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Vision Research 43 (2003) 2697–2705

Vision
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Effects of stimulus size and luminance on oscillopsia in congenital nystagmus

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Received 12 December 2002; received in revised form 3 April 2003

Abstract

Although the absence of oscillopsia is a common feature of congenital nystagmus (CN), it is occasionally noted by patients under poor viewing conditions and has been provoked in laboratory settings with stabilised images. In the present study, the effects of reductions in background stimulus size and luminance on perceptual stability in CN were examined. Sixteen CN subjects were first interviewed using a structured questionnaire about whether they ever experienced oscillopsia and, if so, under what circumstances and with what perceptions. They next fixated an LED centred in projected images of three sizes ($21 \times 14^\circ$, $10 \times 6^\circ$ and $7 \times 4^\circ$) and four luminance levels (115.5, 24.5, 2.7 and 0.1 cd/m², with contrasts from 96 down to 20%). Eye movements were recorded with a limbal tracker. They were asked after viewing each image “whether anything happened to the image while they watched it.”

Occasional oscillopsia was reported by 12/16 of the CN subjects on the questionnaire. In the laboratory, 13/16 subjects experienced oscillopsia in some manner for at least one of the stimuli. 8/13 CN subjects experienced it for the dimmest and smallest slides. 11/13 perceived certain parts (either the LED or background) of the visual stimuli as moving, with the perception of LED movement most pronounced at low background luminance. Foveation did not differ when trials with and without reported oscillopsia were compared (independent samples *t*-test, $p > 0.05$).

Oscillopsia may occur in CN with normal viewing of bright fixation targets against dim backgrounds. Under these conditions, the oscillopsia may be spatially inhomogeneous. Luminance differences between the fixation point and surround may have caused transmission time differences as the image moved across the retina, therefore leading to the perception of motion in one portion of the scene and not the other.

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Keywords: Congenital nystagmus; Oscillopsia; Luminance; Contrast; Efference copy; Foveation

1. Introduction

Congenital nystagmus (CN) is an involuntary ocular oscillation presenting at birth or shortly afterwards (Abadi & Bjerre, 2002; Dell’Osso, 1985). In normal individuals, perceptual stability is maintained only as long as retinal image motion is less than approximately $4^\circ/\text{s}$ (Bedell & Currie, 1993; McKee & Welch, 1985). In CN, despite slow phases that may exceed $100^\circ/\text{s}$, perceived motion of the environment—oscillopsia—is seldom a complaint and individuals report spatial constancy. Indeed, Leigh, Dell’Osso, Yaniglos, and Thurston (1988) reported that fewer than ten of the more than 450 CN subjects tested in their laboratory spontaneously re-

ported oscillopsia. In contrast to CN, oscillopsia is a common complaint of subjects with acquired types of nystagmus despite the similarly moving retinal images (Grünfeld, Morland, Bronstein, & Gresty, 2000). However, careful questioning of CN patients may elicit the comment that sometimes the environment is seen to move, particularly if gaze is directed away from the null position into the non-preferred direction (Leigh et al., 1988), if their nystagmus has worsened due to stress or anxiety (Abel, Williams, & Levi, 1991) if the visual environment is low-contrast and relatively unstructured (Tusa, Zee, Hain, & Simonsz, 1992) or among patients with congenital periodic alternating nystagmus, if their nystagmus has reached a peak in its cycle (Abadi & Dickinson, 1986). Indeed, our preliminary report of this study (Abel & Tkalcevic, 2001) noted that 75% of subjects reported at least *occasional* oscillopsia in daily life. A recent retrospective study of the records of 224

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patients with CN and LMLN found that 39% had experienced oscillopsia whilst viewing binocularly (Abadi & Bjerre, 2002).

Several mechanisms have been suggested that could contribute to the perceived stability of the visual world in CN. The most widely supported is the use of an extra-retinal signal (Abadi, Whittle, & Worfolk, 1999; Dell'Osso, Averbuch-Heller, & Leigh, 1997; Goldstein, Gottlob, & Fendick, 1992) to cancel out the effects of the retinal image motion. Such signals include efference copy and, to a lesser degree, proprioceptive input from the extraocular muscles. This is supported by the observation that a stabilised image on the retina of a CN patient will give rise to the perception of motion (Leigh et al., 1988). Other proposed mechanisms include a dependence upon times when the eyes are moving relatively slowly during foveation periods, with a degree of suppression at other times (Abadi & Worfolk, 1989; Dell'Osso & Leigh, 1992a, 1992b), reduced sensitivity to retinal image motion (Dieterich & Brandt, 1987), post-saccadic backward masking of motion signals (Leigh et al., 1988) and adaptation to the retinal image motion (Shallo-Hoffmann, Bronstein, Acheson, Morland, & Gresty, 1998). It remains to be determined to what extent each mechanism contributes to preventing oscillopsia. Some individuals appear to utilise one mechanism more than others (Abel et al., 1991).

