Infantile nystagmus syndrome: Broadening the high-foveation-quality field with contact lenses

Giovanni Taibbi¹,4,5, Zhong I Wang¹,3, Louis F Dell’Osso¹,3

¹The Daroff-Dell’Osso Ocular Motility Laboratory, Louis Stokes Cleveland Department of Veterans Affairs Medical Center and CASE Medical School; ²Department of Neurology; ³Department of Biomedical Engineering, Case Western Reserve University and University Hospitals Case Medical Center, Cleveland, OH, USA; ⁴Pediatric Ophthalmology and Strabismus Unit, San Paolo Hospital, Milan, Italy; ⁵University of Milan, Milan, Italy

Abstract: We investigated the effects of contact lenses in broadening and improving the high-foveation-quality field in a subject with infantile nystagmus syndrome (INS). A high-speed, digitized video system was used for the eye-movement recording. The subject was asked to fixate a far target at different horizontal gaze angles with contact lenses inserted. Data from the subject while fixating at far without refractive correction and at near (at a convergence angle of 60 PD), were used for comparison. The eXpanded Nystagmus Acuity Function (NAFX) was used to evaluate the foveation quality at each gaze angle. Contact lenses broadened the high-foveation-quality range of gaze angles in this subject. The broadening was comparable to that achieved during 60 PD of convergence although the NAFX values were lower. Contact lenses allowed the subject to see “more” (he had a wider range of high-foveation-quality gaze angles) and “better” (he had improved foveation at each gaze angle). Instead of being contraindicated by INS, contact lenses emerge as a potentially important therapeutic option. Contact lenses employ afferent feedback via the ophthalmic division of the V cranial nerve to damp INS slow phases over a broadened range of gaze angles. This supports the proprioceptive hypothesis of INS improvement.

Keywords: contact lenses, infantile nystagmus, visual function

Introduction

Infantile nystagmus syndrome (INS) is an ocular motor disorder characterized by involuntary oscillations that are typically bilateral, conjugate, uniplanar, horizontal-torsional, pendular or jerk with increasing velocity slow phases (CEMAS 2001). Current INS treatment relies on optical (eg, spectacles, contact lenses), medical (eg, baclofen, memantine), and surgical procedures (eg, extraocular muscle tenotomy and reattachment at the original or a moved insertion) and is aimed at improving vision and visual function of individuals affected by INS. It has been demonstrated that the extraocular muscle tenotomy procedure, which is embedded in all extraocular muscle movement procedures, broadens and improves the range of high-foveation-quality gaze angles in INS (Dell’Osso et al 1999; Hertle et al 2003, 2004; Wang et al 2006). Convergence has also been shown to broaden and improve the range of high-acuity gaze angles in INS (Serra et al 2006). Both therapeutic methods allow INS patients to see “more” (a wider range of high-foveation-quality gaze angles) and to see “better” (improved foveation at each gaze angle).

Contact lenses have been shown to improve visual acuity and to damp the nystagmus intensity (Sédan 1966; Abadi 1979; Allen and Davies 1983; Dell’Osso et al 1988; Golubovic et al 1989); however, their effects on foveation characteristics have not been documented. In this study, we carefully examined a subject to determine whether contact lenses are able to broaden and improve the range of high-foveation-quality gaze angles. In order to gain more insight into the mechanisms of contact lenses’ effects on INS, we also compared the effects to those of convergence in the same subject.
Methods
Recording
We used a digital video system (EyeLink II, SR Research, Mississauga, ON, Canada) for the recording. The system had a linear range of ± 30° horizontally and ± 20° vertically. System sampling frequency was 500 Hz, and gaze position accuracy error was 0.5°–1° on average. The data from this system were digitized at 500 Hz with 16-bit resolution. Immediately after the subject’s prescribed contact lenses were inserted, the EyeLink signal from each eye was calibrated with the other eye behind cover to obtain accurate position information; the foveation periods were used for calibration.

