Original Article

Clinical and electrophysiological results of eye muscle surgery in 17 patients with downbeat nystagmus

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Purpose: To test the hypothesis that eye muscle surgery in treatment of patients with acquired downbeat nystagmus results in improvement measures of visual and ocular motor function. **Methods:** This is a prospective, interventional case series analysis of clinical and electrophyisological data before and after eye muscle surgery in 17 patients with acquired downbeat nystagmus who did not respond to medical treatments. Outcome measures included: 1) routine demography and clinical characteristics, 2) subjective oscillopsia (SO), 3) binocular best-corrected visual acuity in the null position (BVA), 3) primary position strabismic deviation (SD), 5) anomalous head posture (AHP), 6) contrast sensitivity function (CS), and 7) nystagmus slow phase velocity (SPV). All patients were followed at least 12 months. Parametric and non-parametric statistical analysis of outcome measure data above pre- and post-treatment were perfomed using standard software on grouped data using computerized software. **Results:** Patients' age ranged from 5 to 85 years (average 27 years). About 59% were male. Follow up ranged from 1–10 years (average 2.0 years). Around 70% had an associated central nervous systemic diagnosis, 100% had an AHP, oscillopsia and decreased CS, 53% had other eye disease, and 59% had strabismus. There were no complications from surgery. There were signficant post-treatment improvements in mean/median group BVA, SO, SD, AHP, CS, and SPV. **Conclusion:** This study supports the hypothesis that eye muscle surgery as treatments for patients with acquired downbeat nystagmus can result in improvements in multiple aspects of ocular motor and visual functions.

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Downbeat nystagmus (DBN) is one of the most frequent forms of acquired nystagmus.[1] Affected patients suffer from postural imbalance, cerebellar ataxia, oscillopsia, and reduced visual acuity.[2] Most patients have associated cerebellar or neurodegenerative disorders.[3] Nystagmus disorders have been classified in many ways, resulting in some confusion and disagreement among clinicians and scientists. The National Eye Institute sponsored workshop, Classification of Eye Movement Abnormalities and Strabismus (CEMAS), has attempted to resolve some of the confusion with a publication that defines the various types of nystagmus.[4] The published definition of DBN by the CEMAS working group has been used in this study.[4] The major clinical characteristics of DBN are; a conjugate, upward drift of the eye followed by a downward fast phase that does that not depend on vertical head position but has a gravity-dependent component, and a horizontal gaze-evoked nystagmus component.[5] The intensity of the oscillation is usually worse in eccentric gaze horizontally and in down gaze. Pathophysiologic hypotheses include; deficient cerebellar inhibition of the brainstem, imbalance of central vertical vestibular pathways, impaired vertical smooth pursuit, a variant of gaze evoked nystagmus, and a gravity-dependent anomaly of normal upward drift.[5] The most likely mechanism is damage of inhibitory, vertical, gaze-velocity sensitive Purkinje cells in the cerebellar flocculus.[5]

Non-medical therapy of symptoms associated with DBN include; retinal image stabilization techniques, real-time computer-based visual feedback, physiotherapy, and, occupational and speech therapy.[6] Medical treatment of symptoms associated with DBN most commonly involve trials of a host of neurologically active medications.[2,7,8]

Although eye muscle surgery for nystagmus in infancy and childhood is associated with the infantile forms has a long history and, is standard of care, there are less reports of eye muscle surgery to treat, acquired forms of nystagmus.[9] Wang *et al*. reported, using a different surgical procedure from that reported in this paper (tenotomy with reattachment of two horizontal recti and recession of two horizontal recti for associated strabismus) improved measures of nystagmus and visual acuity and reduced diplopia and oscillopsia in only one patient with DBN.[10] The purpose of this report is to summarize

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a case series of patients who had eye muscle surgery in an attempt to improve the signs and symptoms associated with DBN.