Anecdotal evidence suggests that the size, visual structure and luminance levels of a scene are important in helping prevent the breakdown of perceptual stability in CN. Tusa et al. (1992) also described some unusual nystagmus patients with CN-like waveforms who could voluntarily suppress their nystagmus in a well lit environment but who developed more prominent nystagmus and oscillopsia in a dimly lit environment. Although oscillopsia has been induced by stabilising the retinal image in CN patients either via the use of afterimages or by optical means (Leigh et al., 1988), to date no studies have systematically varied physical characteristics of visual stimuli presented under normal, unstabilised viewing conditions. Leigh et al. (1988) observed that when several subjects viewed a scene whose centre was optically stabilised and whose periphery was seen normally, either the surround or centre was seen to move. Although this is a highly unnatural stimulus, comments made by some individuals with CN that they at times experience oscillopsia of only part of their visual environment suggest that oscillopsia suppression may not necessarily be spatially homogeneous. In the present study we systematically varied the size, brightness and contrast of stimuli to determine whether changes in any of these parameters would provoke oscillopsia in subjects with CN and, if so, what the nature of the perceived motion was. We also analysed the subjects' foveation during viewing to determine whether loss of perceptual stability was related to reduced foveation.

2. Methods

The eye movements of 16 subjects with CN, aged between 9 and 20 years (median age, 12.4 years), were examined. The diagnosis of CN was initially made by the referring ophthalmologist and was confirmed on the basis of clinical examination and eye movement recording analysis carried out by the authors. Eleven subjects were classified as idiopathic and five as albino (two ocular, three oculocutaneous) based on their clinical records. Seven individuals, aged between 11 and 26 years (median age, 19.3 years), with no oculomotor or visual abnormalities served as controls. All participants were naive with respect to eye movement measuring techniques and the experimental hypotheses of the study. Written informed consent was obtained according to the declaration of Helsinki. Pertinent characteristics of the subjects are given in Table 1.

Prior to testing, a questionnaire (Appendix A) was administered to each CN subject to ascertain whether they ever had or currently did experience oscillopsia. It was later used to compare these real world situations of oscillopsia with their perceptions in the laboratory.

Eye movements were recorded using a Microguide binocular infrared oculographic system (Kumar & Krol, 1992). Horizontal eye position data were digitised at 400 Hz with a 12-bit analogue to digital converter for later off-line analysis. Testing was done without refractive correction and no subject wore contact lenses. Eye movements were calibrated by sequentially presenting light emitting diodes (LEDs) from -19° to $+19^\circ$ mounted on an arc 160 cm in radius and positioned the same distance in front of the subject. Fixation data were scaled using a best-fit regression line. A chin and headrest were used to stabilise head position during recording.

Following calibration, subjects were instructed to fixate the steadily illuminated 0° LED of the arc. The stimulus presentation lasted 5 s. At the completion of this 5 s trial, the researcher asked subjects, "did anything happen to the light whilst watching it?" The wording was chosen to ensure that it did not imply oscillopsia. Responses were recorded.

Subjects then viewed images projected on a wall-mounted 184 cm by 184 cm white screen, positioned 280 cm from them. Image size was varied in an effort to determine whether this contributed to a breakdown in perceptual stability. The images subtended $21^\circ \times 14.3^\circ$, $10^\circ \times 5.7^\circ$ and $6.5^\circ \times 3.7^\circ$. A fixation LED subtending 0.1° and with a luminance of 443 cd/m^2 was positioned in the centre of the screen. The slides consisted of random black and white shapes of varying sizes (Fig. 1). The slide projector was fitted with one fixed and one rotatable polaroid circular filter (HOYA 52 mm polarising filter). The moveable filter, when turned, controlled the amount of light emitted from the projector.

