Preliminary Results of Performing the Tenotomy Procedure on Adults with Congenital Nystagmus (CN) – A Gift from "Man's Best Friend"

L.F. Dell'Osso, R.W. Hertle*, E.J. Fitz Gibbon, F.A. Miles*, D. Thompson** and D. Yeng***

Ocular Motor Neurophysiology Lab, VAMC & Department of Neurology
Case Western Reserve University, Cleveland Ohio, U.S.A.
** Lab. of Sensorimotor Research, NEI, NIH, Bethesda, Maryland, U.S.A.*
*** NEIS Coordinating Center, EMMES Corp, Potomac, Maryland, U.S.A.*

Summary

We determined the effectiveness of the tenotomy procedure in decreasing the nystagmus of 5 selected adult subjects with CN. The primary outcome measure was the "eXpanded Nystagmus Acuity Function" (NAFX), calculated in masked fashion from eye-movement data. During binocular viewing, all subjects had postoperative (6 weeks) increases in the NAFX of their fixing (preferred) eye (19.7%, 42.9%, 57.9%, 60.6%, and 62.7%; average = 48.8%). Visual acuity increased in 3 subjects (including 1 albino) and was unaffected in the other 2 albinos. However, the NEI-VFQ showed an improvement in vision-specific mental health in 4 subjects (including 2 albinos). There were no adverse reactions. Tenotomy caused significant damping of their CN, including APAN; it also changed the APAN cycle. The lack of measurable Snellen-acuity changes in 2 subjects with albinism, reflects the significant effect of their sensory deficit.

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 damped the CN and that resulted in a subjective improvement in vision, no significant improvement may be measured at the Snellen chart. Assuming that the waveforms were measured in a low-stress environment (e.g., viewing LED's in a dimly illuminated room), the pre-operative baseline CN was intensified during acuity measurements yielding a value that was lower than might be experienced in the real world during less stress. Post-operatively, the damped, low-stress CN waveform improves the latter acuity, accounting for the reported increase. However, the stress induced increase in the CN during acuity measurement may cancel that out and no improvement may be measured.

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 (Phase 1) and five children (Phase 2). This report comes from a preliminary analysis of the pre- and post-operative data of the first five adults.

Materials and Methods

The five CN subjects (ages, 29-49 years old) had no other treatment options and included individuals with varied associated sensory defects (3 had foveal hypoplasia) and additional nystagmus types (2 had asymmetric (a)periodic alternating nystagmus (APAN)).

All subjects had ocular motor recordings made pre- and 1 and 6 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 primary position and at various gaze angles. Secondary motility measures included the CN foveation characteristics, waveforms, and average amplitudes and frequencies were also calculated.

Clinical 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, ocular oscillations (in primary position, at near and at all gaze positions during both monocular and binocular viewing conditions).

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

Results

S1 was a 38 year-old male with albinism but no strabismus. His average pre-tenotomy NAFX in primary position was 0.243 (20/70-) and increased to 0.395 (20/40+) post-tenotomy, a 62.7% increase. Measured binocular visual acuity was unchanged at 20/160. The NEI-VFQ showed an increase in 3 categories, 1 category was unchanged, and a decrease in 2 categories.

S2 was a 29 year-old male with albinism and esotropia. His average pre-tenotomy NAFX in primary position was 0.330 (20/50+) and increased to 0.472 (20/35+) post-tenotomy, a 42.9% increase. Measured binocular visual acuity was unchanged at 20/60. The NEI-VFQ showed an increase in 0 categories, 2 categories were unchanged, and a decrease in 4 categories.

S3 was a 37 year-old male with esotropia. His average pre-tenotomy NAFX in primary position was 0.187 (20/100+) and increased to 0.280 (20/60+) post-tenotomy. However, these values did not correlate with his measured acuity (20/30). He had a better CN waveform at 20° left gaze. His average pre-tenotomy NAFX at 20° left gaze was 0.327 (20/50+) and increased to 0.517 (20/30+) post-tenotomy, a 57.9% increase. Measured binocular visual acuity increased from 20/30 to 20/25, a 19.9% increase. The NEI-VFQ showed an increase in 3 categories, 4 categories were unchanged, and a decrease in 0 categories.

