

# Effect of 4-Horizontal Rectus Muscle Tenotomy on Visual Function and Eye Movement Records in Patients with Infantile Nystagmus Syndrome without Abnormal Head Posture and Strabismus: A Prospective Study

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## Abstract

**Purpose:** To evaluate the effect of tenotomy on visual function and eye movement records in patients with infantile nystagmus syndrome (INS) without abnormal head posture (AHP) and strabismus

**Methods:** A prospective interventional case-series of patients with INS with no AHP or strabismus. Patients underwent 4-horizontal muscle tenotomy. Best corrected visual acuity (BCVA) and eye movement recordings were compared pre and postoperatively.

**Results:** Eight patients were recruited in this study with 3 to 15.5 months of follow-up. Patients showed significant improvement in their visual function. Overall nystagmus amplitude and velocity was decreased 30.7% and 19.8%, respectively. Improvements were more marked at right and left gazes.

**Conclusion:** Tenotomy improves both visual function and eye movement records in INS with no strabismus and eccentric null point. The procedure has more effect on lateral gazes with worse waveforms, thus can broaden area with better visual function. We recommend this surgery in patients with INS but no associated AHP or strabismus.

**Keywords:** Infantile Nystagmus Syndrome, Congenital Nystagmus, Nystagmus without Abnormal Head Posture, Tenotomy, Extraocular Muscle Surgery, 4-Horizontal Rectus Muscle Tenotomy

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## Introduction

Infantile nystagmus syndrome (INS) is an involuntary and conjugate oscillation of both eyes with unknown etiology. This disorder can be present at birth or develop within the first six months of life. Its prevalence has been reported from 3 in 1,000 to 1 in 20,000,<sup>1</sup> so it is estimated that between 3,500 to 210,000 individuals are affected in Iran.

INS increases in intensity with attempted fixation and dampens with convergence. The nystagmus has different intensity in different gaze positions and there is a gaze angle with lowest intensity which is called "null point" and can be whether "eccentric" or at "primary position". This can make patient to prefer an abnormal head posture (AHP) in which he has diminished oscillation and thus better visual acuity (VA). INS is usually associated with strabismus and refractive errors; also can be accompanied by other congenital ocular disorders such as albinism, achromatopsia or congenital cataract.<sup>2,3</sup>

There are various extraocular muscle (EOM) surgical options with proven efficacy on INS associated AHP, eccentric null point or strabismus. These include recessions, resections, or their combination such as Anderson-Kestenbaum. But for patients with INS who have no AHP, eccentric null point or strabismus there is no definite treatment yet. Retro-equatorial 4-horizontal rectus large recessions (to 12 mm posterior to their insertions) was proposed as an option for these patients by Bietti in 1956. It seems this large recessions lead to some changes in length-tension curvature which decrease speed of eye movements<sup>4</sup>; however, these large recessions have technical difficulties.

In recent two decades, previous investigators noticed that beside mechanical effect of EOM displacements -to bring eccentric null point to primary position or correct ocular deviations- best corrected visual acuity (BCVA) is also improved after these surgical manipulations. Based on these observations, in 1998 Dell'Osso and Hertle introduced "tenotomy" (i.e. simply disinserting and reattaching EOMs at their original insertions) as a hypothetical treatment for congenital nystagmus.<sup>5</sup> It is hypothesized that tenotomy probably changes proprioceptive inputs which control resting tension of the

EOMs.<sup>6,7</sup> So, it can reduce the slow-phase gain of nystagmus.

Recently, some interventional case-series have reported 4-muscle tenotomy efficacy in improvement of BCVA and nystagmus waveforms.<sup>8-10</sup> It is advantageous over large rectus recessions because it is technically easier and has less adverse effects, such as diplopia in extreme lateral gaze.

We performed this study to evaluate the effect of 4-horizontal muscle tenotomy on visual function and eye movement records in patients with INS without AHP and strabismus.

