

Velocity Discrimination in Infantile Nystagmus Syndrome

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PURPOSE. Research on infantile nystagmus syndrome (INS) and velocity discrimination is limited, and no research has examined velocity discrimination in subjects with INS at their null position and away from it. This study aims to investigate how individuals with INS perform, compared with controls, when carrying out velocity discrimination tasks. Particularly, the study aims to assess how the null position affects their performance.

METHODS. INS subjects ($N = 21$, mean age 24 years; age range, 15–34 years) and controls ($N = 16$, mean age 26 years; age range, 22–39 years) performed horizontal and vertical velocity discrimination tasks at two gaze positions. Eighteen INS subjects were classified as idiopathic INS and three had associated visual disorders (two had oculocutaneous albinism, and one had congenital cataract). For INS subjects, testing was done at the null position and 15° away from it. If there was no null, testing was done at primary gaze position and 15° away from primary. For controls, testing was done at primary gaze position and 20° away from primary. Horizontal and vertical velocity discrimination thresholds were determined and analyzed.

RESULTS. INS subjects showed significantly higher horizontal and vertical velocity discrimination thresholds compared with controls at both gaze positions ($P < 0.001$). Horizontal thresholds for INS subjects were elevated more than vertical thresholds ($P < 0.0001$) for INS subjects but not for controls. Within the INS group, 12 INS subjects who had an identified null position showed significantly lower horizontal and vertical thresholds at the null than at 15° away from it ($P < 0.05$).

CONCLUSIONS. Velocity discrimination was impaired in INS subjects, with better performance at the null. These findings could assist in understanding how INS affects the daily activities of patients in tasks involving moving objects, and aid in developing new clinical visual function assessments for INS.

Keywords: infantile nystagmus syndrome, motion perception, velocity discrimination, null position, psychophysics

Infantile nystagmus syndrome (INS) is an involuntary, constant, rhythmic eye oscillation which usually presents at or near birth and persists throughout life. Its waveform parameters can vary with gaze angle, leading many patients to the adoption of an abnormal head posture to enhance their vision.¹ The gaze position with minimal nystagmus intensity and better visual performance is known as the null position.^{1–7} Nearly all the research on vision in INS has focused on static visual acuity and the time needed to get the eyes onto the desired target (i.e., target acquisition time).^{8–13} While these are important properties, they are not sufficient to reveal more complex visual functions entailed in real-life visual activities. Therefore, it is important to study how individuals with INS perform when they carry out a range of visual tasks and to examine how performance is influenced by the variability of INS at different gaze positions.^{4,10,11,14}

In everyday life, we are constantly presented with objects in motion. These moving objects require us to identify them and to estimate their speed of motion. For example, when driving a car merging into traffic, accurate estimation of velocity difference between vehicles on the main

road is crucial to avoid accidents. This real-life visual activity demands accurate velocity discrimination.

To date, only one previous study by Shallo-Hoffmann et al.¹⁵ has investigated motion perception deficits in INS using a (vertical) discrimination task and a (horizontal and vertical) detection task. In the discrimination task, subjects were asked to indicate if test and reference gratings that moved vertically had the same or different velocities. The reference velocity was fixed (1, 3, or 6°/s), while the test velocities varied (either matched the reference velocity or being slower or faster by 15% or 30% of the reference velocity). The discrimination thresholds of accuracy were recorded and analyzed. In the detection task, subjects were required to identify the drift direction of either a vertically or horizontally moving grating. The detection thresholds were recorded and compared. Overall, findings from the study suggested that subjects with congenital nystagmus showed poorer discrimination performance (i.e., higher discrimination accuracy thresholds) compared to controls, and they had higher motion detection thresholds when the motion was parallel to nystagmus eye movements. However, in their



study, the investigators did not measure the velocity difference discrimination threshold, which is essential for accurate velocity estimation. In addition, they did not evaluate the effect of gaze at subjects' null positions or elsewhere on velocity discrimination performance.

The null position in INS is of interest since it is possible to have different performance at the null position or at some specific distance away from it. A recent study by Fadardi et al.¹⁴ demonstrated that increased cognitive demands can affect visual acuity of INS subjects, and the performance differed between different gaze positions. From low to high cognitive demand, the deterioration of acuity was greater for INS subjects at the null position compared to 15° away from it. The authors suggested that the larger effects at the null position might be due to the maximal foveation period duration at the null position, which allows more scope to deteriorate than at 15° away from it, where foveation may already be minimal. Thus, the null position in INS allows better performance at it than elsewhere.

In the present study, we investigated how individuals with INS perform velocity discrimination tasks. Particularly, we assessed how the null position affects their performance. To achieve this, we analyzed velocity discrimination thresholds at two different gaze positions for INS and control subjects. Three hypotheses were tested: 1) INS subjects will perform poorly compared to controls in velocity discrimination tasks; that is, INS subjects have higher velocity discrimination thresholds than controls; 2) Thresholds will be elevated more when the velocity discrimination task was performed in the same plane as the nystagmus; that is, INS subjects have higher horizontal than vertical velocity discrimination thresholds; 3) The null position in INS subjects will have a positive effect on velocity discrimination performance.