Table 1
Clinical data for the 16 CN subjects

Subj.	Age and sex	Clinical diagnosis	Binocular visual acuity (LogMAR)	Waveform	Ampl. (°)	Freq. (Hz)	Null angle (°)	Foveation (% $\pm 2^\circ$ and $\leq 4^\circ/\text{s}$)
JD	11 M	Idiopath	0.1	Jef	2.2	3.5	5	22.4
LT	15 F	Idiopath	0.3	PC	1.6	4	5	24
VC	14 F	Idiopath	0.5	PC	8.9	3.5	-10	3.9
NP	9 F	Idiopath	0.4	PPfs	7.3	5	0	28
CE	14 M	Idiopath	0.2	Pfs	2.3	5.5	-5	11
SH	9 M	Idiopath	0.5	J	9.6	3.5	0	7.3
SM	9 M	Idiopath	0.5	PP	1.8	4	10	17.4
MM	9 M	Idiopath	0.9	PC & DJ	2.5	7.5	5	12
MW	20 M	Idiopath	0.1	Jef	0.75	4	0	44
DM	12 M	Idiopath	0.2	J	1.1	6.5	5	10.3
DH	9 M	Idiopath	0.2	J	3	6.5	5	14
KS1	11 F	TPOCA	0.5	PC & J	1.64	3.5	0	20
PJ	12 M	TNOCA	0.7	PC & DJ	13.2	4	5	3
JM	15 M	TNOCA	0.6	Jef	1.7	4	5	42
KS2	18 M	TPOCA	0.6	PC	1.7	4.5	0	5.4
JS	11 F	TPOCA	0.5	Pfs	1.3	3.5	5	20

Subj. = subject; Ampl. = amplitude; Freq. = frequency; TPCOA = tyrosinase-positive oculocutaneous albino; TNOCA = tyrosinase-negative oculocutaneous albino. Nystagmus waveforms were: jerk (J), jerk with extended foveation (Jef), pseudo-cycloid (PC), pseudo-pendular with foveating saccades (PPfs), pendular with foveating saccades (Pfs), and dual jerk (DJ).

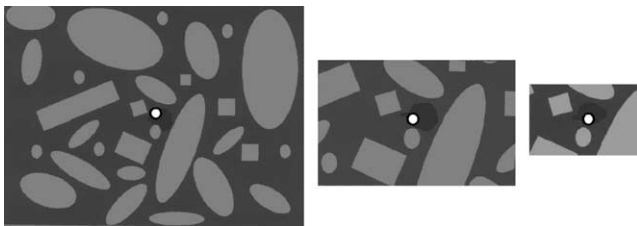


Fig. 1. Representations of the visual stimuli presented. Actual sizes were $21^\circ \times 14.3^\circ$, $10^\circ \times 5.7^\circ$ and $6.5^\circ \times 3.7^\circ$, respectively. The white circle in the centre represents the green LED.

Maximum transmission was 115.5 cd/m^2 when the filters were aligned and minimum transmission was 0.1 cd/m^2 when they were at 90° . Four levels of attenuation were used.

A photographic light meter (set to 50 ASA) was used to determine exposure value (EV) light meter readings in the bright and dark areas of the three slides, so that contrast could be calculated. Contrast was calculated as $(L_{\text{max}} - L_{\text{min}})/(L_{\text{max}} + L_{\text{min}})$. EV light meter readings obtained by the photometer were converted to luminance values (cd/m^2) (Wyszecki & Stiles, 1982). Because both light and dark regions of the image were affected by the filters, the resultant four contrasts were 96%, 94.6%, 93% and 20% as maximum luminance was set at 115.5, 24.5, 2.7 and 0.1 cd/m^2 . When the stray light from the projector and the additive light given from the central LED were subtracted from the original light meter readings during the 0.1 cd/m^2 setting, contrast increased from 20% to 82%. The luminance ratio between the fixation target and the dimmest surround was 4430:1; the luminance ratios for the other three backgrounds and the centre were 164:1, 48.1:1 and 3.8:1,

respectively. On the outside of the slide projector, markers were attached to the fixed polaroid filter to ensure that stimuli were consistent across subjects.

Subjects were instructed to fixate the LED in the centre of the screen. The viewing time allowed for each slide at each contrast setting was 10 s. Subjects first viewed the $21^\circ \times 14.3^\circ$ slide at the minimum luminance setting to help avoid afterimages. Subjects were allowed a 60 s rest between each slide setting to further ensure that afterimages did not elicit a perception of oscillopsia. After viewing the image, subjects were asked “did anything happen whilst watching the slide?” Subjects’ perceptions were recorded. The recording sequence for this study took between 15 and 20 min. The control subjects served to determine whether the autokinetic illusion would affect their perception of the stability of the stimuli.