## Methods

This study was approved by ethical committee of Eye Research Center, Farabi Eye Hospital, Tehran University of Medical Sciences (TUMS), Iran. We performed this prospective interventional case-series from 2011 January till 2012 December and it was compliant with the principles of the Declaration of Helsinki. All participants were adults and signed the informed consent before recruitment to study.

### Recruitment of patients

We recruited patients older than 18 years old with clinical diagnosis of INS without AHP or strabismus, and binocular BCVA of  $20/400$  to  $20/30$  at distance for this study. They were able to undergo a complete ophthalmic evaluation and standard eye movement recording. Exclusion criteria were: 1) history of EOM surgery, 2) other ocular disorders such as cataract, glaucoma, 3) presence of any systemic disorder such as developmental delay or cerebral palsy, 4) significant constant AHP ( $>10^\circ$ ) or eccentric null point which requires surgical correction, 5) large angle horizontal ocular deviation ( $>45$  prism diopter) which needs surgery on more than two EOMs, 6) vertical ocular deviation more than 5 prism diopter or dissociated vertical deviation which should be corrected with surgical management, 7) follow-up less than three months.

### Ophthalmic evaluation and eye movement recordings

Two investigators evaluated all patients separately, in at least two different visits within two weeks before surgery. Ophthalmologic

evaluation consisted of a complete slit-lamp examination of anterior and posterior segments, measuring intraocular pressure - by Goldman tonometry, "monocular and binocular BCVA" - by using a Snellen chart at distant (six meters) and near (33 cm), "cycloplegic refraction" - by using a combination of retinoscopy and subjective techniques after full medical cycloplegia, "fusion" at distance (six meters) and near (33 cm) - by using worth-4-dot, "stereo vision" - by using Stereo FLY and RANDOT Stereo Tests at 33 cm, and "ocular alignment" at six meters and 33 cm.

A masked investigator performed eye movement recordings using a videonystagmography machine (GN Otometrics A/S, MADSEN, AURICAL, ICS, Chartr 200, version 6.8.2). This machine traces data with frequency of 60 hertz (Hz) and resolution of 0.1 degree. We asked patients to look at a stationary target in the straight head position at 120 cm from the patient and at 20° gaze positions with both eyes open. Duration of each trial, considering the patient cooperation, was as long as possible but less than one minute to minimize patient's inattention which causes nystagmus intensity reduction.

### ***Surgical procedure, and post-op care and assessment***

One surgeon performed all operations. "4-horizontal muscle tenotomy" was performed under general anesthesia. Limbal incision localized peritomy was done on medial and lateral rectus muscles areas. After dissecting the muscles from tenon capsule and surrounding tissues like other EOM surgeries, their attachments to sclera - at their insertion - were cut (disinsertion). Then the muscles were sutured by vicryl 6.0 to sclera at their original insertion sites (reattachment). Peritomies were sutured in the same manner of all EOM surgeries.

All patients received topical steroid and antibiotic drops four times daily for seven to 10 days. Two investigators, masked to patients' preoperation information, visited all patients at one day, one week, one month, three months, and every three months postoperatively. They performed a complete ophthalmologic exam as mentioned for preoperation evaluation. Also, any complaints

of patients or side effects such as muscle movement limitation were documented. The same investigator obtained postoperation records of eye movements which were performed at least three months after the procedure.

### ***Signal analysis***

Two investigators selected best foveation segments of eye movement records in each trial to avoid noise signals (due to blinkings) and inattention periods. Average amplitude (defined as peak to peak excursion of the oscillation in degree), and frequency (number of nystagmus cycles per second) of nystagmus records were calculated for each patient and for each different gaze angle. We also measured maximum of slow phase velocity (Peak SPV) which is the velocity (degree/second) of eye movement during its slow phase as an indicator of nystagmus intensity. Data were analyzed in SPSS 11.5 using non-parametric tests.