Methods

All testing, data collection, analysis, and reporting were approved by the Institutional Review Board of Akron Children's Hospital, Akron, OH, USA and The Children's Hospital of Pittsburgh of UPMC, Pittsburgh, PA, USA. All procedures observed the declaration of Helsinki and informed consent/assent was obtained from all patients/parents. Inclusion criteria included; clinical and electrophysiological diagnosis of acquired DBN with or without associated strabismus, age >7 years and previously failed systemic medical therapy. Exclusion criteria included, visual system disease other than strabismus, DBN and ametropia, neurological diseases prohibiting subjective examinations, medications used during the study, and follow-up period affecting the ocular motor system (e.g., neuroleptics, benzodiazepines, aminopyridines, antipsychotics, antidepressants, and narcotics), and follow up <12 months.

Eye muscle surgery

The goals of eye muscle surgery were to move the up-gaze eccentric null position associated with DBN closer to primary position by moving both eyes down without creating a secondary cyclovertical incomitancy or gaze paralysis, and treat any coexisting strabismus. This was accomplished with bilateral superior rectus recessions of 5.0 mm combined with bilateral inferior oblique myectomies in all patients. The reason that we have preferred bilateral oblique and recti operation for chin-up postures is that this combination will *prevent* secondary alphabet and cyclotorisonal deviations more than combined vertical rectus recession and resection procedures due to enhanced or diminished secondary actions of adduction and abduction. The key to surgical success in this group of patients is attention to detail with *equal* oblique and recti surgery on each eye. The only consequence of surgery is usually a 15–20% comitant, limitation of vertical gaze with some lid retraction.[22-24] Those with associated horizontal strabismus had additional horizontal rectus surgery. Standard methods of preoperative evaluation, anesthesia, and fornix-based eye muscle surgery were employed in the surgical treatment of all patients.

Outcome variables

Measures prospectively analyzed for this study included: 1) routine demography and clinical characteristics, 2) subjective oscillopsia (SO), 3) binocular best-corrected visual acuity in the null position (BVA), 3) primary position strabismic deviation (SD), 5) anomalous head posture (AHP), 6) contrast sensitivity function (CS), and 7) nystagmus slow phase velocity (SPV). All patients were followed at least 12 months. Standard clinical monitoring of surgical complications was performed.

Clinical and ocular motor evaluation

Demographic and clinical data reported in this series includes: age, sex, ocular alignment, associated systemic diseases, and refractive error. Cycloplegic refraction, tonometry, and examination of the anterior and posterior segments were performed on all patients. Clinical evaluation of the ocular motor oscillations included changes in the oscillation in primary position distance and near and in all nine diagnostic positions of gaze, under monocular and binocular conditions and over time (~5 min) [Table 1].

Subjective oscillopsia scale

Patients were asked to grade their subjective sensation of oscillopsia after surgery based on three choices; no change, improved, or absent.

Visual acuity testing

Measurements of BVA were obtained both binocularly and monocularly with best spectacle-correction at 3M in the patients' preferred head position (least intense nystagmus gaze position) using the Pediatric Eye Disease Investigator Group (PEDIG) amblyopia treatment studies electronic acuity testing protocol.[11]

Strabismus deviation

Ocular motor examination included a determination of heterophoria/tropia at distance (>3 m) and near (33 cm) using an alternate prism and cover testing in all diagnostic positions of gaze.

Anomalous head posture

AHP was measured while the patients wore a Cervical Range of Motion device (CROM^R) [Fig. 1]. The CROM^R device consists of two gravity-dependent goniometers and one compass dial on a head-mounted frame allowing measurement of range of motion in three planes to ensure that the precision of head position was consistent for AHP measures at baseline and the post-treatment visits.

Contrast sensitivity testing

CS was measured using the CSV-1000E^R sine wave grating test face (Vector Vision^R) with the patient seated 2.5 m from the chart. This test provides for four rows of sine wave gratings. At the recommended test distance, these gratings test spatial frequencies of 3, 6, 12, and 18 cycles/degree (cpd). Testing at all spatial frequencies was administered under monocular and binocular conditions after the patient had adapted to room luminance (85.0 cd/m^2) for 5 min. The patient's refractive correction was in place while allowing use of any anomalous head posture necessary to position their gaze in their null zone. CS values were documented in log units (LU). Sensitivity levels at each frequency ranged from 0.70 to 2.08 LU for 3 cpd, 0.91–2.29 LU for 6 cpd, 0.61–1.99 LU for 12 cpd, and 0.17–1.55 LU for 18 cpd. The results of the patient's first attempt were considered a trial and the values recorded