Eye movement data were analysed for changes in the stability and duration of foveation periods, and whether the CN waveform itself changed during times of oscillopsia. Foveation periods were defined as those periods of the eye movement recording during which eye velocity was $\leq 4^\circ/\text{s}$ and eye position $\pm 2^\circ$ from the point of fixation from cycle to cycle. This positional criterion of $\pm 2^\circ$ was less stringent than the usual $\pm 0.5^\circ$ position setting used in past studies to account for albino subjects who lack a functional fovea (Dell’Osso & Jacobs, 2002; Mezawa, Ishikawa, & Ukai, 1990; Ukwade & Bedell, 1992). Foveation periods were determined by manually positioning the cursor through the beginning of as many slow phases as possible in a given interval of fixation. Points that met the position and velocity criteria for that segment were identified. Blinks and non-fixation points were excluded from analysis.

3. Results

3.1. Questionnaire results

Nine of the eleven idiopathic subjects and three of the five albino subjects experienced occasional real world oscillopsia under specific conditions. The circumstances under which subjects experienced it were varied but almost always related to the object of regard rather than the surrounding scene (Question 2). Dim lighting was relatively frequently associated with the experience (Question 8), as were fatigue or illness (Question 14). Breaking fixation or looking into the preferred null position were effective ways for some subjects to end these periods of perceived motion around them (Question 6). The results for all subjects are summarised in Appendix A.

3.2. Experimental results

The key finding of the study was that some subjects reported that oscillopsia only affected specific parts of

their visual field; e.g., only the central LED appeared to oscillate whilst the background remained motionless. Others saw the background moving whilst the central LED remained stationary. On the other hand, some subjects experienced both the LED and peripheral shapes as moving simultaneously. Some subjects with oscillopsia consistently saw the same parts of the slide oscillating whilst others perceived different oscillating areas of different images. No normal subject perceived motion for any condition.

Trials where oscillopsia was reported were analysed to determine whether background luminance influenced perceptual stability. Reports from the 13 subjects who experienced oscillopsia for at least one stimulus presentation are summarised in Fig. 2. Here, each bar reflects how many times each stimulus of a given size and background luminance/contrast was reported to be seen to move. Reports of movement of the LED, the background or both together are illustrated in Fig. 2A, B and C, respectively. Examination of the influence of background size and background luminance for each of the oscillopsia categories illustrated in Fig. 2 was attempted

