PRELIMINARY RESULTS OF PERFORMING THE TENOTOMY PROCEDURE ON TEN ADULTS WITH CONGENITAL NYSTAGMUS (CN)

RW Hertle[a,b], LF Dell'Osso[c], EJ FitzGibbon[b]
D Thompson[d], D Yang[b], and SD Mellow[b]

Pediatric Ophthalmology, Columbus Children's Hospital, The Ohio State University, Columbus, OH[a] Lab of Sensorimotor Research, NEI, NIH, Bethesda, MD[b] Ocular Motor Neurophysiology Lab, VAMC & Dept of Neurology, Case Western Reserve University, Cleveland OH[c], NEIS Coordinating Center, EMMES Corp, Potomac, MD[d] USA

Abstract

We determined the effectiveness of the tenotomy procedure in decreasing the nystagmus of 10 selected adult subjects with CN. The primary outcome measure was the "eXpanded Nystagmus Acuity Function" (NAFX), calculated in masked fashion from eye-movement data. All subjects had persistent (one year follow-up), postoperative increases in the NAFX of their fixing (preferred) eye (average=48.8%). Secondary outcomes such as optotype visual acuity increased in 3 subjects (including 1 albino) and the NEI-VFQ showed an improvement in vision-specific mental health in all patients. There were no adverse reactions. Tenotomy caused significant damping of their CN, including aperiodic alternating nystagmus (APAN); it also changed the APAN cycle.

Keywords: Nystagmus, Tenotomy, Eye Muscle Surgery

Introduction

Congenital nystagmus (CN) affects visual acuity in both patients with and without associated sensory deficits by reducing the amount of time the target image remains in the foveal area with little or no retinal slip velocity. As the CN intensifies, foveation time is further reduced with the resulting reduction of acuity. Unfortunately, CN intensifies with any stress or excitement including, anxiety, fear, or anger. It is the interplay of these uncontrollable factors with the CN waveforms that determine visual acuity at any given time for a particular visual task. Therefore, it is not surprising to find that after a procedure that clearly damps the CN and that resulted in a subjective improvement in vision, no significant improvement may be measured at the Snellen chart.

Clearly, what is needed is a more realistic measure of the effectiveness of CN therapy vis-à-vis, how it changed the CN waveform, and a prediction of the resulting change in *potential* best-corrected visual acuity. Such a measure is the Nystagmus Acuity Function (NAF)[1] and its recent improvement, the eXpanded Nystagmus Acuity Function (NAFX).[2] Using this function on eye movement data taken under low-stress conditions helps to eliminate both intra- and intersubject variability and facilitates comparisons under different viewing conditions or pre- and post-therapy. The inherent assumption of the NAF(X) is that acuity is solely limited by the CN waveform. It is calculated using both eye position and velocity data to determine a 'foveation window' within which the data most important for acuity reside. The method is independent of the methodology used to gather the data (presuming that they are accurate and noise-free) and of the nystagmus type or waveform.

The hypothesis that a simple tenotomy of the horizontal recti would damp horizontal CN came from observations made from plots of pre- and post-operative eye movement data taken from patients who underwent Anderson-Kestenbaum procedures.[3, 4] The data showed that the CN null was widened as well as shifted and that the off-null CN was also damped. The resulting waveform changes increased foveation time and, with it, potential acuity. Performing tenotomies on a Belgian sheepdog with achiasma, CN and see-saw nystagmus (SSN)[5, 6] tested this hypothesis.[7] The positive results of that study were the foundation for this clinical trial on ten human adults. This report comes from an analysis of the pre- and post-operative data of the first five adults.

Materials and Methods

The ten CN subjects listed below had no other treatment options and included individuals with varied associated sensory defects and additional nystagmus types ((a)periodic alternating nystagmus (APAN)).

PT#	AGE/SEX	CLINICAL NOTES
1	39/M	Albinism, ET, **OS PREFERENCE**
2	30/M	Albinism, ET, **OS PREFERENCE**
3	39/M	Idiopathic CN, Ortho, **NO PREFERENCE**
4	49/M	Albinism, ET, APAN,**OS PREFERENCE**
5	39/M	Idiopathic CN/APAN, Ortho, **NO PREFERENCE**
6	28/F	Albinism, ET, **OS PREFERENCE**
7	39/M	CN/APAN, XT, **OS PREFERENCE**
8	20/M	CN/SSN/Achisama,ET, HT, **OS PREFERENCE**
9	55/F	APAN, Ortho/ **OD PREFERENCE**
10	34/F	Albinism/ET/**OD PREFERENCE**

M = Male, F= Female, ET = esotropia, XT = exotropia, Ortho = orthophoria, OS = left eye, OD = right eye, SSN= see-saw nystagmus, APAN= aperiodic alternating nystagmus

All subjects had ocular motor recordings made pre- and 1, 6 and 52 weeks post-operatively with an electromagnetic induction technique[8] using scleral search coils embedded in silastin. The system bandwidth wad 0--500Hz with <0.03°/hr drift. Eye movements were calibrated using stimuli generated by system software at a distance of 1m from the subjects and at 0 to ±30°. Monocular- and binocular-viewing records were made at all gaze positions, at near, and for prolonged fixation in primary position to test for APAN.