### ***Results***

A total of eight patients (four men and four women) with mean age of 26.1±8.3 year (range 18 to 44 years) completed the study. Their follow-up period was between 3 to 15.5 months (median 7.3 months). Table 1 shows their preoperation information.

Near BCVA was between  $20/50$  and  $20/20$  preoperatively. It improved in two patients postoperatively ( $p=0.07$ ). Table 2 shows comparison of distant BCVA and eye movement records' (at all gazes) of all patients before and after operation. Our participants had preoperative distant BCVA of  $20/100$  to  $20/30$  and four of them showed at least 0.1 logMAR improvement with statistical significance ( $p=0.05$ ). Pre-op overall amplitude at all gazes was in the range of four to 19 degree (median 10 deg) and had an average reduction of 30.7% postoperatively ( $p=0.002$ ). Peak SPV was in the range of seven to 92 degree/second preoperatively and had a 19.8% average decrease, subsequently ( $p=0.001$ ). In contrast to amplitude and SPV, the frequency of nystagmus had a smaller range (2.2 to 5. Hz) preoperatively and we found no clinical significant change in it after the surgery ( $p=0.6$ ).

Table 2 summarizes pre and postoperative data for each patient individually. Patients who

showed BCVA improvement (patients 2, 4, 7, 8) had pre-op BCVA less than or equal to  $20/50$ , and others with better pre-op VA showed no change.

No similar association was observed between nystagmus dampening effect of the procedure and pre-op BCVA. Six of eight patients showed marked (>25%) decrease in nystagmus intensity and one demonstrated moderate reduction (23.6%). One patient had a small increase in amplitude (1.1%).

We also compared eye movement records and amount of their improvement (effect size of tenotomy) between different gazes. Table 3 summarizes these findings. Amplitude and SPV were better at primary position compared with lateral gazes preoperatively ( $p=0.001$ ). Their improvement was marked at right and left gazes and less pronounced at primary position ( $p=0.001$ ).

Pre-op and post-op nystagmus waveforms of patient 8 at each gaze angle are illustrated as an example in figure 1.

Four patients had fusion at near and demonstrated gross amount of streopsis (700-2000 second of arc) preoperatively. No patient lost his fusion or streopsis, also there was no fusion gain or streopsis improvement after tenotomy. Three had a transient or variable AHP which could not be surgically corrected. Postoperatively we found no remarkable change in the amount or duration of their AHP; and also no new AHP developed in others. Six patients had no ocular deviation preoperatively and maintained their alignment after the procedure. Among two other with small angle esotropia, one had ocular alignment after the surgery and the deviation was decreased in the other one.

We detected no complication and patients reported no discomfort during their follow-up period (3 to 15.5 months).

**Table 1.** Preoperative information of patients

Patient No.	Age Mean (SD)	Sex F:M	Clinical findings	FH	OD VA - OS VA	OU BCVA	Dev.	AHP	F/U (m.) median
1	23	M	-	+	20/60 - 20/60	20/40	Ortho	-	12.5
2	27	M	-	-	20/50 - 20/100	20/50	Ortho	Variable turn<10	9.5
3	44	M	-	-	20/30 - 20*/40	20/30	Ortho	Variable turn<10	15.5
4	30	F	Albinoid fundus	-	20/200 - 20/200	20/100	Ortho	-	7.0
5	19	F	-	-	20/40 - 20*/40	20*/40	ET, <8PD	-	3.0
6	22	F	-	-	20/60 - 20/100	20/40	Ortho	Transient, Lt turn<10	7.5
7	26	M	-	-	20/60 - 20/60	20*/60	Ortho	-	4.0
8	18	F	-	-	20/100 - 20/100	20/100	ET, <8PD	-	3.0
Total	26.13 (8.3)	4:4	-	-	-	-	-	-	7.3

