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Visual Fixation in Chiari Type II Malformation

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Abstract

Chiari type II malformation is a congenital deformity of the hindbrain. Square wave jerks are horizontal involuntary saccades that interrupt fixation. Cerebellar disorders may be associated with frequent square wave jerks or saccadic oscillations such as ocular flutter. The effects of Chiari type II malformation on visual fixation are unknown. We recorded eye movements using an eye tracker in 21 participants with Chiari type II malformation, aged 8 to 19 years while they fixated a target for 1 minute. Thirty-eight age-matched healthy participants served as controls. Square wave jerks' parameters were similar in the 2 groups. Saccadic oscillations were not seen. Chiari type II malformation is not associated with pathological square wave jerks or abnormal saccadic oscillations. The congenital nature of this deformity may permit compensation that preserves stable visual fixation. Alternatively, the deformity of Chiari type II malformation may spare parts of the cerebellum that usually cause fixation instability when damaged.

Keywords

visual fixation; square wave jerks; saccadic intrusions; saccadic oscillations; Chiari type II malformation

Chiari type II malformation is a congenital deformity of the brainstem and cerebellum that is associated with spina bifida. In Chiari type II malformation, the posterior fossa is small and its contents are distorted. The inferior vermis herniates below foramen magnum and the superior part of the cerebellar vermis is shifted upwards. Cerebellar weight and volume are decreased. Hydrocephalus requiring shunt diversion occurs in over 85% of cases of Chiari type II malformation.[1–3]

Square wave jerks are horizontal, involuntary pairs of saccades that take the fovea away from the intended position of fixation, and then return the fovea back to the fixation position.[4] Square wave jerks and ocular flutter, the latter defined as bursts of closely spaced or back-to-back saccades in one plane during fixation, are examples of saccadic intrusions and saccadic oscillations that cause fixation instability. Square wave jerks typically have amplitudes between 0.5° and 3° (though they may be up to 10°), and durations of about 200 ms in adults.[5,6] The presence of square wave jerks with larger amplitude and more than

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9,5,7 or 16,[8,9] square wave jerks per minute have been suggested to be abnormal. Abnormal square wave jerks are reported in diseases that affect the cerebellum, brainstem, or cerebral cortex.[4,7,10] Ocular oscillations, for example, ocular flutter, occur in diseases that affect the brainstem and cerebellum.[4]

The effects of the deformity of Chiari type II malformation on square wave jerks and fixation stability have not been previously investigated. Our goal was to determine if pathological square wave jerks and other saccadic oscillations are present in children and adolescents with Chiari type II malformation and to investigate their characteristics.

Methods

Participants

Twenty-one participants (10 females) with spina bifida, Chiari type II malformation, and shunted hydrocephalus were selected randomly from a cohort of participants, who were participants in a spina bifida project funded by the National Institute of Child Health and Human Development. Their mean age (SD) was 14.3 (3.2) years. None had blurred vision, nausea, vomiting, or vertigo. Ten participants had nonparalytic strabismus. All participants had normal pupillary reaction to light, fundus exam, and other cranial nerves function on examination. All participants had lower motor neuron lesion signs on examining their lower limbs including hypotonia, weakness, and reduced or absent deep tendon jerks. Sensory examination of their lower limbs revealed absent sensation to pin prick, vibration, and joint position senses. Nine participants could ambulate but none had a normal gait.

Thirty-eight typically developing children and adolescents (17 females) in the control group were recruited by local advertising. Their mean age (SD) was 13.8 (3.4) years. Three participants had nonparalytic strabismus.

All participants in this investigation had best corrected monocular visual acuity of at least 20/40, a Verbal or Performance IQ of 70 or above. Exclusion criteria were visual field defect on visual field confrontation testing, paralytic strabismus, nystagmus on clinical examination that was present within 30° of central gaze, ear disease, psychiatric disorder, medication with drugs that might interfere with eye movements, symptomatic hydrocephalus, ocular or neurological disorders unrelated to Chiari type II malformation, and syringobulbia on magnetic resonance imaging (MRI).

The study was in accord with the declaration of Helsinki guidelines. Ethical approval for this project was obtained from the Research Ethics Boards at the Hospital for Sick Children and the University Health Network, Toronto. Written consent was obtained from participants or their legal guardian.

Spinal lesion level—Within the spina bifida group, each participant's spinal lesion level was determined from physical examination. Two groups were distinguished: Lower spinal lesion level group (L1 and below) and upper spinal lesion level group (T12 and above), based on the developmental process of neural tube closure.[11] Recent studies on spinal lesion level show that upper spinal lesions are associated with more brain anomalies on MRI, worse behavior and intellectual outcomes.12,13

Hydrocephalus—Three shunt groups were created. Group 1 had no shunt revisions (N = 5), group 2 had 1 shunt revision (N = 9), and group 3 had 2 or more shunt revisions (N = 7) as described previously.[12,14–17]

Nystagmus—Eight participants had nystagmus that was clinically apparent *only* in eccentric gaze. In 7 of these participants, we found on eye movement recording a low amplitude (<2°) and predominantly horizontal gaze-evoked nystagmus. The effect of nystagmus was investigated.