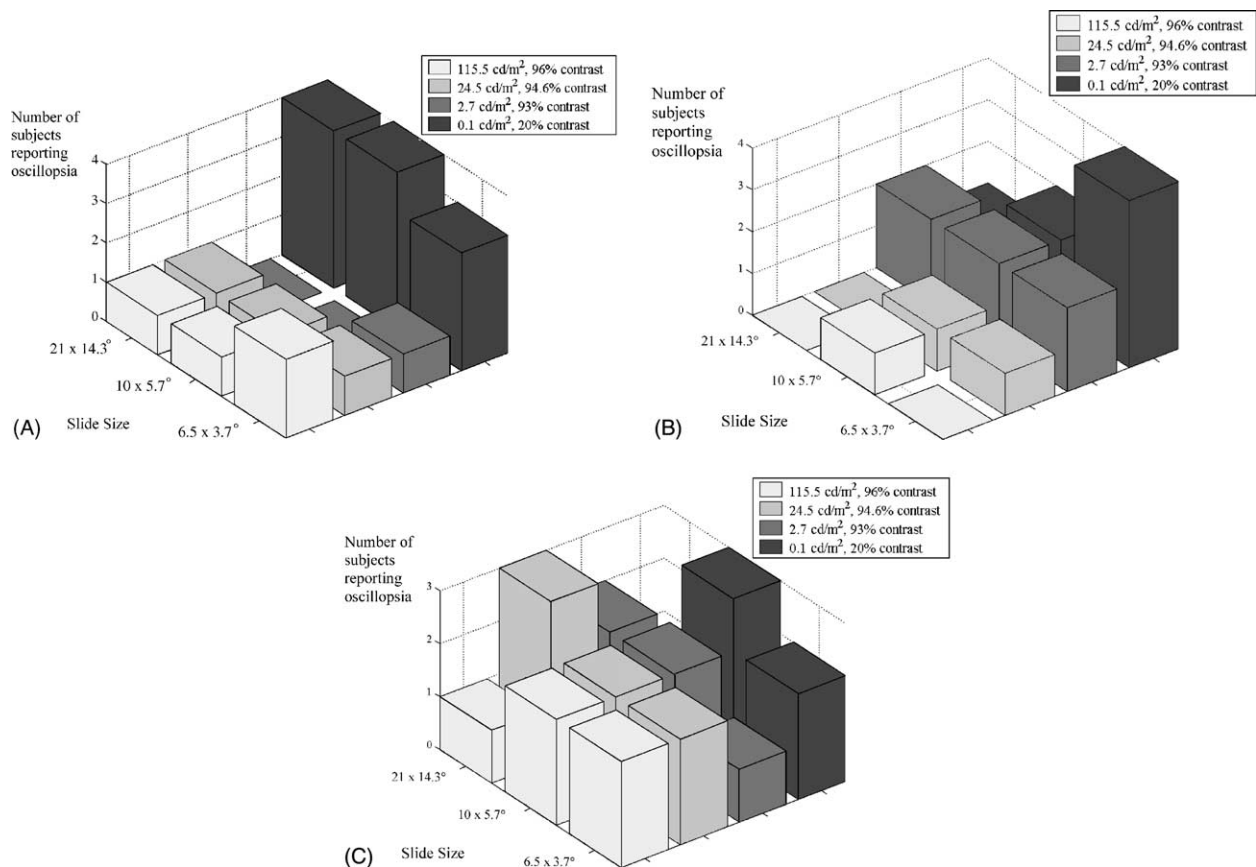


Fig. 2. The number of times that CN subjects perceived (A) the central LED as moving whilst the peripheral shapes remained stationary, (B) the peripheral shapes as moving whilst the central LED remained stationary and (C) both the peripheral shapes and the central LED as moving. For (A) and (B)—the two forms of spatially inhomogeneous oscillopsia—the proportion of occurrences of oscillopsia was greater for the darkest than the brightest background; this was not the case for (C), where the entire stimulus was seen to move. Since each subject saw each stimulus once, the maximum possible count for each bar is 16.

using χ^2 tests but results are not reported because the average expected values were less than five, undermining their validity. Because the strongest influence on oscillopsia appeared to be background luminance, we did, however, examine whether responses differed for the brightest and dimmest backgrounds—the contrast which appears to be the strongest in Fig. 2. For each category of oscillopsia (e.g., everything, LED only, background only, as well as the combination of them all) we examined whether the proportion of occurrences associated with the brightest and dimmest backgrounds differed. For all instances of oscillopsia combined (13 subjects at 3 background sizes for a total of 39 trials), it was reported in 23/39 trials for the dimmest and 11/39 trials for the brightest backgrounds. Comparing these proportions using the normal approximation to the binomial distribution, we found $z = 2.793$; thus the likelihood that these proportions differed was significant at $p < 0.05$. We then looked individually at reports of the LED, background or both moving. We found that for reports of the LED moving against a stationary background, (9 subjects at 3 background sizes, for a total of 27 trials), oscillopsia was reported in 11/27 trials for the

dimmest and 4/27 trials for the brightest backgrounds. This yielded $z = 1.92$; thus the likelihood that the proportions differed fell just short of significance at $p = 0.0548$. For the case where the background alone was seen to move (6 subjects times 3 background sizes, for a total of 18 trials), oscillopsia was reported 7/18 times for the dimmest and 1/18 times for the brightest backgrounds. This yielded $z = 2.06$; thus the likelihood that the proportions differed was significant at $p < 0.05$. Finally, for both the background and LED moving together (3 subjects at 3 background sizes for a total of 9 trials), oscillopsia was reported 5/9 times for both background luminances and clearly did not differ.

χ^2 tests were used to determine whether subject age was associated with oscillopsia. Subjects were divided into younger and older than 13 years and then further compared according to the times oscillopsia was experienced under any of the twelve viewing conditions. Analysis revealed that age was not a contributing factor to oscillopsia for any.