Subject and protocol
This study was approved by the local IRB and written consent was obtained from the subject before the testing. The subject was a calm 66-year-old male with INS; his nystagmus waveforms were well-documented previously and their characteristics representative of others with INS. Of particular importance was the data collected on this patient over a time span of 45 years using several types of infrared reflection, several types of magnetic search coil, and several types of high-speed digital video recording systems. That unique volume of data confirmed the static nature of his INS characteristics under similar recording conditions and allowed comparisons to be made from data sets taken at different times for different studies. The subject had a convergence “null” and wore either 7 PD base-out prisms at different times for different studies. The subject had a and allowed comparisons to be made from data sets taken his INS characteristics under similar recording conditions.

Results
The top panels (a, b, and c) of Figure 1 show the typical waveforms and their corresponding NAFX values at –20°, 0°, and 25°. These panels were generated by the NAFX software program from fixation data at these gaze angles.
Upward eye movements in these panels denote a rightward eye movement. To assist comparison, the y-axis scales were set to be the same in all three panels. At the $-20^\circ$ gaze angle (a), the waveforms were of a pendular with foveating saccade type. The waveforms changed to pseudo pendular with foveating saccade when the eyes were straight ahead (b). At $25^\circ$ (c), the waveforms transitioned to jerk right. The nystagmus peak-to-peak amplitude at $25^\circ$ was the largest among all three gaze angles.

The NAFX value at $0^\circ$ was the highest; the foveation periods were well lined up within a foveation position window of $\pm 0.5^\circ$ (dash-dotted lines). At $-20^\circ$, although the foveation periods were also in a foveation window of $\pm 0.5^\circ$, they had more position variation and higher slow-phase velocity. These factors contributed to a lower NAFX than that at $0^\circ$. At $25^\circ$, the position window was $\pm 1.25^\circ$, much larger than those at $-20^\circ$ and $0^\circ$. However, the NAFX value at $25^\circ$ was comparable with that of $-20^\circ$. The comparison of the NAFX values at the three gaze angles revealed that this subject had better foveation quality at primary position.

The bottom panel (d) of Figure 1 consists of three NAFX versus gaze-angle curves under different viewing conditions. The potential visual acuities corresponding to the NAFX values are listed on the right margin. The “Far with Contacts” curve is a second-order polynomial fit ($r^2 = 0.55$) based on the NAFX values we obtained in this study. The “Far” ($r^2 = 0.86$) and “Conv” ($r^2 = 0.44$) curves were retrieved from a previous study. All three curves demonstrated peaks at $\pm 0^\circ$. When the subject was converging, the curve (dash-dotted) was high and broad (LFD = 50°). When the subject was wearing contact lenses, the curve (dashed) was medium high and medium broad (LFD = 35°). These values are all greater than the $\pm 15^\circ$ range normally used before a head turn is employed. The solid curve (without contact lenses or converging) has the lowest peak and a narrow LFD of 15°. Comparing the dashed curve and the solid curve, the difference

Figure 1 Fixation data (eye position vs. time) for the subject during far viewing with contact lenses at different gaze positions: $-20^\circ$ (a), $0^\circ$ (b) and $25^\circ$ (c). The corresponding NAFX values are also shown. Foveation periods were automatically thickened by the NAFX algorithm. The area between the dash-dotted lines represents the foveation position window used to calculate the NAFX. In (d), plots of NAFX vs. gaze angles for far viewing, far viewing with contact lenses, and while converged (60 PD). Fitted polynomial curves are shown. NAFX-correlated potential visual acuities are adjusted for the subject’s age. Positive gaze angles indicate rightward gaze and “Conv” is convergence.
Discussion