METHODS

Subjects

Twenty-one individuals with INS (mean age 24 years; range, 15–34 years) and 16 healthy control subjects (mean age 26 years; range, 22–39 years) were recruited from two testing sites (Melbourne, Australia, and Jinan, China). Eighteen subjects were classified as idiopathic INS and three had associated visual disorders (two had oculocutaneous albinism, and one had congenital cataract). The diagnosis of INS was first made by the referring ophthalmologists and later confirmed by the investigators with a pretesting clinical examination and analysis of eye movement recording analysis. Subjects with congenital periodic alternating nystagmus were identified by monitoring the nystagmus fast phase direction during their initial examination with extended primary gaze fixation for four minutes,¹⁶ and they were excluded from the study as they generally do not have a fixed null position. The healthy control subjects had to have a corrected visual acuity of 0.0 logMAR or better, and their interocular acuity difference was no more than one logMAR line. They had no history of ophthalmic, neurological, or psychotic illness, and were not taking any medications that could affect their eye movements.

This study complied with the Declaration of Helsinki and was approved by the Human Research Ethics Committees of the Department of Optometry & Vision Sciences, The University of Melbourne, and Shandong Liangkang Eye Hospital, Jinan (Ethics ID: 1749588.5). Informed consent

was obtained from the subjects after explanation of the nature and possible consequences of the study. For subjects under 18 years old, consent was sought from their parents/guardians.

Clinical Demographic Record

For all subjects, basic demographic information was collected before testing. This included age, gender, occupation, and medical history. A basic ophthalmic examination was performed to assess their visual functions. Distance visual acuity was measured at 3 m with a logMAR chart. Near visual acuity was determined at 40 cm using a reading chart. Stereopsis was measured by a Randot Stereotest. A cover test was performed to detect the presence of strabismus. Extraocular muscle excursions were determined at 40 cm in the standard nine cardinal position of gaze to detect any over- or underactions of the muscles. Abnormal head postures and the approximate null positions were also documented. Clinical characteristics of INS subjects are presented in [Table 1](#).

Apparatus

Subjects were seated at 75 cm from a computer monitor in a normally lighted room. The computer screen subtended a visual angle of 44° × 25° with a resolution of 2048 × 1152 pixels and a refresh rate of 60 Hz. Two eye trackers were used to record eye movements at different sites. In Melbourne, the Eyelink 1000 eye tracker (SR Research, ON, Canada) at a sampling frequency of 500 Hz was used, and in Jinan, a head mounted video eye tracker (SmoothEye, New York City, NY, USA) was used at a sampling frequency of 1000 Hz. The experimental protocol was designed and built using PsychoPy v1.85.4.¹⁷ Two metal arcs were made by the investigators to measure the gaze position of the subjects in both Melbourne and Jinan. It was mounted to the top edge of the monitor with targets at ±30° from the center in 5° steps as shown in [Figure 1](#). When subjects were asked to perform the task at 0° gaze position, they were required to put their chin on the chinrest with their eyes looking straight toward the 0° target at the center of the metal arc. When subjects were asked to perform the task at an eccentric gaze position, they were required to put their chin on the chinrest and then turn their head either leftward or rightward with their eyes looking straight toward the designated eccentric target on the metal arc to ensure they performed the task at the required eccentric gaze position.

Stimuli

Stimuli used for velocity discrimination tasks were generated by PsychoPy v1.85.4.¹⁷ The stimuli were sinusoidal gratings, which were presented at a spatial frequency of 0.5 cycles/deg and at a high contrast of 100% ([Fig. 2](#), See Supplementary Video S1 for a moving sinusoidal grating). The stimuli were presented on the screen within a Gaussian window with a diameter of 14.6 cm subtending 10° of visual angle. The gratings moved either horizontally (left or right) or vertically (up or down), which randomly varied from trial to trial within each task. The reference velocity of the stimuli was 5°/s, since INS subjects were reported to experience perceptual stability when retinal slip velocity was < 4°/s,^{18,19} and they were less accurate at discriminating stimulus veloc-

TABLE 1. Clinical Characteristics of INS Subjects

No.	Age/Sex	Diagnosis	Distance VA (logMAR)	Near VA	Stereopsis	AHP
1	21/M	Idiopathic INS	0.6	N8	(-)	(-)
2	16/F	Idiopathic INS, XT	0.6	N6	200"	(-)
3	24/M	Idiopathic INS, XT	0.6	N14	400"	(-)
4	22/M	Idiopathic INS	0.4	N8	400"	(-)
5	21/M	Idiopathic INS, ET	0.7	N14	(-)	(-)
6	15/M	Idiopathic INS	0.6	N14	(-)	Face turn L
7	31/M	INS, OCA	0.6	N14	400"	(-)
8	14/M	Idiopathic INS	0.4	N18	(-)	(-)
9	29/F	Idiopathic INS	0.0	N5	40"	Face turn L
10	26/F	Idiopathic INS	0.4	N6	(-)	(-)
11	26/M	Idiopathic INS, XT	0.2	N5	(-)	(-)
12	19/M	Idiopathic INS, XT	0.0	N4	(-)	(-)
13	30/M	Idiopathic INS	0.0	N4	(-)	Face turn L
14	22/M	Idiopathic INS	0.8	N14	(-)	(-)
15	21/F	Idiopathic INS	0.0	N4	25"	Face turn L
16	34/F	Idiopathic INS, XT	0.2	N4	(-)	Face turn R
17	28/F	Idiopathic INS, XT	0.7	N12	(-)	Face turn L
18	25/F	Idiopathic INS	0.2	N8	140"	(-)
19	22/M	Congenital cataract, INS	1.0	N24	400"	(-)
20	33/M	OCA, INS, ET	0.4	N10	(-)	(-)
21	23/M	Idiopathic INS	0.4	N6	(-)	(-)

M and F refer to male and female. Ages are of years. R and L refer to right and left direction. XT and ET refer to exotropia and esotropia, respectively. OCA refers to oculocutaneous albinism. AHP refers to anomalous head posture. (-) refers to no stereopsis or AHP in INS subjects. N4–N24, N referring to near; 4–24 corresponding to Times New Roman characters, font size 4–24; font size is measured in points; 1 point is equal to 1/72 of an inch.



