Intermittent Oscillopsia in a Case of Congenital Nystagmus

Dependence Upon Waveform

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Spontaneous reports of oscillopsia are rare in cases of congenital nystagmus (CN). We examined the relationship between nystagmus waveform characteristics and oscillopsia in one such case. To reduce the patient's nystagmus, she was fitted with contact lenses. We examined the effects of tactile feedback by applying local anesthetic while she wore the lenses. When she was without lenses, we provided tactile feedback by applying gentle finger pressure to one eyelid. She was also asked to look at a peripheral afterimage. Nystagmus was analyzed for frequency, amplitude, foveation duration, and drift velocity, if foveation was not perfectly stable. Perceived target stability was recorded. The patient noted oscillopsia during the initial baseline recording and with lid pressure. The image was stable with contact lenses with and without anesthesia and during the second session baseline; at these times, drift velocity was <4°/sec and foveation duration was >100 msec. No oscillopsia of the afterimage in dark was noted; she perceived it moving with her gaze as she attempted to look at it.

It appears that in some CN patients, the suppression of oscillopsia operates only within fixed limits of foveation stability and duration. When, because of internal or external factors, their nystagmus exceeds these, oscillopsia results. Invest Ophthalmol Vis Sci 32:3104-3108, 1991

One of the features generally believed to be pathognomonic of congenital nystagmus (CN) is that individuals with CN are normally free of oscillopsia. As has been noted by Leigh et al, this requires the maintenance of a stable visual percept in the face of oscillations in which slow phase velocities may exceed 100°/sec. These far exceed those generally seen in acquired nystagmus, yet the latter is usually associated with oscillopsia, while in CN it is quite rare. Several mechanisms have been suggested to account for this stability of the perceived world in the face of nearly constant motion across the retina. These include visual information sampling only during foveation periods, with suppression at other times; use of an extraretinal signal to cancel out the visual effects of eye motion; central elevation of the motion detection threshold; and post-saccadic backward masking of motion. Separation of the contributions made to suppression of oscillopsia by any or all of the above mechanisms has been difficult.

Oscillopsia occasionally has been noted in real-world conditions in some individuals with CN. Two of the subjects in the above study reported some instances of oscillopsia. One experienced it at gaze angles when nystagmus was maximal. Similarly, Abadi and Dickinson noted that patients with congenital periodic alternating nystagmus (PAN) invariably noted oscillopsia at those points in their cycle when their nystagmus peaked. Kelly et al described two subjects with CN that was present only while they followed leftward-moving objects. During these times, oscillopsia was present. In each of the above instances, CN patients reported oscillopsia only when their nystagmus was present at levels not normally encountered. In none were specific nystagmus characteristics such as frequency, amplitude, foveation time, or foveation stability quantified. The present case is the first in which these have been examined in a CN patient with intermittent oscillopsia and have been related to its presence or absence. Our patient also exemplifies the degree to which the vision of some CN patients may be affected by changes in their nystagmus as a result of mental set.

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Case Report

A 14-year-old-girl known to have CN presented in 1989 with oscillopsia, which had been present for one week after one day of frontal headache. She complained of having had intermittent headache during the previous 12 months and also described intermittent attacks of oscillopsia in previous years. The examination revealed best visual acuity of 6/6 OD (right eye), 6/5 OS (left eye). The color vision, visual fields, and optic fundi were normal. The patient had jerk nystagmus that increased in intensity on gaze to right and left and damped on convergence. She did not exhibit an obvious null angle. The patient stated that in the past she turned her head to the left or right to see better but that self-consciousness about this abnormal head posture led her to keep her head straight. In the primary position, a 2 diopter exotropia was noted. Other congenital abnormalities included the absence of the left kidney and the development of three sets of teeth. The examination in other respects was normal.

