Major Article

# Clinical and ocular motor complications of extraocular muscle extirpation for infantile nystagmus syndrome

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**PURPOSE** To describe the effects of extraocular muscle extirpation performed after previous eye muscle surgery in a 20-year-old woman with infantile nystagmus syndrome (INS) for whom we have 19 years of follow-up data.

**METHODS** Clinical examinations were performed. Eye movement data analysis was carried out using the eXpanded Nystagmus Acuity Function (NAFX) and longest foveation domain (LFD).

**RESULTS** The patient re-presented to the authors at age 20, 2 years after bilateral anterior myectomy of the horizontal rectus muscles, bilateral anterior nasal transposition of the inferior oblique muscle, and bilateral superior oblique recessions. Evaluation revealed deterioration in nystagmus at lateral gaze angles, new incomitant strabismus with severe loss of convergence, limited ductions, saccadic hypometria, slow saccades, and hypo-accommodation. Also, there was a pre- to post-extirpation minimal change of 21% in her peak NAFX, a 50% decrease in LFD, plus a predominant, asymmetric, multiplanar oscillation.

**CONCLUSIONS** It appears that in this patient, horizontal extirpation failed to abolish the nystagmus and caused significant, new, symptomatic deficits interfering with many of the patient's visual functions. (J AAPOS 2018;■:1-5)

The ability of the ocular motor system to shift and maintain gaze, align, and converge the eyes (in nonstrabismic patients), pursue moving targets, and stabilize gaze in the presence of head, body, or environmental movement are essential for good visual function in the real world. Arguably, the most effective surgical treatments for infantile nystagmus syndrome (INS)[1] are those that do not curtail ocular motor functions. The "maximal recession" procedure, which requires large recessions of all four horizontal rectus muscles, was an example of this negative effect.[2,3] Although that procedure was shown to damp the INS, it also significantly compromised the patient's ability to make accurate, rapid saccades, maintain eccentric gaze, align the eyes (especially in lateral gaze), converge the eyes, preserve stereopsis, pursue moving targets, or stabilize the eyes in the presence of head, body, or environmental motion. With the advent of the four-muscle tenotomy and reattachment (T&R) procedure, along with more traditional operations, homeostatic surgical algorithm for treatment of INS[4-8] were developed.[9-11] It has been demonstrated that the beneficial effects of the enthesial tenotomy portion (cutting the tendinous insertion) of any eye muscle surgery improves INS waveforms and dynamic visual functions and also prevents ocular motor system deficits seen in maximal extraocular muscle recessions.[4,12] In 2002 Sinskey extended the presumption underlying the maximal recession procedure to what he may have presumed to be a logical conclusion by proposing total *extirpation and disposal* of the anterior portions of the four horizontal rectus muscles.[13-17] The current case report presents clinical data collected over a 19-year period for a young woman with INS treated with standard eye muscle surgery and subsequently with muscle extirpation surgery.

## Methods

Prior to the recent clinical intervention, the patient had been enrolled in a research protocol in which the collection and reporting of data followed institutional research approval via a research registry protocol with appropriate informed consent and US Health Insurance Portability and Accountability Act of 1996 regulatory requirements. Repeated, routine examinations were

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performed on a routine basis over a period of 19 years. Additional special testing included optical coherence tomography, fundus photography, electroretinography, visual evoked potentials, color vision, visual field, and contrast sensitivity. The recent muscle extirpation surgery was *not* part of this protocol.

Eye movement data were recorded at the Daroff-Dell'Osso Ocular Motility and the Visual and Ocular Motor Physiology Labs. The first recording was made when the patient was 1 year old using an Ober2 infrared system (Permobil, Timra, Sweden, 300 Hz). The video system used in 2001 and 2002 was an EyeLink II (SR Research, Mississauga, ON, Canada) that had a post-calibration linear range of ±30° horizontally and ±20° vertically. The data from these systems were digitized at 500 Hz with 16-bit resolution. Eye positions and velocities (obtained by analog differentiation of the position channels) were also displayed on a strip-chart recording system (Beckman Type R612 Dynograph). The system used when the patient was 6 and 9 years of age was an Ober2; when she was 15 and 20, an EyeLink II. The signal from each eye was calibrated with the other eye covered to obtain accurate position information; the foveation periods of each nystagmus cycle were used for calibration. The subject was seated in a chair with headrest and either a bite board or a chin stabilizer, far enough from an arc of red LEDs or a projection screen to prevent convergence effects (>5 feet). An experiment consisted of from one to ten trials, each lasting under a minute, with resting time allowed between trials.