Equipment and Procedures

The eye tracker—An infrared eye tracker (El Mar Inc, Downsview, ON, Canada) was used to record eye position. The image was sampled at 120 Hz. The system accuracy was 0.5°. The system was free from drift and had a resolution (ie, minimum detectable movement) of 0.1°.[18] Head movements were recorded using a magnetic head tracker (Flock of Birds, Ascension Technology Corp, Burlington, Vt).

Each participant was seated on a chair facing a 45 cm monitor (Samsung, SyncMaster 900 NF), located 57 cm from the participant's cornea. The participant's head was stabilized using a chin rest and adjusted so that the eyes were in the central position when looking at the center of the monitor. The visual target was a 2-mm, white square light that subtended 12 minutes of arc. Its luminance was 65 candela (cd)/m^2 while the monitor background luminance was 0.01 cd/m^2. Eyeglasses were removed because they interfere with the eye tracker. The uncorrected visual acuity in all cases was adequate for seeing the stimulus clearly. Eye movement positions were calibrated for each eye with the fellow eye occluded at 14 fixation light points that were arrayed along the horizontal and vertical axes. Participants' performance and alertness were monitored by TV and by an oscilloscope display of horizontal and vertical eye movements to provide feedback.

Task—An eye patch was used to cover the nonpreferred eye to ensure uniform viewing conditions in all participants. Movements of the viewing eye were measured. Participants were instructed to watch a centrally located target continuously for 1 minute as described elsewhere.[19]

Processing of the Eye Movement Data

Eye and head movements were digitized for offline analysis. Square wave jerks were marked automatically and checked manually using cursors controlled by a computer. A square wave jerk was defined as a saccade, equal to or greater than 0.5° in amplitude that was directed away from the fixation target, followed by a second saccade in the opposite direction that refixated the target, following a period ranging from 100 to 500 ms. Saccades were defined as fast eye movements that had peak velocities > 20°/s. The beginning and end of saccades were marked when eye velocity crossed 20°/s. Duration of square wave jerks was defined as the time interval between the onset of the error-producing (initial) saccade and the completion of the error-correcting saccade. Peak velocity was the maximum saccadic velocity of each square wave jerk saccade.[19] The eye movement traces were also checked carefully to look for other saccadic intrusions or oscillations such as square wave pulses (formerly called macro square wave jerks), defined as horizontal large amplitude (4°–30°) fast eye movements to one side of fixation and back; macro saccadic oscillations, defined as horizontal runs of saccades that straddle the intended fixation position; and ocular flutter.4

Analyses

For each participant, the frequency of square wave jerks per minute, direction (ie, right or left) and mean amplitude of the initial error-producing saccades, mean amplitude and mean duration of all their square wave jerk saccades, were calculated.[19] Peak saccadic velocity varies with small saccadic amplitudes in a linear fashion. Because almost all square wave jerk amplitudes in this study fell within a very narrow range (between 0.5° and 1.8°), mean

peak velocity of all square wave jerk saccades was calculated for this small amplitude range for each participant. Analyses were performed using a Statistical Package for Social Sciences (SPSS Inc, 2001, Chicago, Ill). Normality of data distribution was tested using the mean, median, SD, skewness, kurtosis, and box plots.[20]

Square wave jerks were compared between the control and Chiari type II malformation groups using independent, Student *t* tests for normally distributed data or the Mann-Whitney *U* test for nonparametric data. Square wave jerks' parameters were correlated with age and number of shunt revisions using Spearman's test.[20] Differences in square wave jerks' parameters based on gender, spinal lesion level, and nystagmus were investigated using independent two-tailed, Student *t* tests for normally distributed data or the Mann-Whitney *U* test for nonparametric data. For all tests, significance was defined by *P* values < .05.[20]

Results

All participants completed the task. The frequency of square wave jerks, durations, peak velocities, or amplitudes did not have normal distribution among participants within each group. Therefore, median values are given. An example of square wave jerks is shown in Figure 1. Square wave jerks' characteristics are shown in Table 1.

Square wave jerks occurred more commonly in the control group (89.5%) than in the Chiari type II malformation group (66.7%, *P* = .04). However, square wave jerks' frequencies (ie, the number of square wave jerks/min) in those who had square wave jerks, and their amplitudes, peak velocities, or durations were not significantly different between the control and Chiari type II malformation groups (Table 1).

There were 3 single saccades with amplitudes larger than 2° in 2 participants with square wave jerks in the control group. These saccadic amplitudes were between 2.4° and 4.1° and their peak velocities were between 159° and 189°/s. One of these 2 participants had 2 square wave jerks in 1 minute and the other one had 14 square wave jerks in 1 minute. Mean square wave jerks' amplitudes (mean peak velocities) in these 2 participants were 1.7° (138°/s) and 0.97° (70°/s). There were only 2 single saccades with amplitudes slightly larger than 2° in 1 participant with square wave jerks in the Chiari type II malformation group. These saccadic amplitudes were 2.3° and 2.8° and their peak velocities were 130° and 161°/s. This participant had 9 square wave jerks in 1 minute. Mean square wave jerks' amplitude (mean peak velocity) was 1° (69°/s).