Figure 1: Cervical Range of Motion^R Device. Photograph illustrating measurements of head posture used to collect anomalous head position data. The device is placed on the head prior to visual acuity testing. The instrument ensures accurate determination of the head position (eccentric null zone position) in pitch, yaw, and roll during routine binocular best-corrected distance acuity testing

Table 1: Patient characteristics

PT #	STRAB	Other eye DX	SX DX	Refractive error
1	XT-HT Suppression	INS, ONH	ICP, ataxia, shunt	+3.00 + 2.50 × 90 OU
2	ET-HT Diplopia	None	None	+1.00 OU
3	None Stereo+	Opsoclonus-myoclonus	Ataxia, neuroblastoma, developmental delay	+1.50 OU
4	None Stereo+	None	None	None
5	None Stereo+	None	DM2, CVA, HTN	+0.50 + 0.25 × 105 +0.25 + 0.50 × 89
6	ET Diplopia	None	Cerebellar ataxia, B12 deficiency, pernicious anemia	+1.00 + 0.50 × 90 +1.00
7	ET Diplopia	None	None	−0.50 + 0.50 × 90 −1.25 + 2.00 × 65
8	ET-HT Suppression	Ptosis, rod-cone dystrophy, optic nerve atrophy	Alexander disease, epilepsy, bulbar weakness hypothyroidism, Asperger's disorder, delay in development, Raynaud's disease, SIADH	−2.75 + 2.75 × 105 −1.00 + 1.00 × 90
9	ET-LHT Diplopia		None	−4.00 + 1.25 × 90 OU
10	ET Suppression	Ocular motor apraxia	Austism, sleep apnea	−0.50 sphere OU
11	ET Suppression	None	ADHD, ataxia	−2.00 + 1.75 × 115 −1.25+0.50×90
12	None Stereo+	Vestibular imbalance, Pseudophakia	Migraine Tinnitus, stroke	−2.25 + 2.00 × 180 −3.50+ 2.75 × 180
13	None Stereo+	Pseudophakia Chorioretinal scar	Hearing loss	−2.25+ 2.25 × 4 −1.50+2.25 × 173
14	ET Suppression	Strabismic amblyopia	Seizure, Tourettes, ADHD, ODD, VNS device	+2.00 + 1.00 × 90 +2.00 + 0.75 × 90
15	None Stereo+	None	Depression, ARNOLD Chiari type 1, bone dysplasia, seizures, dwarfism, DD, asthma	−2.25 + 1.75 × 120 −1.50 + 1.50 × 113
16	LHT diplopia	Optic atrophy	Ataxia, thiamine deficiency, dysautonomia wernicke's, gastric bypass	−2.50 + 1.50 × 100 −3.00 + 2.00 × 80
17	None Stereo+	Reading difficulty	None	−0.50 + 3.25 × 75 −0.50 + 3.25 × 105

Clinical characteristics of 17 patients with DBN. PT: Patient, DX: Diagnosis, SX: Systemic, Stereo+: Has demonstrable stereopsis of at least 500 s/arc using Titmus[R] stereopsis testing, XT: Exotropia, HT: Hypertropia, ET: Esotropia, LHT: Left hypertropia, INS: Infantile nystagmus syndrome, ONH: Optic nerve hypoplasia, ICP: Raised intracranial pressure, DM2: Diabetes mellitus type 2, CVA: Cerebrovascular accident, HTN: Systemic hypertension, SIADH: Syndrome of inappropriate secretion of antidiuretic hormone, ADHD: Attention disorder hyperactivity disorder, ODD: Oppositional defiant disorder, VNS: Vagal nerve stimulator, DD: Developmental delay

at the second attempt were used as the CS measurement. The contrast level of the last correct response was taken as the CS threshold.

Eye movement recording and slow phase velocity

All patients had eye movement recordings. The presentation of stimuli, and the acquisition, display, and storage of data were controlled by a series of computers using the standard Microsoft[R] and Matlab[R] software. The horizontal and vertical eye movement recordings were made using the Eyelink[R] remote video eye movement system (Eyelink[R], SR Research Ltd., Mississauga, Ontario, Canada) with a data sampling rate of 500–1000 Hz. Calibration was accomplished monocularly, both online and later by the Matlab[R] computer program. Fixation was clinically supervised between 0 and 30° horizontally and 0 and 20° vertically, both in 5° steps with both eyes open then fixation at 0° with both eyes open for 5–10 min.