Trials with and without oscillopsia did not statistically differ across subjects in terms of the percentage of time for which foveation criteria were met, (independent

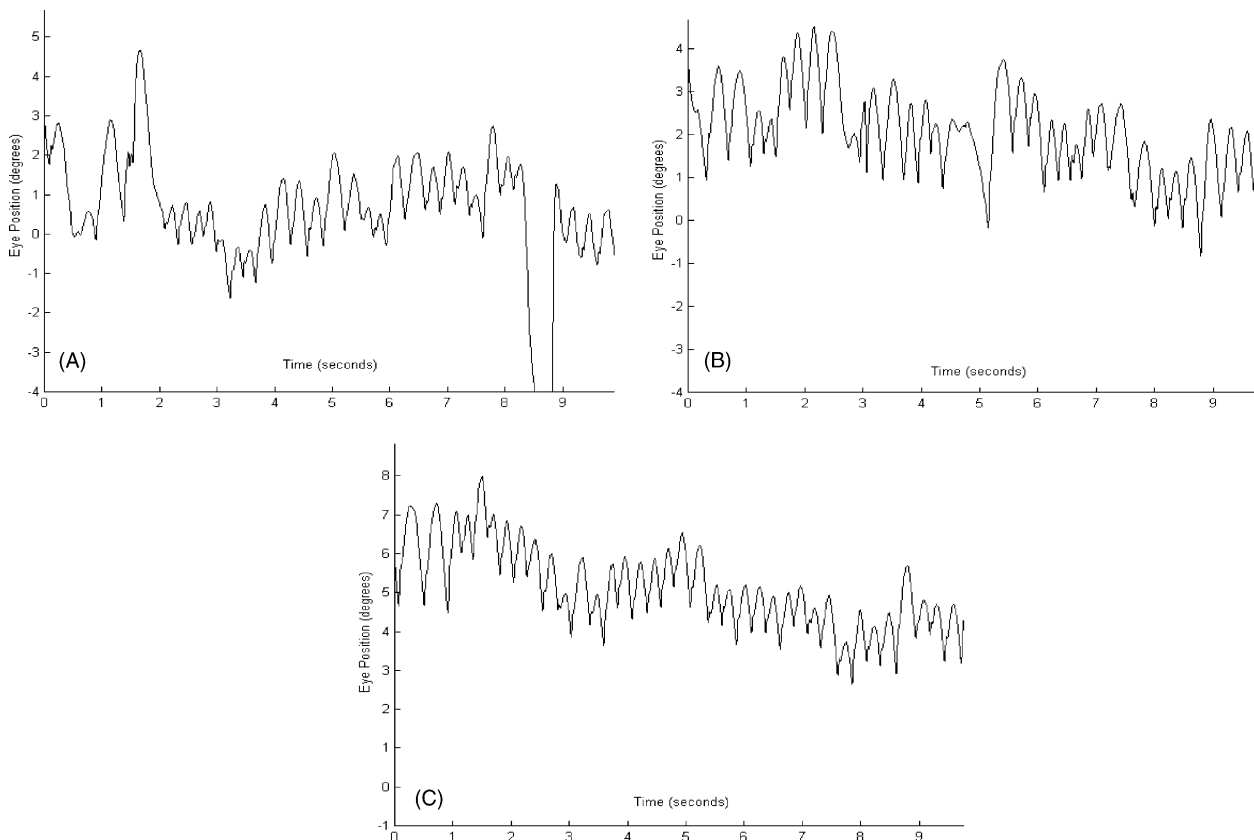


Fig. 3. Recordings of subject KS2, showing minimal changes in foveation time and waveform as perceptual stability varied. (A) Viewing the $10^\circ \times 5.7^\circ$ slide during the 24.5 cd/m^2 , 94.6% contrast setting. KS2 reported “nothing happened”. The % eye velocity $\leq 4^\circ/\text{s}$ and eye position at $\pm 2^\circ = 17.1\%$. (B) Viewing the $21^\circ \times 14.3^\circ$ slide at the 0.1 cd/m^2 , 20% contrast setting. He reported “the LED moved side-to-side”. The foveation criteria were met 18.4% of time. (C) Viewing the $21^\circ \times 14.3^\circ$ slide at the 2.7 cd/m^2 setting. He reported “the background shapes moved side-to-side”. The foveation criteria were met 20.5% of the time.

samples *t*-test, $p = 0.43$). Fig. 3 illustrates the CN waveforms of a subject whose perceptions varied with stimulus conditions but whose foveation time remained nearly constant. The questionnaire responses of this subject indicated that his experiences of oscillopsia occurred in dim light, in primary position but with no indication whether these percepts were spatially inhomogeneous. The percentage of time for which foveation criteria were satisfied did not differ between trials using a steadily illuminated LED in darkness and those using the largest, dimmest background (independent samples *t*-test, $p = 0.61$).

