Article ▸ A Stepwise Approach to Vision Therapy in Nystagmus with Strabismus

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ABSTRACT

Background: Nystagmus is an involuntary movement of the eyes. Infantile Nystagmus Syndrome has a prevalence of 14 per 10,000. While there is no current widely accepted treatment option, vision therapy has been shown to dampen the nystagmus and improve patients' quality of life.

Case Report: A 2-year-old Caucasian female presented with a constant left esotropia (post-strabismus surgery) along with latent nystagmus in her left eye, ultimately contributing to moderate amblyopia and suppression in the left eye (best visual acuity 20/40 OD, 20/100 OS with a right head turn). Treatment progressed in a stepwise approach, beginning with focusing on improving monocular visual acuities, followed by improving motor alignment, then adding prism, and finally breaking suppression, with the goal of improving the patient's vision, binocular system, and postural abnormalities secondary to a constant head turn.

Discussion: Many children with infantile nystagmus have a co-related strabismus or other binocular vision disorder. By explaining the advantages of vision therapy to the patient and their parent and setting forth the understanding of a lengthy therapy program, the patient can reap huge benefits by improving their visual function.

Keywords: infantile nystagmus syndrome, null point, vision therapy, yoked prism

Introduction

Nystagmus is a condition in which involuntary movement of the eyes occurs, and varies widely in its presentation of visual sensory deficits.[1] Symptoms of infantile nystagmus include decreased vision, anomalous head posture, poor visual tracking, oscillopsia, accommodative deficiencies, and photophobia/light interference.[2] While nystagmus is a chronic condition that many infants and children face, it is not commonly discussed in the literature and research. This may possibly be due to the little we understand and the difficulty with treating the condition, as compared to strabismus and other childhood eye conditions. Recently, however, the National Eye Institute supported a new classification system for over two dozen types of nystagmus called the Classification of Eye Movement Abnormalities and Strabismus (CEMAS)[2] (Table 1). One of the major changes in nomenclature includes the change of terminology from Congenital Nystagmus to Infantile Nystagmus Syndrome, with differentiations based off advances in eye movement recording technology on infants and children with nystagmus. For the purpose of this report, only the three classifications of nystagmus with an infantile onset will be discussed.

Infantile Nystagmus Syndrome (INS) (previously Congenital Nystagmus or "Motor and Sensory" Nystagmus) is characterized by the oculomotor recordings showing diagnostic (accelerating) slow phases. The eye movements are usually conjugate and horizontal with a small torsional component. Common associated findings include defects of the visual sensory system (e.g., albinism, achromatopsia, and congenital cataracts), a positive family history, and associated strabismus or refractive error. The nystagmus may decrease with convergence, with a null or neutral zone present. The patient often exhibits an associated head posture or head shaking used to maintain the eyes in a null position. Waveforms may change in early infancy, with a head posture usually evident by 4 years of age. The vision prognosis depends on the integrity of the sensory system.[2]

Table 1. CEMAS Classification of Nystagmus

1. Peripheral Vestibular Imbalance Meniere, Drug toxicity
2. Central Vestibular Imbalance Downbeat, Upbeat, Drug toxicity
3. Instability of Vestibular Mchanisms Periodic Alternating Nystagmus
4. Disorders of Visual Fixation Vision loss, See-saw nystagmus, Drug toxicity
5. Disorders of Gaze Holding Gaze evoked, Acquired Pendular, Drug toxicity
6. Acquired Pedular Nystagmus Central myelin, Oculopalatal, Whipple, Drug toxicity
7. Saccadic Intrusions and Oscillations Square wave jerks, Macro-saccadic oscillations, Opsoclunus, Flutter pulses
8. Miscellaneous Eye Movements Superior oblique myokymia, Ocular motor neuromyotonia
9. Infantile Nystagmus Syndrome "Congenital," "Motor," "Sensory," Idiopathic, Nystagmus blockage
10. Fusion Maldevelopment Nystagmus Syndrome Old "latent, manifest lantent," Nystagmus blockage
11. Spasmus Nutans Syndrome Without optic pathway glioma With optic pathway glioma