Because spontaneous reports of oscillopsia in congenital nystagmus are rare and because the patient described recurrent headache, an ocular motor problem superimposed on the existing CN was suspected. Quantitative eye movement recordings were performed. By the second eye movement recording session, her oscillopsia had resolved. The intermittent headache was considered to be a result of tension. After the first session, the patient, who was quite self-conscious about her nystagmus, was reassured that nothing in her test results suggested any new problem. She was also told that her nystagmus was not conspicuous cosmetically. She was encouraged to not be concerned if her classmates commented on it. She seemed to accept these suggestions, and this may have contributed to the improvement seen in her nystagmus during the second recording session.

Methods

Eye movements were recorded using infrared oculography. The patient sat in a dimly lit room in a chair with a head rest and forehead restraint and viewed red light-emitting diodes mounted on an arc 2 m in radius positioned the same distance in front of her. After obtaining written informed consent from the patient, eye movements were recorded on a rectilinear chart recorder (Graphtec Corp., Japan). The eye movement signals were electronically differentiated and the resulting velocity traces were recorded. The bandwidth of the entire system was DC-85 Hz.

There were two recording sessions, two weeks apart. Calibration was always done monocularly at ±15° and was checked whenever test conditions were changed. For analysis, left eye fixation of the primary position target was compared under the different test conditions. The patient viewed targets between ±15°, in 5° steps. This was done to look for a null angle within this range. She then was asked to fixate the 0° light for over a minute while viewing with her left eye, which had superior visual acuity. She was fitted with a pair of soft contact lenses (−1.0 OD, −0.5 OS) and fixation was recorded. To evaluate the relative importance of tactile feedback provided by the lenses via the touch receptors of the eyelids, a local anesthetic was administered and fixation repeated.

On a second occasion, the patient again viewed the primary position LED with her left eye. After a minute, she was asked to gently place the little finger of her right hand against her right eyelid to enhance any sensations of eye movements detected by the touch receptors of that lid. Left eye fixation of the 0° target was again recorded, as above.

After these recordings were made, an afterimage was placed approximately 15° nasally from her left macula with an ophthalmoscope while she sat in a darkened recording area with no targets present. She was asked to describe its behavior as she attempted to look at it in the dark.

At each point in the study where a test condition was changed, the patient was asked if the target was steady or moving and her response was noted on the chart recording. This enabled us to relate changes in her nystagmus to the perceived stability of her visual environment.

Data were analyzed by evaluating 20-sec segments of each of the five test conditions. Portions were selected starting at least 10 sec into a new test period to allow for the development of a steady state condition. The amplitude of each nystagmus beat was measured and, when the waveform was jerk with extended foveation, the duration of each foveation period was measured. Many beats did not exhibit perfect foveation but began with a relatively constant, low velocity drift that subsequently ran away with exponentially increasing velocity. Therefore, velocities and durations of these initial drifts were noted as well. This broadened the definition of foveation to include those portions of the slow phase during which retinal image motion was reduced but not eliminated. The slow phase still consisted of two components, one of which was an exponential runaway and the other a constant drift. Nystagmus frequency also was computed, using a time interval of 20 sec minus whatever time was spent in blinks or without nystagmus.

For the afterimage presentation, no quantitative analysis was performed. Rather, as the patient’s gaze changed as she attempted to “look at” the afterimage, her description of its motion was noted on the recording and related to her eye movements.

Results

The nystagmus waveform seen in this patient was largely jerk with extended foveation, with periods of jerk beats. There was no indication in her eye movement recordings of any acquired ocular motor disturbance superimposed upon her CN. Nystagmus wave-
forms and behavior (eg, damping with convergence and increasing with effort to see) were pathognomonic of congenital nystagmus.

A summary of the measurements made on the patient's nystagmus is given in Table 1. In two of the test conditions she reported oscillopsia and in three, stable perception of the visual world. The presence of oscillopsia always corresponded to average drift velocities greater than 4°/sec and foveation durations below 100 msec. As expected, her nystagmus improved when contact lenses were applied, with concomitant abolition of the continuous oscillopsia that she noted under the initial test condition. Surprisingly, however, her nystagmus further improved when local anesthetic was administered.