SPV from 5 s fixation intervals under binocular conditions from the patient's preferred eye, from each recording session were averaged and used for the data analysis purposes. The mean SPV for each period was calculated with its standard error from the median velocity of the nystagmus (200 msec period) that occurred for each eye position. The following parameters were used: spontaneous vertical drift velocity on gaze straight ahead with the head erect, up-gaze 20° and down-gaze 20° (SPV in °/s), SPV_{pp} (slow phase velocity primary position), SPV_{up} (slow phase velocity up-gaze), and SPV_{dn} (slow phase velocity down-gaze). The eye movement velocity was calculated using a differentiation of the eye position and a low-pass filtering with a corner frequency of 30 Hz. The high-frequency velocity peaks of the nystagmus' quick phases, saccades, and blink artifacts were removed from the eye velocity using an absolute acceleration threshold of 700°/s^2 and a median filter with a time window of 500 msec.

Statistical analysis

A paired *t*-test was used to determine the difference between baseline (after adaptation to spectacles) and post-treatment (2–6 months after final treatment) group means for BVA and CS. All reported *P* values are two-sided using a significance level of $P < 0.05$. Group mean analysis of baseline and post-treatment AHP was accomplished using a Fisher's exact test. Analyses were conducted using the GB-STAT version 10 (Dynamic Microsystems, Inc[R], 2004). We applied the Shapiro–Wilk test to determine, whether the slow phase nystagmus velocity data were normally distributed. One-way analysis of variance (ANOVA) was used for comparison.

Results

Clinical demographic results

Seventeen patients with DBN evaluated between the years 2006 to 2015 are the subjects of this study and ranged in age from 5.5 to 85.1 years (average 27 years). Fifty-nine percent were male. Follow up after final treatment ranged from 1–10 years (average 2.5 years). Twelve (70%) of the patients had an associated neurological diagnosis, this included; hydrocephalus 1, ataxia 5, neuroblastoma 1, developmental delay 2, B12 deficiency 2, Alexander disease 1, SIADH 1, autism 1, sleep apnea 1, attention deficit disorder 1, migraine 1, cerebrovascular disease 1, seizure disorder 2, Tourette syndrome, 1, vagal stimulator 1, depression 1, Arnold Chiari malformation 1, dysautonomia, and hearing loss 1 [Table 2].

All had decreased BVA, CS, oscillopsia, and an AHP. The AHP was chin-down. Ten (59%) of patients had strabismus. Twelve (71%) had a significant refractive error (> +3.5 sphere, −0.50 sphere, +1.5 cylinder, or 1.5 diopters of anisometropia in any meridian) [Table 1]. All patients were prescribed Ampyra (3,4 diaminopyridine) at some point prior to surgery without success due to intolerance, ineffectiveness, or unwilling to purchase medication.

Surgery

All patients had bilateral, 5.0 mm recession of the superior recti combined with inferior oblique myectomy. In those 10 patients who had additional horizontal deviations, additional horizontal surgery was performed in 5 in whom it was surgically indicated, due to a deviation >10 prism diopters, which consisted of horizontal rectus recession and/or resection. The goals of surgery were to treat combinations of the patient's strabismus, anomalous head posture, and nystagmus. No patients with fusion prior to surgery lost fusion after surgery. Of the five patients with preoperative diplopia four developed postoperative suppression of diplopia or stereopsis. There was no postoperative change in fusion status in all five patients with preoperative suppression and all seven with preoperative stereopsis of at least 500 s/arc. In all patients there was a bilateral, symmetric, minimal, limitation of up-gaze (about 10–15%) without an induced vertical, horizontal, or torsional gaze incomitance. No patient experienced subjective diplopia in free space or with the Worth-dot testing, or needed a second operation for recurrent strabismus or treatment of a new or recurrent head posture as of the last follow-up visit. Only 4 of 17 patients had a small angle residual strabismus (<8 prism diopters) after surgery [Fig. 2].

