Exotropia in Children with High Hyperopia

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Abstract:

Introduction: This study describes the uncommon association of exotropia in children with high hyperopia.

Methods: We retrospectively reviewed the charts of 26 patients seen by our pediatric ophthalmology service between 1990 and 2009 who had an exotropia and 4.00 or more diopters of hyperopia. We analyzed the characteristics of our patient population as well as alignment outcomes with full or partial hyperopic correction.

Results: Twenty-six patients between the ages of 2.5 months and 9 years met study criteria. Fifteen patients in this study had associated medical conditions or developmental delay. Nineteen of 22 patients with measured visual acuities had amblyopia, 10 of which were unilateral and 9 bilateral. Patients also had poor stereopsis, with none demonstrating fine stereoacuity. Twenty-three exotropic children were treated with spectacles. Fifteen children received their full cycloplegic refraction and 10 of these children had improvement in their exotropia. Eight children received partial correction of their hyperopia and only 3 had improvement in their exotropia. Six of 26 patients required strabismus surgery and presented with large poorly-controlled exotropia or no improvement with spectacle correction.

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. Poor alignment with need for strabismus surgery was associated with an initial large angle, poorly controlled exotropia, and poor response to spectacles.

Introduction:

Children with hyperopia greater than 3.50 diopters 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 greater than 4 diopters are either orthotropic or have an esotropia[1] . Rarely, a child with greater than 4 diopters or greater of hyperopia may present with an exotropia[2].

The use of spectacles in children with hyperopia and exotopia contrasts two opposing approaches to visual development and ocular alignment. 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 exotopia usually do not require spectacle correction. With amblyogenic degrees of hyperopia, however, spectacle correction is necessary to allow for 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 high hyperopia in exotropic patients can improve the exotropia.[2].

In an effort to better understand patients with high hyperopia and exotropia we retrospectively reviewed patient characteristics and treatment outcomes of these patients in our service within the past 20 years.

Subjects and Methods:

We retrospectively reviewed the charts of all patients seen by Doctors Sylvia Kodsi and Steven Rubin at the North Shore-Long Island Jewish Health System Department of Ophthalmology between January 1990 and December 2009 who had at least 10 prism diopters of exotropia and at least 4 diopters of hyperopia (spherical equivalent) in one or both eyes by cycloplegic retinoscopy. The exotropia was measured in prism diopters and could be either intermittent with the patient orthotropic at times, or constant. If the exotropia was intermittent, the control was classified as good, fair, or poor. Good control was recorded if the patient became exotropic only after cover testing and rapidly returned to orthotropia with a blink. Fair control was documented if they became exotropic with cover

testing and returned to orthotropia with multiple blinks. Poor control was recorded when a patient had a spontaneous exotropia, or who did not become orthotropic with multiple blinks. The patients could also have coexisiting 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 worse than 20/40 (below the age of five) or worse than 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 preferrence 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 diopter of cycloplegic refraction), partial correction (cycloplegic refraction minus more than 1 diopter 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 prism diopters 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 prism diopters 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 because of the deidentification of patient characteristics in this study.

Results:

Twenty-six patients met our study criteria. Eleven children were male and 15 female. The age of the first eye examination in our office ranged from 2.5 months to 9 years with an average of 4 years old. This does not however reflect the age of onset of symptoms. Patients with follow-up were followed for an average of 4.2 years with a range of 6 months to 13.25 years. The average spherical eqivalent of hyperopia was 6.33 diopters with a range of 3.50 to 10.00 diopters.

Twenty three children had an intermittent exotropia and 3 patients had a constant exotropia. Of the children with a constant exotropia, one patient had a congenital exotropia diagnosed at 4 months of age. The average deviation was 23 prism diopters of exotropia with a range from 10 to 45 prism diopters. Ten patients had a basic type of exotropia with less than a 10 prism diopter difference at distance and near. Six patients had convergence insufficiency with near deviation greater than distance of at least 10 prism diopters. Five patients had divergence excess with distance deviation greater than near of at least 10 prism diopters. We were unable to quantify both distance and near deviation in two 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 since patch testing was not performed in all these patients.

Fifteen patients in this study had developmental delay or other associated medical conditions. Eleven patients were diagnosed with developmental delay. Other associated medical conditions included eyelid hemangioma, periventricular leukomalacia, VATER syndrome, Treacher-Collins Syndrome, Jacobsen syndrome, Down syndrome, and macrocephaly.