Table 2. Distance Visual Acuities Across Progress Exams

Date	Age	Patching Method (OD)	Distance Visual Acuities		Method of Assessing VA
			OD	OS	
02/01/11	24 months	Full-occlusion 2hrs/day	20/50	20/100	Preferential viewing paddles
03/08/11	25 months	Bangerter Filter 2-3hrs/day	20/40	20/40	Preferential viewing paddles
04/12/11	26 months	No patching	20/40	20/125+	Lea symbols matching
06/07/11	27 months	Bangerter Filter 2-3hrs/day	20/30	20/50	Not specified
01/10/12	36 months	No patching	20/25	20/80	Lea symbols matching
06/26/12	41 months	Bangerter Filter 2-3hrs/day	20/20	20/40	Lea symbols
09/18/12	44 months	Atropine OD e.o.d.	20/20	20/20	Snellen single letters
10/16/12	45 months	Atropine OD e.o.d.	20/40	20/40	Snellen single letters
12/04/12	47 months	Atropine OD e.o.d	20/40	20/50	Lea symbols
01/28/13	5 years	Atropine OD e.o.d	20/30+2	20/50+	Lea symbols

Fusion Maldevelopment Nystagmus Syndrome (FMNS) (previously Latent/Manifest Latent Nystagmus) is a benign jerk nystagmus that is most evident under monocular conditions and has an associated strabismus, most commonly esotropia. The ocular motor recordings are the sole way to differentiate FMNS from INS, where FMNS is characterized by decelerating or linear slow phases toward the covered eye and a fast phase jerk in the direction of the fixating eye. Since patients with esotropia usually suppress one eye, the nystagmus is present without covering an eye. The nystagmus may dampen with adduction, so the patient may present with a head turn in the direction of the fixating eye. Visual acuity results are often much better under binocular versus monocular conditions. The intensity may decrease with age.[2]

Spasmus Nutans Syndrome (SNS) also presents at an infantile age, with a disconjugate, small-frequency, low-amplitude oscillation accompanied by an abnormal head posture. The eye movements improve during childhood, and the syndrome must be confirmed with a normal MRI/CT scan of the visual pathways. SNS usually resolves between 2-8 years of age, while the abnormal eye movements are still noticeable on ocular motor recordings.[2]

Nystagmus is caused by disorders within the mechanisms instrumental in holding gaze steady, such as the vestibular system, the gaze-holding mechanism, the visual stabilization system, and the smooth pursuit system.[2] Many of these symptoms and visual mechanisms can be improved through vision therapy, either independently or in conjunction with amblyopia, binocular disorders, and/or visual-perceptual disorders. Thus, vision therapy should be considered as a treatment in a young child with infantile-onset nystagmus.

Studies have shown that the decreased visual acuity in people with nystagmus is not due to motion blur from the uncontrolled eye movements, but rather from the decreased time the image is placed within the fovea.[3] With treatment aimed at increasing foveation and/or reducing the nystagmus intensity, it is possible that the change in nystagmus waveform improves the temporal aspect of visual function.

This case report discusses the lengthy stepwise approach to improving vision and function in an infant with infantile-onset nystagmus and strabismus.

Case Report

MA presented at 2 years of age with her mother and father, who expressed concerns of a residual eye turn following strabismus surgery and decreased vision in her left eye. Upon examination, the patient showed a constant left esotropia (post-strabismus surgery) along with latent nystagmus in her left eye, ultimately contributing to moderate amblyopia and suppression in the left eye. Her best visual acuities upon initial presentation, assessed with preferential viewing paddles, were 20/50 OD, 20/100 OS with a noticeable right head turn. Records from the pediatric ophthalmologist indicated strabismus surgery for a left hypertropia and esotropia when the child was 15 months old. Refractive error, as measured with retinoscopy, was +1.25 diopters sphere (DS) OU, and she wore low hyperopic glasses of +1.00 DS OU. Dilated fundus exams, as performed by the pediatric ophthalmologist, were unremarkable at 10 months and 23 months of age. Current treatment, as directed by the pediatric ophthalmologist, involved occlusion patching to improve visual acuity in the left eye for two hours/day.