FIGURE 1. A metal arc mounted to the top edge of the monitor with targets at $\pm 30^\circ$ from the center in 5° steps. It was used to measure the gaze position of the subject.

ities $< 4^\circ/\text{s}$, which might be a result of the proposed oscillopsia suppression mechanism.^{15,20}

Procedure

At the beginning of the task, a five-point pop-up calibration sequence (four around the periphery and one at the center of the screen) was performed binocularly. No validation procedure was performed. A chinrest and forehead rest were used to stabilize the head of subjects. Investigators also monitored the participants during the whole testing procedure to ensure that their heads were stabilized. For subjects with INS, it is not always possible to have their calibrations

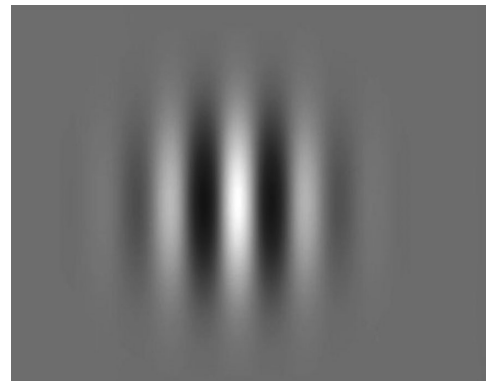


FIGURE 2. A sinusoidal grating employed as the stimulus in the velocity discrimination task.

validated since they are unable to fixate the targets stably for a sufficient period of time. In this case, the calibration was performed by a normally sighted calibrator. This has been reported to be a simple and easily applicable way to get relatively more accurate results compared with other alternative calibration methods.²¹ The calibration performed is sufficient for this study because the eye movement recordings were mainly used to confirm the diagnosis of INS, and to identify the presence and location of the null position. Once calibration was completed, the INS subject was required to fixate on a dot presented horizontally across $\pm 20^\circ$ from the center in 5° steps on the computer screen. Each gaze position was tested twice from right to left and then vice versa, with each presentation lasting for five seconds. Characteristics of INS waveform at the null position and 15° away from it are shown in Table 2. The gaze position with the least nystagmus intensity during this test was determined as the

TABLE 2. Characteristics of INS Waveform

No.	Null Position	Waveform	
		Primary Gaze	15° Away From Primary Gaze
1	(-)	0°/jerk, mixed direction	15°R/jerk, mixed direction
2	0°	0°/jerk R	15°R/jerk R
3	(-)	0°/jerk R	15°R/jerk R
4	(-)	0°/jerk, mixed direction	15°L/jerk L
5	0°	0°/jerk L	15°R/jerk R
6	20°R	20°R/no detectable nystagmus	5°R/jerk R
7	(-)	0°/jerk, mixed direction	15°R/jerk, mixed direction
8	0°	0°/jerk L	15°R/jerk R
9	15°R	15°R/jerk R	0°/jerk R
10	(-)	0°/jerk, mixed direction	15°R/jerk R
11	0°	0°/jerk L	15°R/jerk L
12	(-)	0°/jerk R	15°L/jerk R
13	20°R	20°R/no detectable nystagmus	5°R/jerk L
14	0°	0°/jerk, mixed direction	15°R/jerk, mixed direction
15	25°R	25°R/jerk R	10°R/jerk R
16	15°L	15°L/jerk R	0°/jerk R
17	15°R	15°R/jerk R	0°/mixed jerk
18	0°	0°/jerk L	15°L/jerk L
19	(-)	0°/jerk, mixed direction	15°L/jerk, mixed direction
20	(-)	0°/jerk, mixed direction	15°R/jerk, mixed direction
21	(-)	0°/jerk L	15°R/jerk L

(-) refers to no identified null position. R and L refer to right and left direction.

null position.^{22,23} Following this, all subjects were required to perform the velocity discrimination tasks.

Two velocity discrimination tasks were performed to measure the subject's velocity discrimination thresholds: 1) horizontal velocity discrimination task (gratings moving leftward or rightward); 2) vertical velocity discrimination task (gratings moving upward or downward). Within each task, velocity discrimination thresholds were measured at two gaze positions. For INS subjects with identified null positions, they performed at their null position and 15° away from it (either toward left or right). If the null position was in lateral gaze ($\pm 10^\circ$, $\pm 15^\circ$, or $\pm 20^\circ$), the 15° away position was in the opposite direction to it. If the null position was at or near primary gaze (0° or $\pm 5^\circ$), the 15° away from null position was either toward left or right. For INS subjects without identified null positions, they performed at primary (straight-ahead) gaze position and 15° eccentric position (either toward left or right). For control subjects, testing was done at primary (straight-ahead) gaze position and a 20° eccentric position (either toward left or right). The order of the two tasks was randomized, and within each task, the gaze positions were randomized.

