is also relatively impaired compared to what would be expected in intermittent exotropia. Morrison and colleagues⁶ reported that 76.8% of their patients had better than 200 arcsec versus 29.4% of our patients; none of our patients demonstrated better than 70 arcsec, compared to 21.1% of their study patients.

The increased incidence of bilateral amblyopia and/or reduced binocularity in our study population may be responsible for the reduced accommodative drive in these patients. Maheshwari and colleagues⁷ have shown that amblyopic eyes have reduced accommodation. This reduction in accommodation associated with amblyopia may allow an exotropia to occur instead of an accommodative esotropia. In the children who responded to their full hyperopic spectacles, we theorize that by treating the amblyopia with full hyperopic correction, our children had a clearer retinal image, improved visual acuity, and accommodation, thereby reducing the exotropia. Full hyperopic correction may have been associated with an improvement in control and size of the exotropia compared to partial hyperopic correction.

All 5 patients who underwent strabismus surgery developed an accommodative esotropia postoperatively that was controlled with glasses. All patients had preoperative measurements with correction but not without correction due to their poor uncorrected visual acuity. Preoperatively, however, most caretakers reported that the exotropia was worse with the glasses than without the glasses, suggesting that the patients would develop an accommodative esotropia postoperatively. It is unclear why the exotropia did not respond to the hyperopic correction in these patients or why the strabismus appeared worse with spectacles. This may be related to the fact that these patients had a larger and poorer controlled exotropia at their initial visit.

There are some limitations to our study. First, a tertiary facility may select for a higher incidence of children with developmental delays and other medical conditions. Second, we

included all patients with exotropia whether constant or intermittent, although the vast majority of the patients had intermittent exotropia. Because this was a retrospective study, the patients were not randomly assigned to receive either full or partial correction of their hyperopic prescription. It is possible that control and size of the exotropia would influence the hyperopic prescription given by the physicians. However, the patients were divided fairly evenly between two practices, with one ophthalmologist almost exclusively prescribing within 1 D of the full cycloplegic refraction and the other almost exclusively prescribing much less than the full cycloplegic refraction. Another limitation was lack of consistency in the reduction of the hyperopia prescribed in those patients given partial hyperopic correction. Finally, the sample size of our population was small and statistical analysis could not confirm the possibility of improved alignment with full cycloplegic refraction compared to partial cycloplegic refraction. Many of these issues would be resolved with a prospective randomized study with a larger number of patients in order to achieve statistical significance.

In conclusion, patients with high hyperopia and exotropia were more likely to have developmental delay, amblyopia, and poor binocular function. The children whose exotropia improved the most with the prescribed hyperopic prescription presented with smaller deviations and good control of the intermittent exotropia. Full hyperopic correction may result in a better outcome than partial hyperopic correction, although further larger studies are necessary to confirm this finding. Poor alignment with need for strabismus surgery was more common with initial large angle, poorly controlled exotropia, and poor response to spectacles.

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Table 1Patient outcomes with high hyperopia and exotropia^a

Treatment	N	Resolution (%)	Improvement (%)	No change (%)	Worse (%)	Lost or Noncompliant (%)
Spectacles	23					
Full	15	5 (33)	5 (33)	2 (13)	0	3 (20)
Partial	8	1 (12.5)	2 (25)	3 (37.5)	2 (25)	0
Observation only	1	0	1 (100)	0	0	0
Decreased correction in spectacle-induced exotropia	2	1 (50)	0	0	1 (50)	0

^aPercentages are for outcomes within a treatment category.