In the second session, her nystagmus initially was quite minimal and stable vision was noted. Gentle finger pressure upon her eyelid, however, brought about a marked deterioration in her nystagmus, along with an immediate report of oscillopsia. During this latter condition, her foveation durations were the lowest seen. Comparisons of drift and duration in oscillopsia versus nonoscillopsia conditions using an analysis of variance showed significant differences on both (p < .05). Examples of the patient's nystagmus at points where oscillopsia was present or absent are given in Figure 1.

When asked to look at the extrafoveal afterimage, the patient's gaze moved to the left as she attempted to pursue the temporally perceived target. Her gaze then moved smoothly with her slow phases to the right (Fig. 2).

Her perception of the afterimage's motion reflected the overall direction of her gaze rather than the beat-to-beat motion of her ocular oscillation. Thus, whether she moved smoothly to the right by letting her slow phases go uncorrected or moved saccadically to the left while her slow phases drifted to the right, she perceived the afterimage moving along with her direction of gaze, rather than changing on a beat-to-

<table>
<thead>
<tr>
<th>Test condition</th>
<th>Nystagmus amplitude (°)</th>
<th>Nystagmus frequency (Hz)</th>
<th>Average drift velocity (°/sec)</th>
<th>Average foveation duration (msec)</th>
<th>Oscillopsia</th>
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</thead>
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<tr>
<td>1st baseline</td>
<td>4.3 ± 1.3</td>
<td>3.6</td>
<td>9.7 ± 4.7</td>
<td>58.1 ± 15.8</td>
<td>Yes</td>
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<tr>
<td>With contact lenses</td>
<td>3.0 ± 1.1</td>
<td>3.0</td>
<td>4.0 ± 2.9</td>
<td>107.8 ± 55.2</td>
<td>No</td>
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<tr>
<td>With contact lenses and local</td>
<td>1.8 ± 0.6</td>
<td>3.3</td>
<td>2.0 ± 1.7</td>
<td>130.2 ± 54.2</td>
<td>No</td>
</tr>
<tr>
<td>2nd baseline</td>
<td>2.0 ± 0.7</td>
<td>2.3</td>
<td>2.1 ± 1.7</td>
<td>116.1 ± 67.5</td>
<td>No</td>
</tr>
<tr>
<td>With finger pressure</td>
<td>3.3 ± 1.1</td>
<td>3.4</td>
<td>5.4 ± 4.0</td>
<td>39.4 ± 48.3</td>
<td>Yes</td>
</tr>
</tbody>
</table>

Table 1. Nystagmus characteristics vs oscillopsia

Fig. 1. Examples of the patient's nystagmus when (A) oscillopsia was present, during the first session baseline, and (B) when it was absent, with contact lenses in and local anesthetic administered.

Nystagmus amplitude, frequency, average drift velocity during foveation periods, duration of foveation periods, and presence or absence of oscillopsia are given for the five test conditions. All conditions in which oscillopsia was absent had drifts during foveation averaging less than 4°/sec and durations greater than 100 msec.
beat basis. During this condition, her nystagmus waveform often was pure jerk, with no foveation periods.

Discussion

The relationship between nystagmus and oscillopsia is complex, when the latter is and is not present. In the presence of acquired ocular motor disturbances, the degree of oscillopsia noted is often less than the amount of retinal slip actually occurring. In congenital nystagmus, oscillopsia is rarely reported and, when it is, it is generally in unusual positions of gaze or in periods of increased nystagmus intensity (amplitude \times frequency). Intensity provides no information concerning the presence of periods of little or no retinal slip during a nystagmus beat. Therefore, it might be expected to relate poorly to oscillopsia, in the way that the relationship between intensity and visual acuity is weak. However, nystagmus parameters related to retinal image stability would be expected to bear a more direct relationship to oscillopsia, as has been shown for visual acuity. This has not to our knowledge been previously examined in a patient with otherwise typical CN.