Treatment progressed in the following stepwise approach:

1. Improve monocular VA

The toddler had previously been prescribed full-occlusion patching with a patch or occlusive sticker, which caused behavioral changes and irritability due to heightened nystagmus and worse acuity when monocular. Alternative patching methods were explored, such as fogging the lens by means of a Bangerter foil (20/400 and 20/200 blur) over the right eye for up to 3 hours/day while playing games such as ball rolling, bouncing, and popping bubbles. When the patient's parents noticed that she was looking outside of the Bangerter foil and struggling to keep her glasses on, patching therapy switched to atropine 1% ophthalmic suspension,

Table 3. Distance Visual Acuities Across Progress Exams Throughout Vision Therapy

Stage of Therapy	Date	Progress check after number of therapy sessions/length of break	Distance Visual Acuities	
			OD	OS
1. Improve Motor Alignment & Maintain VA	04/10/13	9 sessions	20/25	20/25
	01/22/14	9 month break	20/25-	20/60
	04/28/14	10 sessions	20/30	20/40
	07/21/14	7 sessions	20/20	20/50
	10/27/14	8 sessions	20/25+	20/40+
	04/06/15	4 month break	20/20	20/20
2. Prism OD: 0.5 BO OS: 3.5 BI Increased prism to OD: 2.0 BO OS: 5.0 BI 3. Break Suppression	07/29/15	8 sessions	20/30-	20/40-
	11/18/15	12 sessions	20/20	20/20
4. Binocularity & Depth Perception	03/02/16	9 sessions	20/20	20/40

with one drop instilled every other day in the right eye. The patient's parents agreed that the atropine was a way to ensure that the patient was adhering to the patching regimen without causing a heightened nystagmus. With the patient's low hyperopic refraction and no need for a lens to be fogged, it was not necessary for the patient to wear correction.

Visual acuities in the left eye varied from 20/125+ to 20/40 across progress exams over nearly two years with on-and-off patching therapy (Table 2). After two years of treatment with various occlusion techniques, the visual acuity in the left eye reliably improved to 20/40; however, the patient continued to exhibit a right head turn and suppression of her left eye.

2. Improve Motor Alignment & Maintain VA

At 4 years old, when the patient had matured enough to participate in active vision therapy, she was enrolled in an academic-based therapy program. The initial focus of therapy was on maintaining proper visual acuities and improving motor ocular alignment, which then shifted to breaking suppression.

All monocular therapy activities were performed with a Bangerter foil or frosted occluder and included activities such as eye control, SVI rotator and saccades, and pointer straw.

After 9 vision therapy sessions, visual acuity was 20/25 in both eyes, and the patient's parents decided to take a break from therapy for several months to allow the patient to mature and to improve cooperation with at-home activities. Nine months later, MA showed signs of reduced visual acuity in the left eye (20/60), and therapy resumed. Through a total of 44 sessions of vision therapy over a two-year period, the patient's acuity improved to 20/25 in the left eye, and motor alignment shifted to a small intermittent exotropia (8 pd). Table 3 describes distance visual acuity as it was assessed across progress checks throughout the duration of treatment for the patient.

3. Prism

The patient maintained a noticeable head turn to the right, which placed the left eye in its null position of slight temporal gaze, and her parents grew concerned about longterm effects of torticollis or other neck and/or back problems. Prismatic null position therapy was initiated by placing yoked prism base right in hopes of shifting the image towards the null point of the nystagmus to compensate for the head turn and to increase the duration of foveation. Prism was ground into plano polycarbonate lenses, yoked base right, with slightly more over the left eye (0.5 BO OD, 3.5 BI OS); these were to be worn as often as possible. The patient continued with weekly therapy sessions throughout this phase, and the prism was later increased to 2.0 BO OD and 5.0 BI OS after 12 sessions. At this point, visual acuity was 20/20 in the left eye, without a head turn, and with dampened nystagmus. Within two weeks, the patient's parents were able to appreciate a decreased head turn, while visual acuities remained stable at 20/20.