The means and standard errors were calculated for foveation time percentages for the four possible oscillopsia percepts. These were: LED only— $16.36 \pm 14.88\%$; background only— $28.23 \pm 25.78\%$; both together— $11.86 \pm 11.35\%$ and neither— $17.35 \pm 9.91\%$. A one-way analysis of variance was used to determine whether percentage foveation time was associated with the region of the stimulus perceived as oscillating. There was a significant main effect for the region that was perceived as oscillating ($F = 2.912$, $df = 3$, $p = 0.04$). Post hoc pair-wise comparisons using the Newman–Keuls multiple comparison test were only significant between ‘background’ and ‘both’ ($p < 0.05$).

4. Discussion

4.1. Questionnaire results

Responses to the oscillopsia questionnaire support recent reports (Abadi & Bjerre, 2002; Abel & Tkalecic, 2001) that oscillopsia is not as infrequent in CN as assumed (Bedell & Currie, 1993; Leigh et al., 1988). Twelve of the sixteen CN subjects (nine idiopathic and three albino) reported occasional real world oscillopsia under specific conditions. The direct questioning employed in this study is a probable explanation for the positive responses elicited about the presence of oscillopsia. Although only rarely associated explicitly with maximal nystagmus, its occurrence with primary position viewing (away from the null position), reading, fatigue or illness suggest that in the bulk of instances where oscillopsia is reported, it is associated with an exacerbation of the nystagmus waveform. That 12/16 CN subjects reported at least occasional real world environmental motion emphasises the need for more detailed studies so as to better determine in which CN patients oscillopsia occasionally occurs and under what conditions it does so.

Several authors have suggested that an abnormal neuro-developmental process induced by a visual defect at birth or in early infancy may influence the efficiency and sensitivity of perceptual stability in CN (Abadi & Pascal, 1991; Abadi et al., 1999). We found that CN

subjects with normal or near-normal visual acuity complained of oscillopsia as often as those subjects with an associated visuosensory defect. With age, CN waveforms and mechanisms responsible for suppressing oscillopsia improve (Abadi et al., 1999; Reinecke, Guo, & Goldstein, 1988). However, we found no influence of age on the presence of oscillopsia.

Although infrequent for most subjects, oscillopsia was at times problematic for some study participants. Although oscillopsia would be highly unlikely as a presenting complaint in congenital nystagmus, it may nonetheless be a somewhat uncommon but troublesome part of the condition for some individuals.

4.2. Perceptual results

This study is the first to have examined oscillopsia in typical CN patients whilst systematically varying the size, brightness and contrast of visual stimuli under normal, not retinally stabilised viewing conditions. Prior studies of oscillopsia in CN have stabilised all or part of the visual stimulus with afterimages (Dell’Osso et al., 1997; Kommerell, 1986; Leigh et al., 1988) or either mechanical or optical stabilisation methods (Abadi et al., 1999; Leigh et al., 1988). Irrespective of subject age, waveform type and visual acuity, it appeared that dim background stimuli appeared to be more frequently associated with oscillopsia. Subjects also perceived oscillopsia more frequently with a central LED and dim surround (11/16 trials) than with an LED viewed alone in a dark room (3/16 trials). As expected, no normal subject perceived environmental motion for any viewing condition. This appears to concur with Tusa et al. (1992) who found that their subjects’ perceptual stability broke down with a single 0.1° LED (1.5 mcd/m^2) in an otherwise dark room, when the slow-phase velocity of their nystagmus reached its maximum value. However, their subjects were atypical for CN, in that their nystagmus was suppressed under normal viewing conditions and only became manifest under reduced lighting. Whether they indeed had CN is difficult to determine. Participants in the present study had both clinical presentations and nystagmus waveforms consistent with CN. Stimulus size and brightness also varied over a wide range.