The primary motility measure was the NAFX, which is linearly related to predicted, best-corrected visual acuity; it was calculated in masked fashion in primary position and at various gaze angles. Secondary motility measures included the CN foveation characteristics, waveforms, and average amplitudes and frequencies were also calculated.

Secondary outcomes included visual function, assessed pre- and post-operatively using a masked measure of visual acuity (ETDRS and the NEI-Visual Function Questionnaire (VFQ). All subjects were also tested for binocular function (Worth 4-Dot at distance and near and Randot Preschool Stereoacuity Test), ocular motility (prismatic deviations at 6m and 33cm), color (Ishihara color plates), visual fields, tonometry, slit lamp examination of the anterior and posterior segments, indirect fundus examination, photographic documentation of the optic nerve head and posterior pole, and ocular oscillations in primary position in all gaze positions during both monocular and binocular viewing conditions.

All data analyses and clinical evaluations were masked. Simple tenotomies of the four horizontal recti and immediate reattachments at their original insertions sites were made using standard surgical techniques.

Results

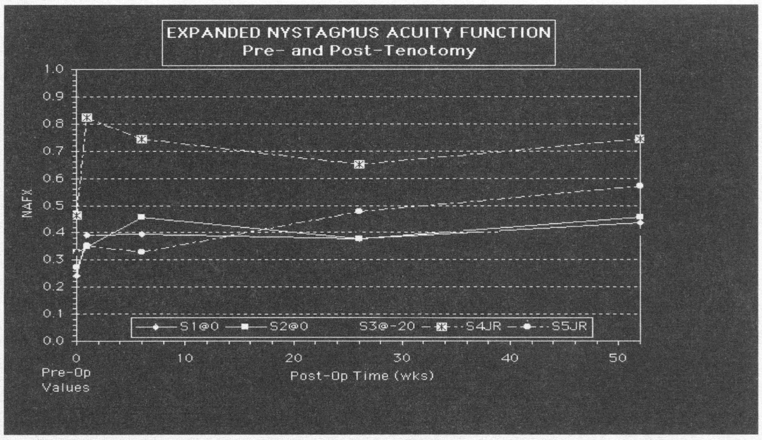

Fig. showing primary outcome measure: This shows the persistent increase in NAFX of the first 5 adults (pt's 1-5) who have complete motility analysis for one year.(S= subject and #=patient number).

The tenotomy procedure resulted in an average increase in the NAFX for all 5 subjects of 48.8%. All subjects had increased foveation time (ranging from insignificant to 79.6%). The APAN cycle also appeared to be changed.

PT#	AGE	VA OU Pre	VA OU Post
1	39	20/160	20/125
2	30	20/60	20/60
3	39	20/30	20/25
4	49	20/100	20/100
5	39	20/40	20/40
6	28	20/80	20/80
7	39	20/50	20/50
8	20	20/80	20/80
9	55	20/50	20/40
10	34	20/80	20/80

Table showing the results of the masked visual outcome measure with ETDRS chart preoperatively and at one year postoperatively in the 10 patients. (VA = visual acuity, OU = both eyes open, Pre= preoperatively, Post=postoperatively.) Three adults with improved acuity are shaded.

All patients also had increases in the vision-specific categories of the NEI-VFQ.

Conclusions

The tenotomy procedure resulted in a persistent (one year) average increase in the NAFX for the first 5 subjects of 48.8%. All subjects had persistent, increased foveation time (ranging from insignificant to 79.6%). All non-APAN CN was damped. The APAN cycle appeared to be changed; the significance of this is unknown. S3 also showed a broadening of his region of highest NAFX (encompassing 20° left gaze), demonstrating the same null-broadening effect of tenotomy seen in the Anderson-Kestenbaum results.[3] Three subjects (2 with CN only, of which 1 had APAN and 1 with CN and APAN plus albinism) had persistent increases in their measured visual acuity and all showed persistent increases in the vision-specific categories of the NEI-VFQ.

At the time of this report we are in the process of analyzing the data from one year post-tenotomy recordings in the remaining 5 adult patients. If the results continue to be positive, four-muscle tenotomy should may be employed to damp CN in patients who have no gaze-angle or convergence null (including APAN); added to the artificial divergence procedure (i.e., add a bilateral tenotomy); and combined with strabismus procedures when CN is also present (e.g., either a bimedial recession plus a bilateral tenotomy or a four-muscle recess-resect procedure for strabismus).

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