4. Break Suppression

Once the motor alignment had improved to a point where second-degree fusion would be possible, and in conjunction with the addition of prism, therapy was more heavily focused on anti-suppression. Monocular fixation in a binocular field (MFBF) activities ensured that the eyes had alternating roles in fixating on a specific task, while both eyes could appreciate the peripheral field biocularly. Activities involved red/green glasses and acetate filters with Hart chart near/far rock, column jumping, red red rock, and various card games with Sherman playing cards (Figure 1). Over 12 sessions, the patient was no longer suppressing her left eye, as tested with Worth 4-dot, but she exhibited no stereopsis when tested with Randot.

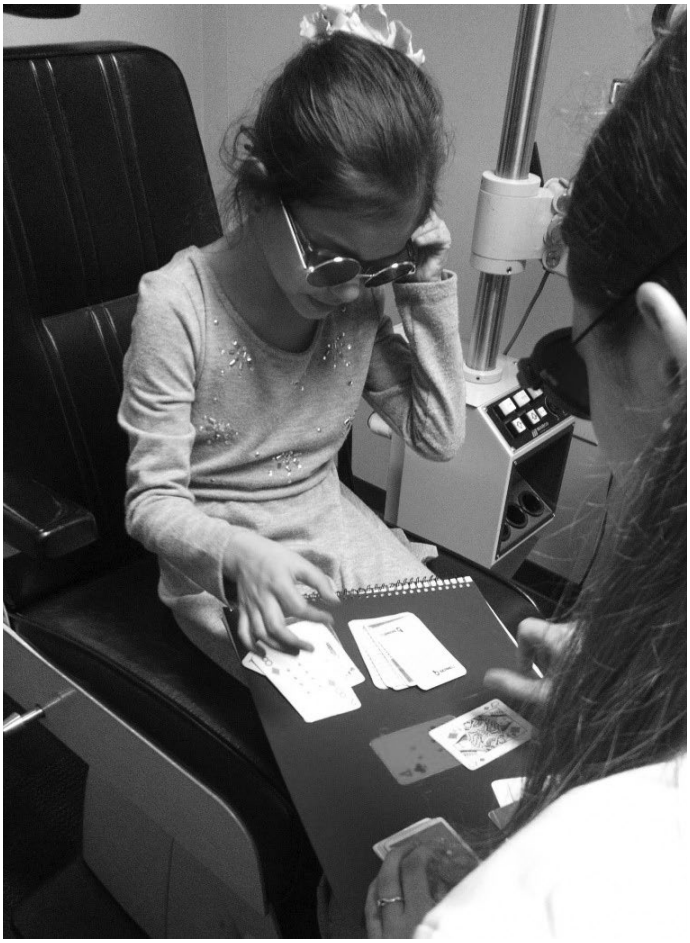

Figure 1. *Patient MA performing Sherman playing cards, an anti-suppression activity*

5. Binocularity & Depth Perception

The final step in therapy was to attempt to gain bi-foveation and appreciation of depth, or stereopsis. Activities performed included movable forest, vectograms, lifesaver circles, and VisionBuilder randot (base in and base out). Over 10 therapy sessions, the patient was unable to appreciate stereo or work her vergence system accurately; however, suppression did not appear to reoccur. At this time, it was suggested to give the patient a break from therapy and to re-evaluate the possible gains of therapy in four to six months.

Discussion

Nystagmus is often intimidating when seen in-office, and it takes some time and special attention to decide if and how to treat. Without the use of ocular motor recordings, it is indeed very difficult to properly classify the nystagmus this patient exhibited, although she appeared to have fusional maldevelopment nystagmus syndrome. While the patient developed a head turn to the right, placing her right eye in an adducting position, the nystagmus was rarely noted in the right eye. However, the null gaze of her left eye was in an abducting position, with nystagmus heightened under adduction. Upon a brief assessment, one would not consider this an association of FMNS (decreased nystagmus with adduction), but it is possible that the patient was adducting the right eye to a point that the nystagmus was not clinically noticeable and was constantly suppressing the left eye. While this updated classification system provides more information on the smaller details of nystagmus, it may confuse a clinician, who most likely lacks the technical instrumentation to measure ocular motor movements accurately.