S4 was a 49 year-old male with albinism, esotropia and APAN. His average pre-tenotomy NAFX in primary position during his best interval of jerk-right APAN was 0.463 (20/35+) and increased to 0.743 (20/20+) post-tenotomy, a 60.6% increase. Measured binocular visual acuity increased from 20/100 to 20/80, a 25% increase. The NEI-VFQ showed an increase in 2 categories, 4 categories were unchanged, and a decrease in 0 categories.

S5 was a 38 year-old male with no strabismus and APAN. His average pre-tenotomy NAFX in primary position during his best interval of jerk-right APAN was 0.275 (20/60-) and increased to 0.329 (20/50+) post-tenotomy, a 19.7% increase. Measured binocular visual acuity increased from 20/40 to 20/30, a 33.4% increase. The NEI-VFQ showed an increase in 4 categories, 2 categories were unchanged, and a decrease in 1 category.

Conclusions

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%). All non-APAN CN was damped (ranging from insignificant to 34.7%). The APAN cycle appeared to be changed; the significance of this is unknown. S3 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 increases in their measured visual acuity and 4 (2 with albinism, of which 1 had APAN and 1 other with APAN) showed increases in the vision-specific categories of the NEI-VFQ.

At the time of this report, all 15 subjects had been operated upon and we were in the process of gathering and analyzing the data from post-tenotomy recordings and examinations. If the results continue to be positive, four-muscle tenotomy should 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 esotropia).

References

[1] N.V. SHETH, L.F. DELL'OSSO, R.J. LEIGH, C.L. VAN DOREN, and H.P. PECKHAM: The effects of afferent stimulation on congenital nystagmus foveation periods, *Vision Res, 35*, 2371-2382, 1995.

[2] J.B. JACOBS and L.F. DELL'OSSO: An expanded nystagmus acuity function [ARVO Abstract], *Invest Ophthalmol Vis Sci, 39*, S149, 1998.

[3] L.F. DELL'OSSO and J.T. FLYNN: Congenital nystagmus surgery: a quantitative evaluation of the effects, *Arch Ophthalmol, 97*, 462-469, 1979.

[4] L.F. DELL'OSSO: Extraocular muscle tenotomy, dissection, and suture: A hypothetical therapy for congenital nystagmus, *J Pediatr Ophthalmol Strab, 35*, 232-233, 1997.

[5] L.F. DELL'OSSO and R.W. WILLIAMS: Ocular motor abnormalities in achiasmatic mutant Belgian sheepdogs: Unyoked eye movements in a mammal, *Vision Res, 35*, 109-116, 1995.

[6] L.F. DELL'OSSO, R.W. WILLIAMS, J.B. JACOBS, and D.M. ERCHUL: The congenital and see-saw nystagmus in the prototypical achiasma of canines: comparison to the human achiasmatic prototype, *Vision Res, 38*, 1629-1641, 1998.

[7] L.F. DELL'OSSO, R.W. HERTLE, R.W. WILLIAMS, and J.B. JACOBS: A new surgery for congenital nystagmus: effects of tenotomy on an achiasmatic canine and the role of extraocular proprioception, *J Am Assoc Pediatr Ophthalmol Strab, 3*, 166-182, 1999.

[8] D.A. ROBINSON: A method of measuring eye movement using a scleral search coil in a magnetic field, *IEEE Trans Bio Med Electron*, *BME*, 137-145, 1963.

Supported in part by Veterans Affairs Merit Review

Neuro-ophthalmology at the Beginning of the New Millennium

Proceedings of the International Neuro-ophthalmology Society Meeting

Toronto, Ontario, Canada, September 10-14, 2000

Editor

JAMES A. SHARPE

MEDIMOND

Medical Publications

www.medimond.com