F: Female, M: Male, FH: Family history of nystagmus, VA/BCVA: Distant monocular and binocular best corrected visual acuity, Dev: Ocular deviation, Ortho: Orthotropia, ET: Esotropia, PD: Prism diopter, AHP: Abnormal head posture, Lt: Left, F/U: Follow-up duration

**Table 2.** Preoperative best corrected visual acuity and nystagmus characteristics; and their changes after tenotomy overall and for each patient individually

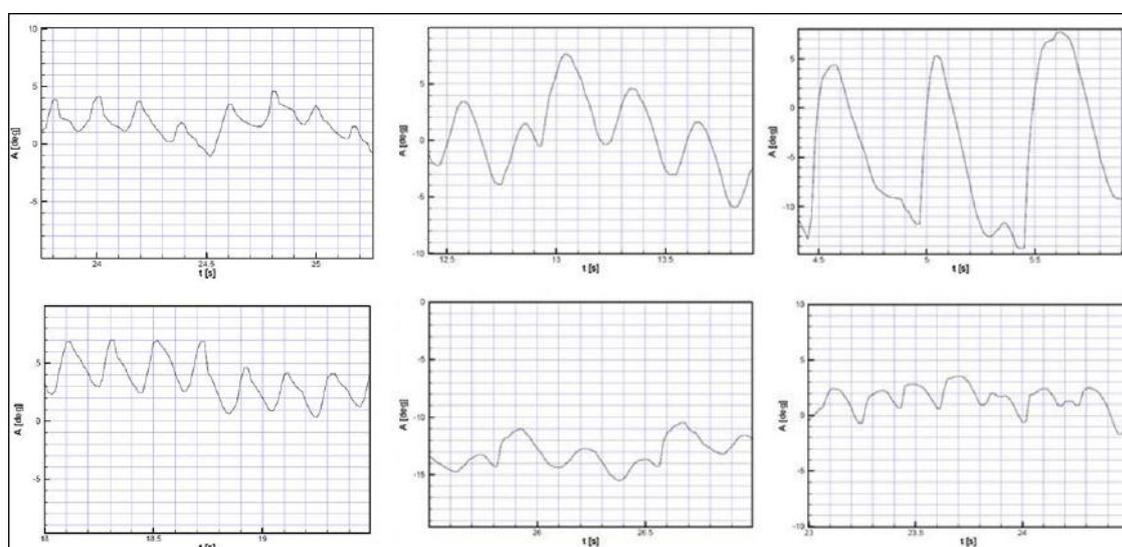
Patient No.	Pre-op Dev.	Post-op Dev.	Pre-op BCVA	Change in VA logMAR	Pre-op Amp (%decreased)	Mean Freq pre/post	Pre-op Peak SPV (%decreased)
1	Ortho	Ortho	20/40	0	8.7 (-1.1%)	5.0 / 5.2	46.7 (-11.3%)
2	Ortho	Ortho	20/50	-0.1	19.6 (41.3%)	3.9 / 3.4	81.7 (35.9%)
3	Ortho	Ortho	20/30	0	6.7 (40.3%)	5.2 / 6.2	63.0 (39.7%)
4	Ortho	Ortho	20/100	-0.1	14.4 (23.6%)	4.2 / 4.9	92.0 (21.7%)
5	ET, <8PD	ET, <8PD	20*/40	0	4.0 (27.5%)	2.2 / 2.6	7.0 (-10.0%)
6	Ortho	Ortho	20/40	0	11.4 (41.2%)	4.4 / 4.3	57.3 (56.4%)
7	Ortho	Ortho	20+/60	-0.1	5.9 (27.1%)	4.9 / 4.1	15.3 (-11.1%)
8	ET, <8PD	Ortho	20/100	-0.2	19.0 (46.3%)	3.8 / 4.5	57.3 (37.2%)
Mean Relative decrease (SD)				-	30.7% (15.3)	-	19.8% (27.0)
p-value*				0.05	0.002	0.6	0.001