### Data Analysis

All analysis was carried out in MATLAB, using the eXpanded Nystagmus Acuity Function (NAFX)[18-20] and other OMtools software (omlab.org). Two-dimensional (radial) NAFX values were compared to those using the predominantly horizontal nature of the oscillations. The longest foveation domains (LFD) were calculated as the range of gaze angles, where the NAFX was ≥90% of the peak NAFX value.

## Results

The patient's examinations have consistently and repeatedly shown normal color vision, visual field, lids, adnexa, anterior segment, and intraocular pressure; and abnormal (paradoxical) pupils, decreased foveal reflex, scattered peripheral, pigmentary disturbance, and mildly anomalous optic nerves. She always had a variable small-angle multiplanar strabismus (exotropia and hypertropia) and no stereopsis. She had no abnormal head posture.

At age 5, an associated congenital, stable, rod-cone dystrophy was diagnosed based on an abnormal electroretinogram and normal visual evoked potentials. Her best-corrected binocular visual acuity over 19 years' follow-up ranged between 20/80 and 20/60. She has had significant anisometropic astigmatism (at the time of this evaluation, −4.50 + 4.25 × 100 in the right eye and −4.50 + 2.00 × 170 in the left eye).

At age 6 years she underwent uncomplicated bilateral medial rectus T&R for the INS combined with a bilateral lateral rectus recessions of 3.0 mm for the exotropia, performed by one of the authors (RWH). After T&R surgery she was able to read easily and comfortably; she eventually obtained a driver's license and had no trouble driving. She did not have oscillopsia, head turn, or diplopia.

At age 18, believing her nystagmus could be stopped completely, she underwent an "anterior myectomy of the right and left medial and lateral rectus, anterior nasal transposition of the right and left inferior oblique and recession of the superior oblique right and left" at another institution. Each horizontal rectus was "allowed to retract into the sleeve where it disappeared from view" (from surgical notes). We are unaware of any IRB-controlled research protocol for that procedure.

At age 20, at the time of her most recent visit to our clinic, 2 years after extraocular muscle extirpations, she had the following subjective complaints: (1) persistent asymmetric, multiplanar oscillopsia and diplopia; (2) an anomalous and pronounced chin-down head posture; (3) incomitant vertical strabismus; (4) difficulty with reading and driving, and (5) motion sickness. Additionally, objective evaluation after extirpation revealed the following: (1) profound monocular asymmetry (right eye worse than left) in the horizontal, vertical, and torsional planes of the nystagmus (which increased in intensity at near); (2) incomitant strabismus (incomitant right hypertropia; (3) right head tilt; (4) no convergence, and incomitant exotropia; (5) limited ductions (see Figure 1); (6) a chin-down anomalous head posture; (7) saccadic hypometria; (8) low vestibular ocular reflex and horizontal pursuit gain; and (9) significant hypo-accommodation.

### Eye Movement Data

The patient's eye movements were recorded six times, from 1 year to 20 years of age. A timeline for the recordings and two surgeries is provided in Table 1. The initial preoperative eye movement recordings, made when the patient was 1, 5, and 6 years of age, showed INS with both horizontal and vertical components (horizontal amplitudes usually greater than vertical), resulting in circular/elliptical/oblique motion. The horizontal components of both eyes were conjugate and the vertical components varied from 0° to 180°; when the vertical components were 180° out of phase, the oscillations had a see-saw component. The waveforms were pendular, pendular with foveating saccades, triangular, and pseudo-jerk. Neither horizontal nor vertical gaze changed the INS substantially; that is, there were no "nulls" in either plane. Figure 2 shows eye position versus time and scan paths of both eyes (before the standard surgery) during attempted right eye foveation of a target at 0° with the left eye with exotropia of about 10° and hypotropia of about 2°.

The scan paths of both eyes during attempted fixation of targets at 0° and ±20° horizontally and at 0° and ±10° vertically before standard surgery may be seen in eFigure 1. The scarcity or even absence of repeated cycles with accurate foveation illustrates the difficulty (more