Visual acuity

Visual acuity outcome data reported in this study included baseline and at least 2 months and not longer than 6 months after surgery. We used 2–6 months' post-treatment acuity data to allow for complete adaptation to treatments and to avoid natural age-related changes in the younger age group due to visual system maturation. The average group mean BVA improved significantly ($P < 0.001$) after treatment (baseline average preoperative logMAR BVA = 0.42 ± 0.23, average

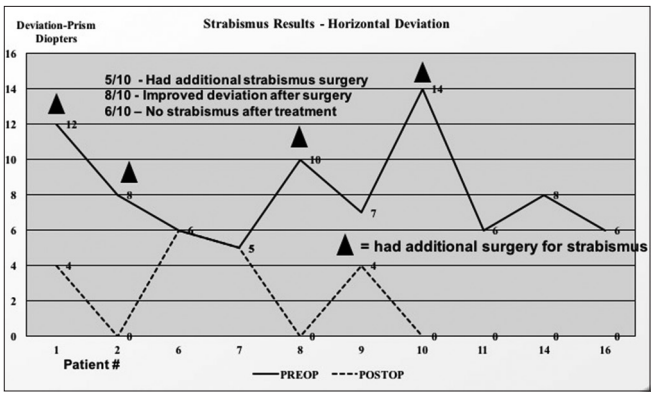

Figure 2: Strabismus results. Graphic results of changes in deviation after eye muscle surgery in those 10 of 17 patients with DBN who had an associated horizontal strabismus. Five of 10 had no horizontal rectus surgery due to having only a small angle strabismus while the remaining 5 had additional horizontal rectus surgery. At last follow up, 6 of 10 were orthophoric and the remaining 4 patients had <8 prism diopters of primary position horizontal deviation on alternate prism cover testing (Y axis is deviation in prism diopters using alternate prism cover testing, X axis show the 10 patients and their patient number who had associated horizontal strabismus)

Table 2: Classification of eye movement abnormalities and strabismus characteristics of downbeat nystagmus

Disease name	Central vestibular imbalance nystagmus
Criteria	Mixed horizontal-torsional trajectory; usually beats away from the side of a vestibular lesion, associated neurologic signs and symptoms Usually acute onset of vertigo, nausea, dizzy, oscillopsia, Associated with other signs of vestibulocerebellar involvement
Common associated findings	Downbeat, upbeat, torsional, horizontal, jerk seesaw, slow phases may have linear-, increasing-, or decreasing-velocity waveforms, poorly suppressed by fixation of a visual target, may be precipitated or exacerbated or changed in direction, by altering head position, vigorous head-shaking (horizontal or vertical), or hyperventilation, convergence may increase, suppress or convert to upbeat to downbeat nystagmus and vice versa, commonly associated with impaired smooth pursuit, gaze-evoked nystagmus, gait instability, and ataxia. MRI/CT scan of brain reflects underlying disease, ocular motility recordings show linear ("constant velocity") slow phases
General comments	Prognosis depends on underlying disease

MRI: Magnetic resonance imaging, CT: Computed tomography

post-treatment BVA logMAR 0.16 ± 0.14) [Fig. 3]. Although group means showed improvement in visual acuity, individual patient acuity may have varied ± 0.1 logMAR based on chance alone. This is inherent in the testing methodology and age of the patients involved in this report and the reason we chose to perform group mean analysis.

Subjective oscillopsia

SO was present in 100% of patients before surgery and there was no change in 2/17, (11%), improvement in 11/17 (65%), and absent in 4/17 (24%).

Anomalous head posture

The AHP at which BVA was obtained improved significantly (P = 0.0018) in all patients (average 24° preoperatively to 6° postoperatively), and was within 8° of straight in all patients [Fig. 4].

Contrast sensitivity

Table 3 shows changes in CS at all cycles per degree (cpd) stimuli before and post-surgery (final) conditions which was statistically significantly better at all stimuli.

Slow phase velocity

The group mean change in SPV values after surgery significantly improved in all three positions of gaze [Fig. 5a–c]. From 2.59 ± 0.83 °/s to 1.36 ± 0.98°/s in up-gaze; from 3.63 ± 0.78 °/s to 1.75 ± 0.69 °/s in primary gaze; from 4.84 ± 1.03 °/s to 2.64 ± 0.94 °/s in down-gaze (signed rank test, Z = 3.7; P = 0.0013) [Figs. 5–7].