One advantage of contact lenses over eyeglasses for INS patients (especially those with eccentric nulls) is that they allow fixation through the optical sweet spot at all times, thereby preventing distortion. Inspection of Figure 3 in the 1988 paper demonstrating that contact lenses damped INS over a range of gaze angles suggests that the curves were also broadened (Dell’Osso et al 1988). Our findings confirmed that suggestion by documenting a 133% (from LFD = 15° to LFD = 35°) broadening of the NAFX curve, which is directly correlated to improvements in visual acuity. Thus, contact lenses broaden the range of high-foveation-quality gaze angles in addition to any improvement in primary-position visual acuity they may provide. Figure 1 demonstrates that contact-lens broadening was equivalent to that of convergence but that the latter improved the NAFX to a greater extent. This subject did see “more” (a wider range of high-foveation-quality field) and “better” (improved foveation at each gaze angle) with contact lenses. This suggests that contact lenses do far more to improve visual function than has been previously appreciated and is a potentially important therapeutic option for INS patients.

The NAFX values shown in Figure 1 for −20° and 25° are essentially equivalent, indicating that the corresponding potential visual acuities are also. However, the amplitude of the INS at 25° is approximately four times greater. This reaffirms the poor correlation between nystagmus amplitude and potential visual acuity (Dell’Osso and Jacobs 2002) and illustrates why amplitude is only a reliable measure of cosmetic improvement, not visual function.

INS can be highly dependent on emotion and attention. However, using the same low-stress, non-acuity dependent recording conditions and protocols as we did in this study and all previous studies, if there is no additional sensory deficit acquired later in life, the baseline of INS does not change in the patient and the measurements of waveforms do not change either. We have over 40 years of eye-movement data from this subject, as well as multiple studies of other patients over years to decades, which demonstrate the stability of baseline INS over time. It was, therefore, justifiable to utilize historical data as the non-contact-lens control in this study. This study documented how contact lenses changed the subject’s waveforms, especially compared to convergence-induced changes; studying the broadening effects of contact lenses on more INS patients will determine the range of idiosyncratic foveation changes.

Contact lenses affect eye movements via afferent information carried on the ophthalmic division of the V (trigeminal) cranial nerve, not due to the mass of the lenses nor the refractive correction (Dell’Osso et al 1988). That was demonstrated by the use of topical anesthetics to limit the proprioceptive input from the contact lenses. Tenotomy has also been hypothesized to utilize proprioceptive information carried via the III, IV, and VI cranial nerves in addition to the V cranial nerve (Dell’Osso et al 1999; Hertle et al 2003, 2004). The absence of a proprioceptive effect on eye movements was the conclusion of an early study that only measured the effects on a specific class of ocular motor neurons (Keller and Robinson 1971). However, new studies have identified a second class, more amenable to taking part in a proprioceptive feedback loop (Büttner-Ennever et al 2001, 2002; Ugolini et al 2006).

Specifically, a reduction of the steady-state muscle tension effectively reduces the muscle responsiveness to the small signals driving the slow phases (ie, small-signal gain reduction) (Wang et al 2006). The broadening effects we documented in this study provide evidence that the therapeutic effect of contact lenses on INS was not only due to their ability to correct refractive errors or to their optical properties (eg, lower spherical and chromatic aberrations than spectacles), but also due to their interference with the neurological mechanisms affecting the resulting nystagmus characteristics.

The broadening effect of contact lenses took place within the 30-minute time interval between insertion of the lenses and recording of the data. Previous studies of the effects of afferent stimulation of the V cranial nerve (pressure, touch, electric, vibration, and even air flow over the forehead – personal observation) have documented that this type of INS damping was immediate (ie, within milliseconds) (Dell’Osso et al 1988, 1991; Sheth et al 1995). Because contact lenses stimulate the same pathway, we conclude that their salutary effects in this study, as in the 1988 study (Dell’Osso et al 1988), also occurred instantaneously. This suggests that afferent stimulation of proprioceptive pathways modulates slow eye movements (eg, INS slow phases) in an
on-line manner, not unlike a proprioceptive stretch reflex. We therefore, expect to find that contact lenses broaden the high-NAFX range of gaze angles in others whose INS is damped by them.

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Disclosure
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References