If the primary mechanism for suppression of oscillopsia were the use of an extra-retinal signal (either proprioceptive or corollary discharge), dependence of oscillopsia upon waveform characteristics would not be expected a priori. Whatever the nature of the slow phase, it would be subtracted from the visual signal, leaving a stable percept. Furthermore, the use of an extra-retinal signal for nystagmus cancellation would be expected to induce oscillopsia when an afterimage is viewed in the dark. This was not the case in our patient or in the two subjects studied by Leigh et al, who occasionally reported “real world” oscillopsia.

Although conclusively identifying all of the relevant mechanisms used in the suppression of oscillopsia is impossible, our quantitative assessment of the CN in this patient, coupled with her careful reports of her perceptions, provide some additional information about this phenomenon. While correlation does not prove causality, the critical elements appear to be a retinal slip during foveation of <4°/sec, along with a duration >100 ms. Whether duration or retinal slip alone was the critical factor could not be determined, because more stable foveations were invariably longer in our patient. In a case with some similarities to ours, a CN patient who developed intermittent oscillopsia after a sudden loss of consciousness perceived image movement when minimum slow phase velocities were >4°/sec and position errors were >0.5° in a jerk right waveform. Perceptual stability was experienced when the waveform was jerk left with extended foveation and both position and velocity errors fell below the preceding values. This case, coupled with the fact that many CN patients have foveation times well below 100 msec and still experience perceptual stability, suggests that foveation duration is not the critical factor. It seems likely that patients such as ours and the one described above make lesser use of extraretinal feedback, as evidenced in our patient by her failure to see an afterimage as oscillating. Temporal low-pass filtering has been suggested as the cause of reduced perceived afterimage motion in normals as saccade frequency increases. The reduced extraretinal feedback proposed in our patient could be caused by an abnormal low-pass cutoff frequency and could account for afterimage stability.

It is difficult to determine to what extent visual sampling occurred during foveation periods. While Jin et al found that detection of brief flashes was not more likely during foveation than during other portions of the slow phase, a difference in sensitivity during foveation could still exist, at least in some patients, including those susceptible to periods of oscillopsia. Such patients may rely more on other means of suppression, including an increase in the motion detection threshold and on visual sampling. Gottlob et al found that retinally stabilized flashes presented at different points in the CN slow phase were associated perceptually with their correct spatial, not retinal, locations. This suggests that at least some CN patients may suppress oscillopsia through a continual remapping of their visual space. Perhaps patients such as ours can do this only when sufficiently stable visual input is available.

In the case reported here, oscillopsia arose whenever the nystagmus waveform continuously fell outside specific velocity and duration criteria, whether these changes were a result of external manipulations
such as lid pressure or from internal causes such as anxiety or self-consciousness about nystagmus. Such a limited suppression mechanism is clearly not present in all CN patients, because many with far worse waveforms than our patient’s least stable periods still do not note environmental movement. These limits relate to mean values of drift velocity and duration. While the standard deviations of these measures in our patient were such that values for particular beats under different test conditions were identical, perception corresponded to the average value for that trial, rather than varying on a beat-to-beat basis. This integration across beats is consistent with the normal perceptual continuity experienced by CN patients whose visual experience is uninterrupted by the presence several times a second of retinal slip velocities that may exceed 100°/sec.

This case also reflects the effects of variability in a patient’s CN on clinical intervention, as seen in our patient’s seemingly paradoxical response to tactile feedback. She had previously expressed a desire for contact lenses, predisposing her toward them. Perhaps because of her embarrassment when made aware of her nystagmus, the same tactile feedback that has been reported to reduce CN in other patients12 exacerbated hers. This led to an apparently optimal state with lenses and anesthesia and a pronounced deterioration when tactile feedback via finger pressure was provided. Whether all of the CN patients who sometimes experience oscillopsia under normal conditions are similarly sensitive to becoming conscious of their nystagmus merits further study, as does the foveation behavior of other CN patients who note oscillopsia only under exceptional conditions.

Key words: nystagmus, CN, oscillopsia, visual perception

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