Discussion

DBN is a frequent form of acquired nystagmus and manifests with oscillopsia, postural instability, and cerebellar gait disruption.[1] Additional ocular motor signs such as gaze-evoked nystagmus, deficient smooth pursuit eye movement, and strabismus are often associated.[3,12,13] In the majority of patients DBN is caused by bilaterally impaired function of the cerebellar floccular lobe or its connections to the midbrain.[5,14] Visual function in patients with acquired nystagmus is affected by unpredictable combinations of ocular motor (nystagmus, visual-vestibular, strabismus) and sensory system deficits (ametropia, diplopia, optic nerve, and/or retinal disease).[15,16] Symptoms include impaired visual acuity, oscillopsia, diplopia, anomalous head posture, gait disturbances, poor ocular motor pursuit, and reading skills.[6,17]

Different GABAergic substances have been used to treat DBN, with moderate success.[7,18] 3,4-diaminopyridine (3,4-DAP), which is a non-selective blocker of the Kv family of voltage-gated potassium channels, effectively suppresses DBN probably via an inhibition of potassium channels of Purkinje cells. 4-aminopyridine (4-AP) also alleviates the symptoms of DBN.[7,18] Based on the current state-of-the-art RCTs, the use of 4-AP in a dosage of 5 mg two to four times per day is generally recommended for the treatment of DBN.[7,18] The non-selective activator of small conductance calcium-activated potassium

Table 3: Contrast sensitivity testing - results

Frequency exam time	Mean±SD^	Minimum^	Maximum	P*
3 cpd				
Preoperative^	1.38±0.47	0.70	2.00	<0.001
Postoperative	1.85±0.18	1.44	2.08	
6 cpd				
Preoperative	1.63±0.45	0.91	2.19	<0.001
Postoperative	2.07±0.18	1.63	2.29	
12 cpd				
Preoperative	1.27±0.49	0.61	1.79	<0.0002
Postoperative	1.71±0.19	1.34	1.88	
18 cpd				
Preoperative	0.92±0.52	0.17	1.48	<0.0004
Postoperative	1.28±0.25	0.66	1.48	

*Stimuli in cycles/degree, ^Contrast sensitivity values in Log units (LU). Summarized the group mean response to CS testing with the CSV 1000^R chart at each of the four testing stimuli. There were significant group mean improvements in all four spatial frequency targets after treatment. Frequency: Cycles/degree test pattern, Exam: Preoperative: Preoperative baseline or postoperative: Postoperative, final, ± SD. *Cpd: Cycles per degree, SD: Standard deviation

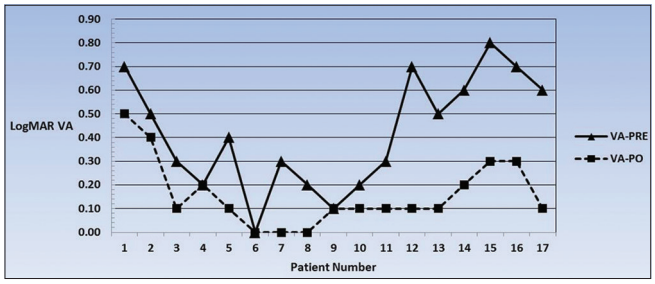

Figure 3: Visual acuity at baseline and after treatment. This figure shows a comparison plot of individual patient and group mean best-corrected binocular acuity logMAR acuity at baseline and post-treatment. The average group mean BVA improved significantly (P < 0.001) after treatment (baseline average preoperative logMAR BVA = 0.42 ± 0.23, average post-treatment BVA logMAR 0.16 ± 0.14) (Figure 3). Y axis shows logMAR values from 0 to 0.9 and X axis shows patient number. VA-PRE: Visual acuity preoperatively, VA-PO: Visual acuity postoperatively