In 22 of our 26 patients, we were able to obtain a vision either by optotypes or fixation preference testing. The other four patients did not have a fixation preference documented because of difficulty with the examination from developmental delay. Of the 22 patients, 19 (86%) had amblyopia as defined previously and 10 of these children had unilateral amblyopia. Nine of the 19 children had bilateral amblyopia with 3 of these 9 children having worse amblyopia in one eye than the other. Only 3 of 22 patients had vision measured with optopypes that was not defined as amblyopia.

We were able to measure binocular function with near stereoacuity in 17 of our 26 patients. Of the 17 patients, 4 had no measurable stereoacuity, 3 had only 3000 seconds of arc, 5 patients had 400-800 seconds of arc, and 5 had 70-200 seconds of arc. No patients had any fine stereoacuity (less than 70 seconds) at time of initial therapy.

Of the 26 patients, 6 children were already wearing glasses. Eighteen of the 20 children with no prior treatment had a 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 one patient and improved the exotropia in the other patient. 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 two children were lost to follow-up. Of the 6 patients that presented to our service already wearing spectacles with an exotropia, one patient required surgery and one additional patient needed surgery but was lost to follow-up.

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

Five of our 26 patients eventually underwent strabismus surgery with the surgical planning based on preoperative measurements with correction. One of these 5 patients had a congenital exotropia. The 4 other patients 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 prism diopters (range 20 to 45 prism diopters) compared with the average exotropia of 21 prism diopters for the 21 patients who did not have surgery. All 5 patients that had strabismus surgery for exotropia developed an accommodative esotropia postoperatively which was controlled with their hyperopic spectacle correction.

Discussion:

Our case series suggests that developmental delay or other associated medical conditions may be more common in children with exotropia and high hyperopia. One study showed the incidence of hyperopia of at least 3.50 diopters in the general population to be only 5% between the age of 6-9 months, with the incidence decreasing on follow-up[3]. Nielsen and colleagues showed a higher incidence of high levels of hyperopia in children with developmental delay. He found more than 3 diopters spherical equivalent in 15.3% of children with developmental delay and an even greater incidence of 21.8% in children with IQs less than 50[4]. Alternatively, the higher prevalence of developmental delay in our patient series may also 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. In a study by Colburn and colleagues, only 19% of patients with at least 3.75 diopters of hyperopia had amblyopia at presentation, and 38% of these patients without spectacle correction eventually developed amblyopia[1]. In comparison, our patients had an incidence of amblyopia of 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 intermittent exotropes. 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 vision better than 20/40[5]. This may be related to the high incidence of developmental delay in our study population.

Similarly, the level of stereoacuity is also relatively impaired compared to what would be expected in intermittent exotropia. In a study by Morrison and colleagues, 76.8% of their patients had better than 200 sec of arc versus 29.4% of our patients. None of our patients demonstrated better than 70 sec of arc compared to 21.1% of their study patients[6].

The increased incidence of bilateral amblyopia and/or reduced binocularity in this population may be responsible for the reduced accommodative drive in these patients. Maheshwari and colleagues have shown that amblyopic eyes have reduced accommodation[7]. This reduction in accommodation associated with amblyopia may allow for 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 acuityand accommodation, thereby reducing the exotropia. Although not statistically significant, prescribing the full hyperopic correction was possibly associated with an improvement in control and size of the exotropia compared to partial hyperopic correction. .

For the 5 patients who had strabismus surgery, all 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 vision without correction. Preoperatively, however, most caretakers noticed 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, the study was from a tertiary facility which may select for a higher incidence of children with developmental delays and other medical conditions.

Second, we included all patients with exotropia whether it was constant or intermittent, although the vast majority of the patients were intermittent exotropes. Since this was a restrospective 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 physician practices with one physician almost exclusively prescribing within one diopter of the full cycloplegic refraction while the other almost exclusively prescribed 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 giving partial cycloplegicrefraction. 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 summary, patients with high hyperopia and exotropia in our study population 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 possibly have 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.

TABLE 1: Patient Outcomes

Patients with high hyperopia and exotropia were separated into three categories: Spectacle correction (either full or partial cycloplegic refraction), observation only, or reducting hyperopic spectacle correction due to an exotropia that developed only after the child was put in hyperopic spectacles. Percentages are for outcomes within a treatment category.

Treatment	N	Resolution (%)	Improvement	No change	Worse	Lost or Non-compliant
Spectacles	23					
Full	15	5 (33.3)	5 (33.3)	2 (13.3)	0 (0)	3 (20)
Partial	8	1 (12.5)	2 (25)	3 (37.5)	2 (25)	0 (0)
Observation only	1	0 (0)	1 (100)	0 (0)	0 (0)	0 (0)
Decreased correction in spectacle-induced exotropia	2	1 (50)	1 (50)	0 (0)	0 (0)	0 (0)

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