For both tasks, each trial began when subjects were asked to fixate a black fixation dot ($1.44^\circ \times 1.44^\circ$ of visual angle). Following the fixation dot, subjects viewed a pair of gratings moving in the same direction, which were presented successively at different velocities. Each grating was presented for 650 ms, and the interval between the two gratings was 500 ms. After viewing the two gratings, subjects were asked to indicate which of the two gratings moved faster by pressing one of two designated buttons on the keyboard. Each response elicited an audio tone from the program. A correct response generated a high tone, and an incorrect response generated a low tone. Subjects were instructed about these tones so that they could be encouraged to be accurate and be alert. The threshold of each subject's motion perception was measured by a three-down/one-up two-alternative

forced-choice staircase procedure to estimate the 79.4% correct velocity difference discrimination.²⁴ The velocity of the reference grating was 5°/s. The initial velocity of the faster grating was set at 10°/s, which was a 100% velocity difference. The velocity difference between successive stimuli then was decreased by 30% of the current level if the subjects made three consecutive correct responses or increased by 30% of the current level if a single incorrect response was made. The experimental session terminated after eight reversals of staircase directions. The velocity discrimination thresholds were determined by a Weber fraction ($\Delta V/V$), of which ΔV is defined as the just-noticeable difference between two gratings and V is the reference velocity of the stimuli. The threshold was calculated by the last six of eight reversal point values of the Weber fraction ($\Delta V/V$).

Before formal testing began, each subject received several practice trials to ensure they understood the task procedure. The practice trials started from 100% velocity difference of which the faster grating was easy to detect. Subjects were also told the task would become harder, and that it was important that they tried their utmost to identify which one moved faster and pressed the key as accurately as possible. However, if some trials of the task were too difficult for them to discriminate which one moved faster, they were instructed to guess.

Statistical Analysis

We analyzed velocity discrimination thresholds utilizing SPSS version 21 (IBM Corporation, Armonk, NY, USA) and GraphPad Prism Version 8 for Windows (GraphPad Software, San Diego, CA, USA). An outlier analysis (ROUT ($Q = 1\%$)) was used to detect the outliers, and outlier values were removed for subsequent analyses.²⁵ Two-way mixed ANOVAs were used to measure the effect of INS

on the velocity discrimination task performance. Two-way repeated-measures ANOVAs were used to measure the effect of the null position and stimulus motion direction on the velocity discrimination performance. The eye movements at different gaze positions were recorded, and the direction of the slow phase of jerk waveform was noted along with the direction of horizontal stimulus motion (leftward or rightward) during the task. Velocity discrimination thresholds when these directions were concordant or discordant were compared using a two-tailed paired *t*-test.

RESULTS

The velocity discrimination thresholds measured by horizontal and vertical tasks were analyzed for control subjects (primary gaze (straight-ahead) and 20° eccentricity), and INS subjects (primary gaze (null or straight-ahead if no null present), and 15° eccentricity). One outlier was removed from leftward discrimination threshold data at primary gaze position for INS subjects. The thresholds are shown in percentage of Weber fraction ($\Delta V/V$ (%)). Data were presented as mean \pm SD%.

When comparing the discrimination thresholds between control and INS subjects, a 2-way mixed ANOVA showed that the INS subjects had significantly higher horizontal (right and left) ($37.59 \pm 18.56\%$) and vertical (up and down) ($28.12 \pm 12.40\%$) thresholds than the control subjects (horizontal: $19.85 \pm 10.06\%$, vertical: $19.75 \pm 9.39\%$) at both primary and eccentric gaze positions (Fig. 3 and Fig. 4) (Primary: F [1, 35] = 16.30, P = 0.0003; Eccentric: F [1, 35] = 15.34, P = 0.0004).

As this study aimed to investigate the effect of the null position on velocity discrimination in INS, the INS group was further divided into two subgroups: 1) 12 INS subjects with a null (subgroup A), 2) Nine INS subjects without a null (subgroup B). A 2-way repeated-measures ANOVA showed

that subgroup A had significantly lower horizontal ($27.00 \pm 7.90\%$) and vertical ($23.08 \pm 8.58\%$) thresholds at the null position than at 15° away from it (horizontal: $37.61 \pm 18.08\%$, vertical: $28.89 \pm 9.23\%$) (Fig. 3 and Fig. 4) (Horizontal: F [1, 11] = 7.859, P = 0.0172); Vertical: F [1, 11] = 8.035, P = 0.0162). For subgroup B and the control group, 2-way repeated-measures ANOVAs showed no differences between different gaze positions (Subgroup B: F [1, 8] = 1.407, P = 0.2695, Control group: F [1, 15] = 2.656, P = 0.1240).

When comparing horizontal and vertical thresholds within INS and control subjects, a 2-way repeated-measures ANOVA revealed the INS subjects had significantly higher horizontal thresholds compared to vertical thresholds at both primary and eccentric gaze positions (Fig. 5) (F [1, 41] = 24.99, P < 0.0001). There were no differences for the control subjects (F [1, 31] = 0.006949, P = 0.9341).

Since the INS subjects had their nystagmus only in the horizontal plane, thresholds measured in the horizontal task (right and left) were compared with regard to the nystagmus slow phase direction for INS subjects. Thirteen INS subjects at primary gaze and 16 INS subjects at eccentric gaze had pure jerk left or right nystagmus that were analyzable (see Table 2 for data presentation). A two-tailed paired *t*-test showed that when stimulus motion was in the same direction as the nystagmus slow phase, thresholds ($38.34 \pm 19.67\%$) were not significantly different from the thresholds of when stimulus motion direction was opposite to the nystagmus slow phase direction ($38.08 \pm 21.38\%$) (Fig. 6) (t [28] = 0.09718, P = 0.9233).