The different options for patching therapy are typically easily interchangeable in amblyopia and strabismus; however, when a latent nystagmus is involved, the list of options narrows, and one must consider the consequences that can arise from full-occlusion patching with a heightened nystagmus. Non-opaque methods of patching include a high-plus lens over the better-seeing eye, Bangerter filters (range in blur from approx. 20/20 to light perception, across nine logarithmic densities), or tape (translucent or medical-grade) applied to the lens. Atropine 1% ophthalmic solution can be used every other day in the non-amblyopic eye to induce blur through inhibiting accommodation.[4] This option is especially favorable in people with nystagmus, since it does not fully occlude the better-seeing eye, and thus does not increase the amplitude or frequency of latent nystagmus.

Prismatic null position therapy involves the use of yoked prism with the base in the same direction as the head turn, which enables the patient to keep their eyes in an eccentric null position while decreasing the magnitude of the head turn.[5] In some cases, this may require large amounts of prism (over 20 pd), and the added weight and optical distortion from the prism may compromise acuity. In this case, relatively small amounts of prism (five prism diopters and less) were successful.

With a common association of strabismus and nystagmus, these patients will often find themselves in a pediatric/vision therapy practice. The goals of active vision therapy are to "improve fixation ability, reduce amblyopia, eliminate suppression, correct anomalous correspondence, improve sensory fusion (including stereopsis), and increase facility and range of accommodative and vergence responses."[6]

Other optical options for the treatment of nystagmus take into consideration the dampening of eye movements under adduction or convergence postures. If the patient is orthophoric, the use of base out prism over each eye and over-minusing will achieve a convergence posture. Hertle describes a trial of 7.0 prism diopters of base out prism over each eye, with an over-correction of -1.00 DS in both eyes,[2] although the magnitude of prism and minus lenses can vary. The overall convergence-dampening effect could improve distance acuity in this subset of people with nystagmus.

Medical treatment for nystagmus involves the chronic use of oral central nervous system (CNS) medications to influence the firing of neurotransmitters. Studies show that the gaze-holding mechanism is controlled by neurons within the medulla for horizontal gaze and the midbrain for vertical gaze.[7] Medications such as baclofen, clonazepam,

valproic acid, and gabapentin are amongst the more commonly used.[8] However, the large list of side effects of the CNS medications limits their use, especially within a pediatric population.

Recent studies have shown that people with nystagmus take longer to recognize moving visual targets and demonstrate an increased saccadic latency toward peripheral targets. However, the delay to recognize/respond to peripheral targets is not a visual processing issue; it is solely one of saccadic latency.[9] A slowed time to perform a saccade, while certainly not assessed on visual acuity testing, may severely affect one's function in high-stimulus situations such as driving or playing sports. Thus, measuring temporal visual function may give a better assessment of treatment outcomes in patients with nystagmus.

When assessing the importance of the treatment of nystagmus at the social level, studies have found that nystagmus has a significant impact on the perception of human characteristics. Individuals with nystagmus are being judged negatively with regard to personality traits and attractiveness, potentially placing them at a disadvantage for professional opportunities and other significant areas of everyday life.[10]

Conclusion

While infantile-onset nystagmus is not a large topic or everyday finding in practice, it is important to be able to differentiate a congenital, benign form versus one with potentially damaging associations. The new classification system, although more detailed and thorough, may make the clinical diagnosis of nystagmus type more difficult and confuse the clinician. With the high association of strabismus or other binocular vision disorders with nystagmus, addressing all visual problems is paramount in providing people with nystagmus the best treatment. Where reduced visual acuity in people with nystagmus may have an amblyogenic component, vision therapy can assist in improving the visual system in this class of patients. When a patient presents with an infantile-onset nystagmus, one should properly identify the location of a null point, any abnormal head posture, and the presence of binocular vision disorders, as well as aim to assess the eye movements to the best of their ability. Treatment is focused on improving vision through enhancing foveation time and addressing concomitant binocular vision issues. One must be mindful of visual limitations and stay committed to a lengthy therapy program. Education with parents and patients is crucial in helping them understand the goals of therapy as they progress.

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