The frequency of square wave jerks, their duration, peak velocity, or amplitude did not correlate with age in either group. Gender, spinal lesion level, or nystagmus did not significantly affect square wave jerks' parameters. However, square wave jerks' durations increased with increasing number of shunt revisions on Spearman's correlation tests (correlation coefficient = 0.628, *P* = .016, N = 14; Figure 2).

Other types of saccadic intrusions or oscillations, for example, square wave oscillations (ie, regular periodic square wave jerks), square wave pulses, macro saccadic oscillations, and ocular flutter were not recorded in any participant.

Discussion

Square wave jerks occur in most typically developing children and adolescents. They also occur in many children and adolescents with Chiari type II malformation. Participants with Chiari type II malformation did not have a pathological increase in square wave jerks' frequency or amplitude as reported previously in some cerebellar diseases.[7,10] Other saccadic intrusions or saccadic oscillations were also not recorded in any participant with

Chiari type II malformation. The congenital and chronic nature of Chiari type II malformation may permit adaptation that preserves relatively stable visual fixation. Chronic nonprogressive lesions of the cerebral cortex are also less likely to produce square wave jerks.[7] Alternatively, the deformity of Chiari type II malformation may spare parts of the cerebellum that usually causes significant fixation instability when damaged.

The second refixation saccade had a latency (reflected in square wave jerks' duration) that increased with increasing number of shunt revisions. More shunt revisions, a surrogate marker of the cumulative effects of raised intracranial pressure,[12,15] may damage parts of the brain involved in saccadic processing, including the frontal and parietal lobes, and cause an increase in the latency of refixation saccades.

Normal and abnormal ocular motor functions have been documented in children and adolescents with Chiari type II malformation.[21–23] This may reflect the anatomic substrate affected by the deformity of Chiari type II malformation and could potentially explain the normal prevalence, frequency, and amplitude of square wave jerks and the lack of abnormal saccadic oscillations in Chiari type II malformation that we found in this investigation. Saccadic amplitude accuracy and saccadic adaptation, which are both processed in vermis lobules VI and VII and the fastigial nuclei, are typically normal in most children and adolescents with Chiari type II malformation.[21,22] However, smooth ocular pursuit gain is subnormal in children and adolescents with Chiari type II malformation and nystagmus but not in children and adolescents with Chiari type II malformation who do not have nystagmus, likely reflecting floccular and parafloccular cerebellar lobe involvement.[23] The location of the anatomical substrate of pathological square wave jerks in cerebellar disorders is unknown. The anatomical substrate of pathological square wave jerks is unlikely to be located in the floccular and parafloccular lobes, because the presence of nystagmus did not influence square wave jerks' parameters. Vermis lobules VI and VII and the fastigial nuclei are important for processing saccades.[24] Because square wave jerks consist of intrusive saccades, we postulate that this part of the cerebellum is also important in processing square wave jerks. Chiari type II malformation is typically not associated with abnormal saccades,[21] and the absence of pathological square wave jerks or other saccadic intrusions and oscillations, is in accord with normality of other saccadic functions in this malformation.

It will be informative to record eye movements during fixation in children with acquired cerebellar diseases that affect different parts of the cerebellum to localize the anatomic site responsible for producing pathological square wave jerks and other saccadic oscillations.

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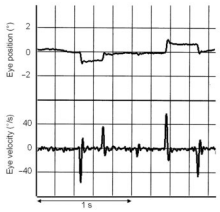

Figure 1.

The figure shows 2 square wave jerks recorded from the right eye of an 8-year-old girl with Chiari type II malformation fixating a visual target. The upper trace shows eye position. Upward deflection represents rightward movement. The bottom trace shows the corresponding eye velocity.

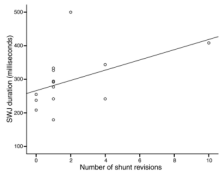

Figure 2.

A scatter plot illustrating duration of square wave jerks (SWJs) and number of shunt revisions. Significant correlation was found between the 2 variables. The equation of the fitted line is $Y = 15.5X + 265$, $R^2 = .23$.

Table 1

Square Wave Jerks' (SWJ) Characteristics [range] in the Control and Chiari Type II Malformation Groups

	Control Group	Chiari II Group	*P* value
Number of participants with SWJ (%)	34 (89.5)	14 (66.7)	.04
Median number of SWJ/min in those with SWJ	3 [1–18]	3.5 [1–17]	.38
Median number of initial rightward SWJ/min	2 [0–11]	1.5 [0–5]	.51
Median number of initial leftward SWJ/min	1.5 [0–12]	1 [0–13]	.40
Median amplitude of the initial SWJ saccades in °	0.83 [0.5–1.6]	0.75 [0.53–1.1]	.25
Median amplitude of SWJ in °	0.81 [0.53–1.7]	0.74 [0.6–1]	.18
Median peak velocity of SWJ in °/s	60 [30–138]	54 [34–80]	.59
Median duration of SWJ in ms	249 [125–421]	284 [179–500]	.12