The effect of visual acuity on velocity discrimination thresholds was investigated. Pearson correlation and linear regression analyses showed no correlation between acuity and velocity discrimination thresholds in INS subjects (horizontal thresholds: r = 0.1676, P = 0.4801, vertical thresholds: r = 0.3034, P = 0.1934). This result demonstrated that visual

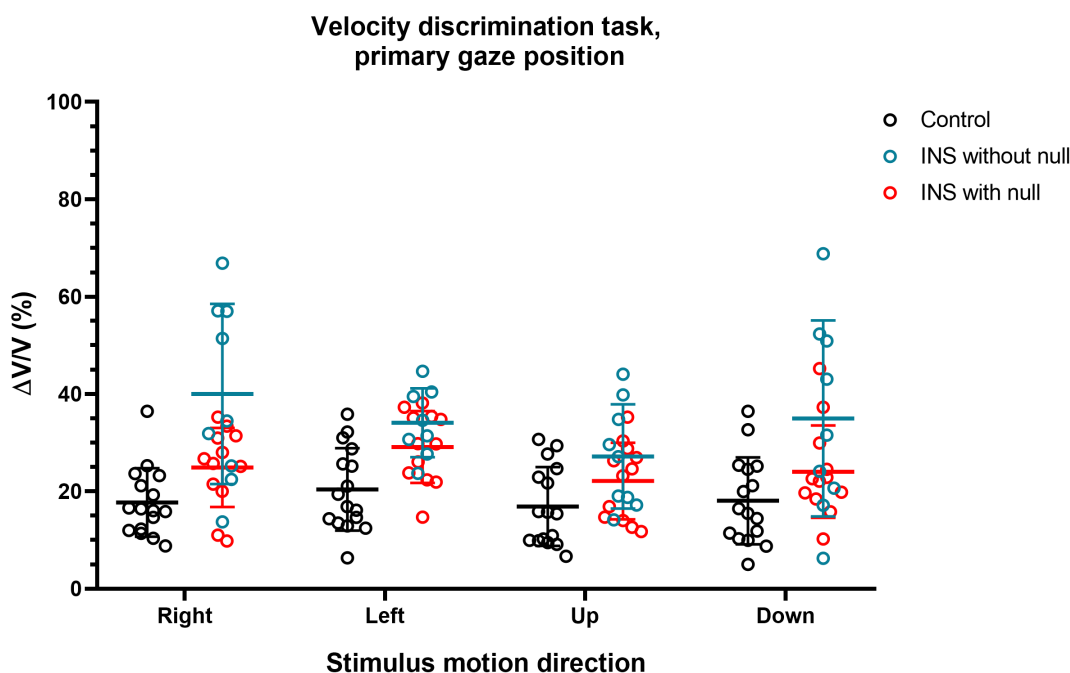


FIGURE 3. Horizontal (right and left) and vertical (up and down) velocity discrimination thresholds for control and INS subjects at primary gaze position. Error bars indicate standard deviation, which holds for the following figures.

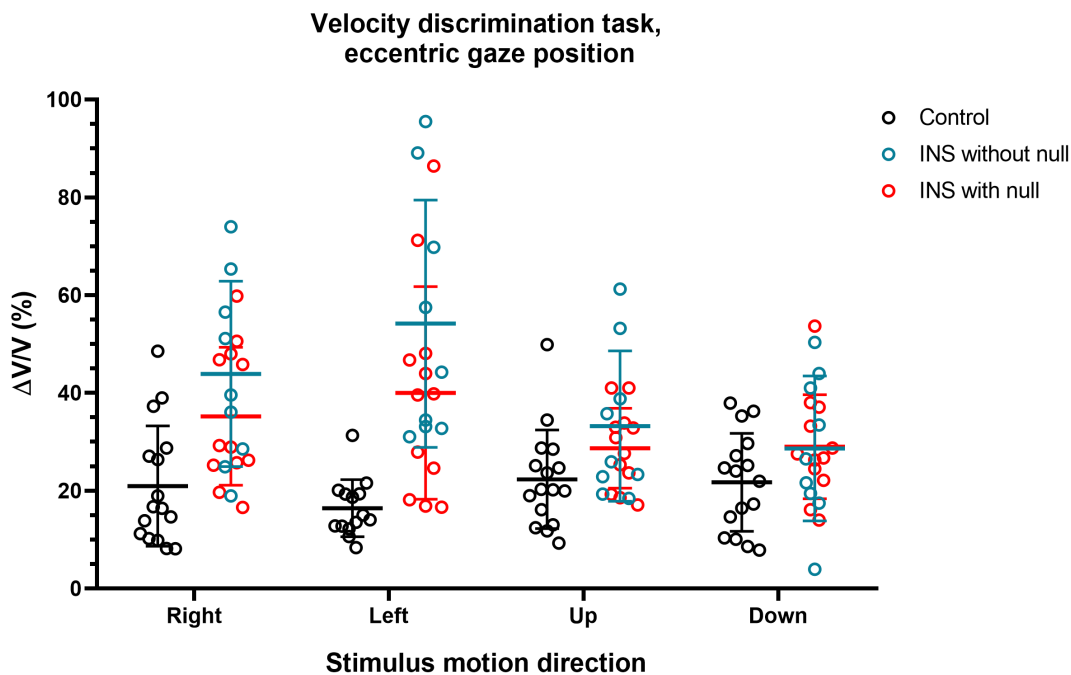


FIGURE 4. Horizontal (right and left) and vertical (up and down) velocity discrimination thresholds for control and INS subjects at eccentric gaze position.