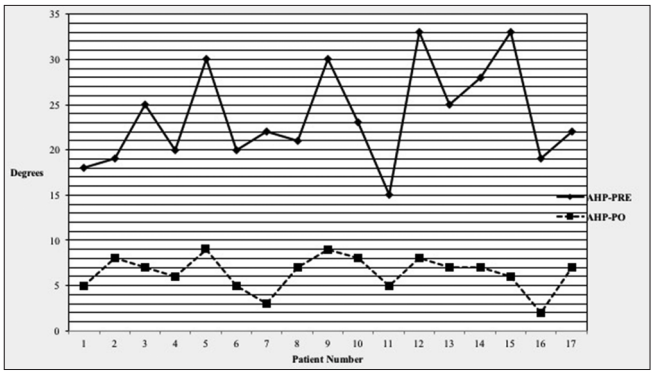

Figure 4: Anomalous head posture at baseline and after treatment. This figure shows a comparison plot of average head posture in degrees where best binocular visual acuity was obtained at baseline and after treatment. There was a significant group mean difference patients pre to postoperatively (P < 0.002) in all patients (average 24° preoperatively to 6° postoperatively), and was within 8° of straight in all patients (Y axis shows head posture deviation from straight in degrees, X axis shows patient number). AHP: Anomalous head posture, PRE: Preoperatively, PO: Postoperatively

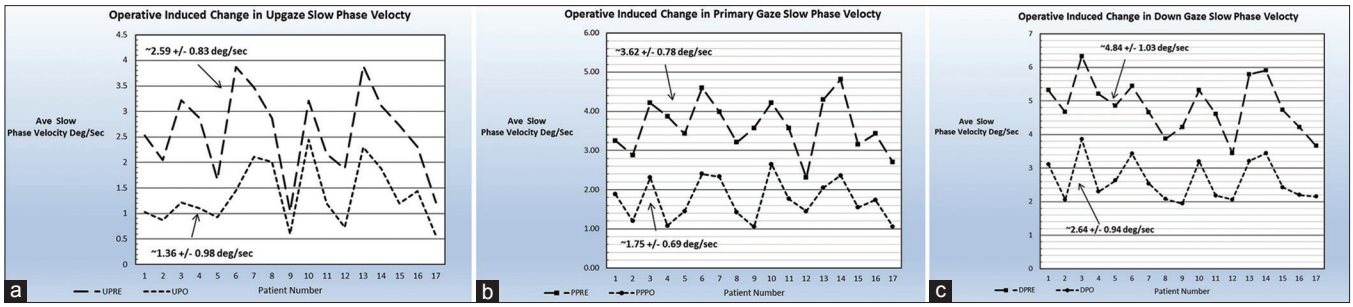

Figure 5: Measurement of slow phase velocity. These graphs show data from group mean average of slow phase velocity as a function of gaze at baseline and after treatment. The group mean change in SPV values after surgery significantly improved in all three positions of gaze. From 2.59 ± 0.83 °/s to 1.36 ± 0.98 °/s in up-gaze (a), from 3.63 ± 0.78 °/s to 1.75 ± 0.69 °/s in primary gaze (b), from 4.84 ± 1.03 °/s to 2.64 ± 0.94 °/s in down-gaze (c) (signed rank test, $Z = 3.7$; $P < 0.002$). UPRE: Up-gaze preoperatively, UPO: Up-gaze postoperatively, PPRE: Primary position preoperatively, PPO: Primary position postoperatively, DPRE: Down-gaze preoperatively, DPO: Down-gaze postoperatively

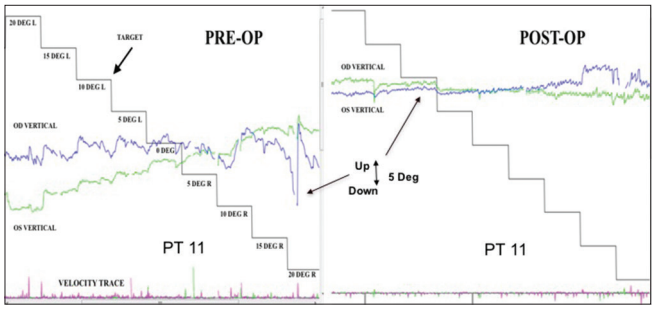

Figure 6: Example of treatment effect on nystagmus during horizontal gaze. This figure shows binocular, vertical position and velocity, eye movement characteristics of patient 11 at baseline and after treatment as a function of horizontal gaze. There is a clear postoperative reduction in nystagmus intensity, both amplitude and frequency, seen across gaze (position trace right and left eye seen on top and velocity tract at bottom). OD: Right eye, OS: Left eye, DEG: Degrees, L: Left, R: Right, PRE-OP: Preoperative recording, POST-OP: Postoperative recording