Dev: Ocular deviation, VA: Distant best corrected binocular visual acuity, Amp: Amplitude, Freq.: Frequency, SPV: Slow-phase velocity, \* Wilcoxon Rank test for comparison of pre and post-op value changes

**Table 3.** Comparison of eye movement records and amount of their improvement (effect size of tenotomy) between different gazes

	Mean amplitude		Mean peak SPV	
	Pre-op (deg.)	%decreased	Pre-op (deg./sec.)	%decreased
Center	8.8	10.2%	38.6	17.0%
Right gaze	12.4	30%	62.0	26.1%
Left gaze	11.7	33.8%	57.0	24.4%
p-value*	0.001	0.001	0.001	0.001

SPV: slow-phase velocity  
\* Friedman test

**Figure 1.** Pre and post-op eye movement records of patient 8 showing marked reduction of nystagmus amplitude and velocity after the procedure. Top level shows pre-op and bottom level shows post-op records. Nystagmus waveforms at left gaze, primary position and right gaze are shown in left, center and right column respectively.

## Discussion

Although the etiology of INS is unknown, it seems an interruption between the developing sensory and motor systems interferes with ocular motor calibration (neural “cross talk”) and results in involuntary oscillations.<sup>3,4</sup> It is hypothesized that tenotomy probably changes proprioceptive inputs which control resting tension of the EOMs.<sup>7,11</sup> So, it can reduce the slow-phase gain of nystagmus. Some recent studies reported supportive information for this theory by introducing palisade endings with proprioceptive role at the tendino-scleral interface (“enthesis”) of EOMs.<sup>12,13</sup>

In our prospective interventional study on eight patients with INS, we found that tenotomy could improve primary position BCVA. We also demonstrated this procedure

markedly diminished eye movement intensity. These are in parallel with other recent studies which have shown beneficial effect of 4-muscle tenotomy on visual performance and waveform characteristics.<sup>8,14</sup>

We found better visual outcome in four patients with poorer pre-op BCVA ( $\leq 20/50$ ), while no obvious change was noticed in those with pre-op BCVA of more than  $20/50$ . Similar association has been observed in other studies too.<sup>8</sup> These evidences suggest that for having a favorable effect on visual functions we should consider an optimum pre-op BCVA; however, we did not find this association between improvement of amplitude and intensity of nystagmus and pre-op BCVA of the patient.

Amplitude and SPV were better at primary position compared with lateral gazes preoperatively; this finding describes worse nystagmus waveforms at lateral gazes compared with primary position which is anticipated in patients with INS without eccentric null point. We found nystagmus dampening effect of the procedure was more at lateral gazes where the waveform characteristics were worse. This results in broadened visual field with low nystagmus intensity; and it means BCVA improvement due to tenotomy is not limited to primary position. The same effect has been reported by Wang et al previously.<sup>10,15</sup>

Although clinical visual gain was moderate in our study patients (four of eight patients), we found notable reduction in nystagmus intensity and amplitude (seven of eight patients, with moderate to high dampening). It seems that nystagmus dampening effects of these procedures are independent of their visual outcome.

## Conclusion

In conclusion, 4-muscle tenotomy in patients with INS without AHP and strabismus has an

overall therapeutic effect on primary position BCVA, diminishes nystagmus intensity at different gazes and broadens area with higher quality vision. We highlight prognostic role of pre-op BCVA on the procedure expected therapeutic effect to improve visual function. Patients with pre-op BCVA of more than  $20/50$  are probably not good candidates for tenotomy to improve their BCVA.

We recommend performing “4-horizontal muscle tenotomy” for patients with INS without AHP and strabismus who have pre-op distant BCVA of equal to or less than  $20/50$ .

Further studies with larger sample size and evaluation of some other visual function indices such as “time to acquisition” and “foveation time” are needed to better identify tenotomy outcome.

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