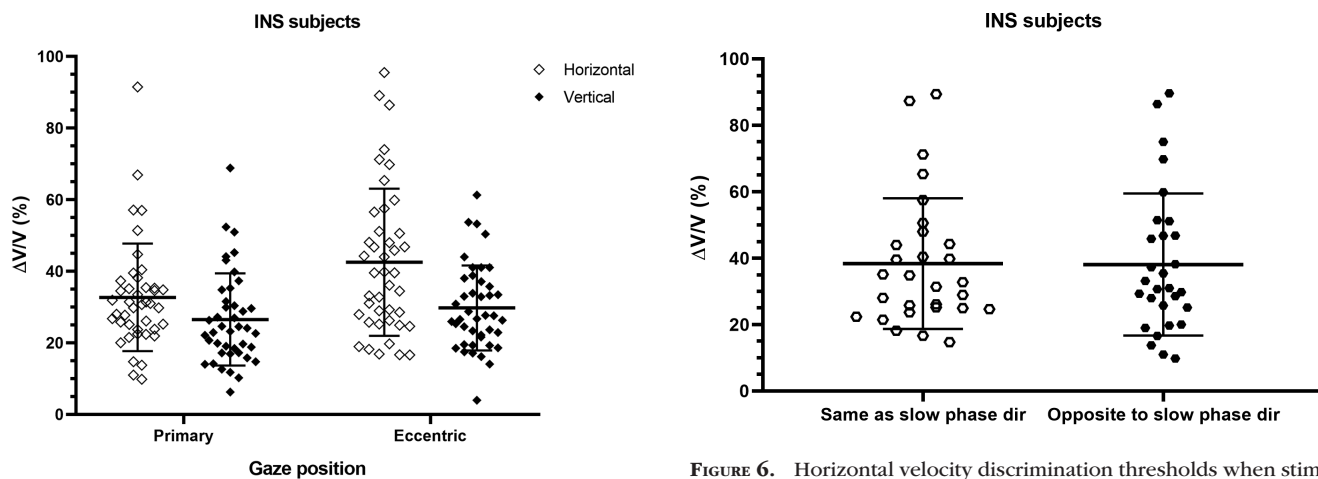


FIGURE 5. Horizontal and vertical motion velocity discrimination thresholds at primary and eccentric gaze positions for INS subjects.

FIGURE 6. Horizontal velocity discrimination thresholds when stimulus motion direction was in the same direction as the nystagmus slow phase direction (dir) or was opposite to the nystagmus slow phase direction (dir).

acuity of INS subjects did not affect their velocity discrimination thresholds.

DISCUSSION

Poorer Velocity Discrimination Performance of INS Subjects Compared to Controls

As hypothesized, INS subjects showed poorer velocity discrimination performance (i.e., elevated velocity discrimination thresholds) compared to controls for both horizontal and vertical motion directions.

Though the findings in the present study were partly in agreement with Shallo-Hoffmann et al.,¹⁵ the method of obtaining the discrimination thresholds was different.

Shallo-Hoffmann et al.¹⁵ measured the accuracy of group responses of velocity discrimination and estimated discrimination thresholds by fitting a Gaussian curve to group responses. In the present study, we measured the Weber fraction for each subject and assessed discrimination thresholds by using a staircase psychophysical method.

In the study by Shallo-Hoffmann et al.,¹⁵ they suggested that the poorer performance of congenital nystagmus subjects compared to controls may be attributed to a mechanism that may be used to avoid oscillopsia at a cost of sensitivity to motion in INS subjects. Hence, it can be speculated that the mechanism of oscillopsia suppression could be a possible explanation for the elevated velocity discrimination thresholds of INS subjects in our study.

For INS subjects, despite having incessant retinal image motion, they rarely report oscillopsia. Several mechanisms have been proposed to account for this perceptual stability in INS subjects. A most widely accepted mechanism of oscillopsia suppression is that retinal image motion is canceled by an efferent copy of the extraretinal signal.^{26,27} Extraretinal signals have been demonstrated to accompany involuntary eye movements in individuals with INS.²⁷ These extraretinal signals were proposed to play a role in alleviating motion smear in subjects with INS and contribute to the absence of oscillopsia in INS.²⁷

Bedell et al.²⁶ assessed the extraretinal signal in four subjects with nystagmus by requiring them to point in the direction of a flashed target in darkness when presented at various phases of the nystagmus waveform. They reported that extraretinal signals were available for approximately 75% of the eye position changes in INS and suggested that the extraretinal signals contributed to the oscillopsia suppression. In another study, Bedell²⁷ evaluated visual performance in persons with INS and normal observers when presented with similar retinal image motion (e.g., target moved at 8°/s). Normal observers reported profound target movement and motion smear, while persons with congenital nystagmus perceived a relatively stable and clear visual world. These differences were attributed to the extraretinal signals that accompany the involuntary eye oscillations reducing the motion smear in subjects with INS.

Another proposed mechanism of oscillopsia suppression is the elevation of the motion detection threshold.²⁸ It has been reported by Dieterich et al.²⁹ that individuals with INS had elevated thresholds for detecting motion at peripheral and central locations compared to controls. Shallo-Hoffmann et al.¹⁵ also demonstrated that, when identifying the drift direction of a horizontally moving grating, INS subjects showed elevated detection thresholds. Leigh et al.²⁸ induced oscillopsia in four INS subjects by stabilizing images on the retina under different conditions and suggested that several mechanisms operate to maintain perceptual stability in INS. Possible mechanisms included the use of extraretinal signals to cancel out the effects of eye motion and the elevation of the motion detection threshold.