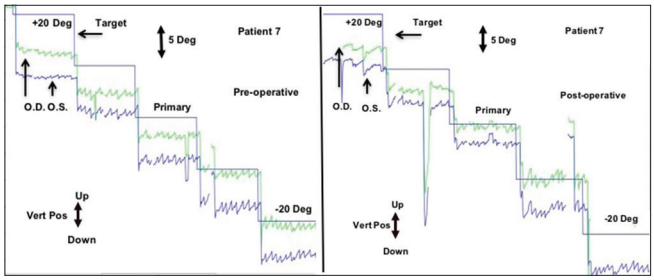

Figure 7: Example of treatment effect on nystagmus during vertical gaze. This figure shows binocular, vertical position, eye movement characteristics of patient 7 at baseline and after treatment as a function of vertical gaze. There is a clear postoperative reduction in nystagmus intensity, both amplitude and frequency, seen in all vertical gaze positions except down-gaze 20°. The postoperative change is especially noticeable in primary position (position trace right and left eye seen on top and velocity tract at bottom). OD: Right eye, OS: Left eye, DEG: Degrees, + = Up-gaze, – = Down-gaze

channels (SK-channels), chlorzoxazone, has recently been demonstrated to be a potential therapeutic agent for treatment of DBN.[19] In general, DBN intensity can be reduced in slightly over 50% of patients with these medical treatments, but many patients may not tolerate, respond to, or cannot afford drug therapy.[2,3,20–22]

Surgery on the eye muscles as treatment for the infantile forms of nystagmus is an accepted practice and its purpose is to reduce the nystagmus intensity, increase foveation time, broaden and deepen the null zone, and straighten the eyes and the head.[23,24] Eye muscle surgery for acquired nystagmus is not a common practice with few studies reporting its use.[9,10] We used multiple outcome measures in an attempt to discover how eye muscle surgery affects the visual system in patients with DBN. Eye muscle surgery for an up-gaze eccentric null position combined with horizontal strabismus correction resulted in improvements in BVA, SO, CS, AHP, SPV, ocular alignment, and binocular function. We chose a bilateral 5.0 mm recession of the superior rectus combined with bilateral inferior oblique myectomy as a result of the author's experience with using this as one procedure to treat dissociated vertical deviation without creating cyclovertical incomitancy. It is not clear how eye muscle surgery results in these changes, although movement of the null position and central nervous system changes in a proprioceptive ocular motor loop after cutting the enthesial portion of the extraocular muscle tendon, have both been postulated.[25–27] It may be that the ideal treatment for these patients are combinations of medical and surgical treatment but those data will probably only be accumulated through multicenter cooperative efforts.

Study limitations

There are several limitations of this case series. This is not a randomized clinical trial and there are no randomized clinical trials evaluating eye muscle surgical treatment of DBN thus, clear evidenced-based guidelines including indications and risks of treating visual system anomalies associated with DBN are based on case/cohort series such as this one. The use of a nominal, 3-part subjective grading system for oscillopsia could not be analyzed using non-parametric testing, thus no statistically significant conclusions can be declared. Although a 5 mm recession of the superior rectus was effective in this series of patients, the anomalous head posture was decreased but not completely eliminated, and, since up-gaze is much less important than down-gaze, further over-correction of the anomalous head position could be considered, and it is possible that this could be achieved by larger magnitude superior rectus muscle recessions. We did not report long term follow-up data or whether the effect diminishes over time but are planning to do this in a later report as all the patients are

continually examined. Although all patients had DBN, we included patients of all ages, varied baseline visual systems, and a few with associated neurological disease. Although the patients reported here consist one of the largest, prospective, surgical treatment cohorts of DBN patients, this remains level 2–3 evidence at best.

Conclusion

This data supports the hypothesis that eye muscle surgery improves pathological signs and symptoms in patients with acquired downbeat nystagmus.

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Conflicts of interest

There are no conflicts of interest.

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