In summary, the various proposed mechanisms of oscillopsia suppression are suggested to operate together to reduce the sensitivity of externally caused retinal motion in INS subjects. This would lead to difficulty in detecting the difference in target velocity, hence resulting in elevated velocity discrimination thresholds in INS subjects.

Poorer Horizontal Than Vertical Velocity Discrimination Performance in INS Subjects

In this study, INS subjects had significantly higher thresholds when the velocity discrimination task was performed in the same plane as the nystagmus. Although no previous study investigated horizontal and vertical velocity discrimination in INS, anisotropies between horizontal and vertical have been stated for motion detection in INS. Bedell³⁰ measured thresholds for detecting horizontal and vertical motion of a light dot target in INS subjects and reported that thresholds were elevated more for horizontal than vertical motion. Shallo-Hoffmann et al.¹⁵ assessed motion detection performance using both horizontally and vertically moving gratings and found that INS subjects had higher thresholds when the motion was parallel to nystagmus eye movements.

In the present study, though the experimental setup and method of analysis were different from the abovementioned studies, the higher horizontal than vertical discrimination thresholds in INS subjects detected in the current study were consistent with previous findings.^{15,30} A possible explanation for these findings is that thresholds are elevated more for motion in the meridian of eye movement because the constant movement of the retinal image caused by nystagmus renders additional movement of the target difficult to detect.

Null Position Effect

It was proposed that the null position would have a positive effect on velocity discrimination performance in INS. Results of this study showed significantly reduced velocity discrimination thresholds at the null position compared to 15° away from it in INS subjects.

Velocity discrimination has been stated to improve with the duration of the motion.³¹ This may suggest that the better velocity discrimination performance at the null position might be due to its longer foveation duration compared to 15° away from it in INS subjects. However, for 18 of the 21 INS subjects who were tested in Jinan, the eye tracker was not available at the time of testing to document their waveforms. It would be of benefit to further explore the correlation between foveation duration and velocity discrimination thresholds for INS subjects at the null position and 15° away from it to better understand the null position effect.

This finding also raises the general question about why individuals with INS prefer to use their null position. A recent study by Dunn et al.¹⁰ assessed the impact of the null position on visual acuity in subjects with idiopathic INS and reported that although the improvement in visual acuity at the null position was statistically significant, its magnitude (0.08 logMAR) was much smaller than might be expected from the larger improvement in nystagmus parameters like foveation duration. So why do individuals with INS adopt an abnormal head posture, if they gain only very small improvement in visual acuity at the null? This might be driven by improvements in multiple aspects of visual function, such as velocity discrimination, visual processing time, or recognition time.

Velocity Discrimination in the Real-Life Activities of INS Subjects

Velocity discrimination is often required for real-life activities that involve motion. One example is driving, an activity that is of vital importance in daily life. To date, several studies have evaluated visual function in INS with respect to real-life activities including driving using questionnaires.^{32,33} McLean et al.³² reported that 19 of the 21 interviewees with nystagmus had difficulties with driving, and they discussed that the reduction in visual acuity in nystagmus may account for the difficulties of driving. Das and coworkers³³ found that nearly half of their participants with INS (17/35) met the driving standard of a visual acuity of 0.3 logMAR in the UK. However, only seven of them were regular drivers at the time of the interview. This suggested that the reduced visual functioning in driving was not only associated with reduced visual acuity but also a result of other impaired aspects of visual function in INS. The elevated velocity discrimination thresholds shown in the present study could be one factor

accounting for the difficulties of driving for INS subjects. Participation in ball sports may be hindered not only by reduced visual acuity but also by difficulties in estimating the velocity of the ball or of other players. Even forms of computer gaming require accurate velocity estimates of game elements. Findings of the present study could help us to further understand how people with INS perform daily visual activities and assist us in developing new clinical visual function assessment tools for INS, since assessing visual performance of INS patients solely by measuring their visual acuity in the clinic tends to underestimate the effect of INS on visual function in real-life activities.

In summary, velocity discrimination was impaired in INS subjects, with elevated thresholds seen for both horizontal and vertical motion. Furthermore, the thresholds were elevated more for horizontal motion. These findings suggest that the mechanisms employed to suppress oscillopsia in INS elevated motion discrimination thresholds, especially for the motion in the same meridian as the nystagmus. The null position had a positive effect on discrimination thresholds. This is another visual function which is improved at the null, in addition to the sometimes modest improvement in visual acuity.¹⁰ It would be of great benefit to investigate the correlation between nystagmus parameters (e.g., foveation duration, intensity) and discrimination thresholds in a future study where recording during the task was available to better understand the underlying mechanism of null position effect in velocity discrimination for INS subjects.

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References

1. Abadi RV, Whittle J. The nature of head postures in congenital nystagmus. *Arch Ophthalmol*. 1991;109(2):216–220.
2. Abadi RV, Worfolk R. Retinal slip velocities in congenital nystagmus. *Vision Res*. 1989;29(2):195–205.
3. Stevens DJ, Hertle RW. Relationships between visual acuity and anomalous head posture in patients with congenital nystagmus. *J Pediatr Ophthalmol Strabismus*. 2003;40(5):259–264.
4. Abadi RV, Bjerre A. Motor and sensory characteristics of infantile nystagmus. *Br J Ophthalmol*. 2002;86(10):1152–1160.
5. Dell'Osso LF, Daroff RB. Congenital nystagmus waveforms and foveation strategy. *Doc Ophthalmol*. 1975;39(1):155–182.
6. Dell'Osso LF. Congenital, latent and manifest latent nystagmus—similarities, differences and relation to strabismus. *Jpn J Ophthalmol*. 1985;29(4):351–368.
7. Dell'Osso LF, Flynn JT, Daroff RB. Hereditary congenital nystagmus. An intrafamilial study. *Arch Ophthalmol*. 1974;92(5):366–374.
8. Barot N, McLean RJ, Gottlob I, Proudlock FA. Reading performance in infantile nystagmus. *Ophthalmology*. 2013;120(6):1232–1238.
9. Dunn MJ, Margrain TH, Woodhouse JM, Ennis FA, Harris CM, Erichsen JT. Grating visual acuity in infantile nystagmus in the absence of image motion. *Invest Ophthalmol Vis Sci*. 2014;55(4):2682–2686.
10. Dunn MJ, Wiggins D, Woodhouse JM, Margrain TH, Harris CM, Erichsen JT. The effect of gaze angle on visual acuity in infantile nystagmus. *Invest Ophthalmol Vis Sci*. 2017;58(1):642–650.
11. Hertle RW, Maybodi M, Reed GF, Guerami AH, Yang D, Fitzgibbon EJ. Latency of dynamic and gaze-dependent optotype recognition in patients with infantile nystagmus syndrome versus control subjects. *Ann N Y Acad Sci*. 2002;956(1):601–603.
12. Wang ZI, Dell'Osso LF. Eye-movement-based assessment of visual function in patients with infantile nystagmus syndrome. *Optom Vis Sci*. 2009;86(8):988–995.
13. Wang ZI, Dell'Osso LF. Being “slow to see” is a dynamic visual function consequence of infantile nystagmus syndrome: model predictions and patient data identify stimulus timing as its cause. *Vision Res*. 2007;47(11):1550–1560.
14. Fadardi MS, Bathke AC, Harrar SW, Abel LA. Task-induced changes in idiopathic infantile nystagmus vary with gaze. *Optom Vis Sci*. 2017;94(5):606–615.
15. Shallo-Hoffmann JA, Bronstein AM, Acheson J, Morland AB, Gresty MA. Vertical and horizontal motion perception in congenital nystagmus. *Neuro-Ophthalmology*. 1998;19(4):171–183.
16. Gradstein L, Reinecke RD, Wizov SS, Goldstein HP. Congenital periodic alternating nystagmus. *Ophthalmology*. 1997;104(6):918–929.
17. Peirce JW. PsychoPy—psychophysics software in Python. *J Neurosci Methods*. 2007;162(1–2):8–13.
18. Abel LA, Williams IM, Levi L. Intermittent oscillopsia in a case of congenital nystagmus. Dependence upon waveform. *Invest Ophthalmol Vis Sci*. 1991;32(12):3104–3108.
19. Dell'Osso LF, Leigh RJ. Ocular motor stability of foveation periods: required conditions for suppression of oscillopsia. *Neuro-Ophthalmology*. 1992;12(5):303–326.
20. Bedell HE, Bollenbacher MA. Perception of motion smear in normal observers and in persons with congenital nystagmus. *Invest Ophthalmol Vis Sci*. 1996;37(1):188–195.
21. Harrar V, Le Trung W, Malienko A, Khan AZ. A nonvisual eye tracker calibration method for video-based tracking. *J Vis*. 2018;18(9):13.
22. Abadi RV, Dickinson CM. Waveform characteristics in congenital nystagmus. *Doc Ophthalmol*. 1986;64(2):153–167.
23. Dell'Osso LF. Fixation characteristics in hereditary congenital nystagmus. *Am J Optom Arch Am Acad Optom*. 1973;50(2):85–90.
24. Wetherill G, Levitt H. Sequential estimation of points on a psychometric function. *Br J Math Stat Psychol*. 1965;18(1):1–10.
25. Motulsky HJ, Brown RE. Detecting outliers when fitting data with nonlinear regression—a new method based on robust nonlinear regression and the false discovery rate. *BMC Bioinformatics*. 2006;7:123.
26. Bedell HE, Currie DC. Extraretinal signals for congenital nystagmus. *Invest Ophthalmol Vis Sci*. 1993;34(7):2325–2332.
27. Bedell HE. Perception of a clear and stable visual world with congenital nystagmus. *Optom Vis Sci*. 2000;77(11):573–581.
28. Leigh RJ, Dell'Osso LF, Yaniglos SS, Thurston SE. Oscillopsia, retinal image stabilization and congenital nystagmus. *Invest Ophthalmol Vis Sci*. 1988;29(2):279–282.

29. Dieterich M, Brandt T. Impaired motion perception in congenital nystagmus and acquired ocular motor palsy. *Clin Vis Sci*. 1987;1(4):337–345.
30. Bedell HE. Sensitivity to oscillatory target motion in congenital nystagmus. *Invest Ophthalmol Vis Sci*. 1992;33(5):1811–1821.
31. Vaina LM, Gryzwacz NM, Saiviroonporn P, LeMay M, Bienfang DC, Cowey A. Can spatial and temporal motion integration compensate for deficits in local motion mechanisms? *Neuropsychologia*. 2003;41(13):1817–1836.
32. McLean RJ, Windridge KC, Gottlob I. Living with nystagmus: a qualitative study. *Br J Ophthalmol*. 2012;96(7):981–986.
33. Das A, Quartilho A, Xing W, et al. Visual functioning in adults with idiopathic infantile nystagmus syndrome (IINS). *Strabismus*. 2018;26(4):203–209.