It should be highlighted that when viewing conditions induced oscillopsia, nystagmus characteristics did not vary concurrently (e.g., Fig. 3). Whilst most subjects who noted oscillopsia did so when viewing dim, smaller backgrounds, 3 of the 12 experienced it with the brightest background as well. This was consistent, however, with two of these subjects’ questionnaire responses. The absence of a relationship between perceptual stability and foveation is of interest, since oscillopsia has previously been described when congenital periodic nystagmus reaches a maximum (Abadi &

Bjerre, 2002; Abadi & Dickinson, 1986) or in a patient with unusually labile nystagmus (Abel et al., 1991). The patients reported by Tusa et al. (1992) would also fit this category, if they indeed had CN. These previous studies, however, noted uniform oscillopsia over the entire visual field; the observations in the present study that in most instances only part of the stimulus was moving distinguish it from its predecessors.

Some subjects commented that only the central LED oscillated whilst the peripheral background shapes remained motionless (Fig. 2A). Others reported the opposite (Fig. 2B). Yet other subjects experienced both the LED and peripheral shapes as moving simultaneously (Fig. 2C). In contrast, spatially inhomogeneous oscillopsia in Leigh et al. (1988) reflected their spatially inhomogeneous electronic or optical image stabilisation. Therefore, their subjects' ability to suppress the stabilised retinal image but experience oscillopsia of the non-stabilised portion of their visual field or vice versa is less surprising than our results. In addition, some subjects perceived different parts of the slide to oscillate as size and luminance/contrast changed. Although there was no change in the CN waveform seen to account for the perception of different parts of the slide oscillating during the various viewing conditions, it is possible that these subjects redirected their attention to different portions of the stimuli during each of the various viewing conditions. This may have evoked different perceptions of the same visual stimulus, as occurs with illusions which involve ambiguous perceptions of form (Tsal, 1994) such as the sketch which may be seen either as a duck or a rabbit. Similarly, the subject able to control which part of the incompletely stabilised visual field he saw as stationary (Leigh et al., 1988) may have done so via redirection of his attention without a concomitant refixation.

As noted previously, a number of previous reports have noted that nystagmus exacerbation may lead to oscillopsia (Abadi & Bjerre, 2002; Abadi & Dickinson, 1986; Abel et al., 1991; Leigh et al., 1988). The present study only recorded subjects in primary position and not either in nulls or in gaze positions where nystagmus was maximal. Those subjects who did not perceive oscillopsia under any of the 12 viewing conditions may have done so in gaze positions which exacerbated their nystagmus. Whether such incipient oscillopsia would be spatially homogeneous is unknown. The interplay of gaze position, distribution of visual attention, and stimulus characteristics in the triggering of oscillopsia all remain objects of future study, as does the trial-by-trial variability of the phenomena reported in this study. However, whether or not a change in attentional allocation alters the perception of a given stimulus, the perceptions reported herein still require explanation, even if their longer-term stability has not been examined.

The various mechanisms proposed to contribute to perceived stability of the visual world in CN do not explain why such stability should break down in a spatially inhomogeneous fashion. Although efference copy may suffice to suppress oscillopsia in most CN patients (Abadi et al., 1999; Bedell & Currie, 1993; Goldstein et al., 1992; Leigh et al., 1988), the findings of the present study suggest that its efficacy may break down under degraded viewing conditions. Furthermore, as foveation did not influence perception, waveform characteristics cannot account for the perceptual outcomes in this study. A possible explanation for oscillopsia affecting only certain parts of the visual field is that motion thresholds are spatially variable. Shallo-Hoffmann et al. (1998) has observed reductions in the motion aftereffect in individuals with CN and ascribed this to reduced motion sensitivity, but this provided no information as to whether such reductions apply equally across the visual field.

An alternative explanation involves the possibility that the higher luminance portions of the stimulus would arrive sooner in visual cortex because of intensity-dependent transmission time differences from retina to cortex (Allik & Kreegipuu, 1998). The flash-lag phenomenon, where synchronously presented flashed and moving stimuli are seen as if the flash lags behind the moving target, have been explained on the basis that moving stimuli have shorter latencies than static ones (Patel, Ogmen, Bedell, & Sampath, 2000; Whitney, Murakami, & Cavanagh, 2000). Other visual illusions related to the longer latencies of responses to dimmer stimuli are the monocular Hess and binocular Pulfrich effects (Williams & Lit, 1983). If fixation target and background luminance differences led to differing cortical arrival times of their respective representations, the perceived spatial relationships between them could be affected. There were considerable differences in luminance between the central LED and background, particularly for the dimmest trials. Consistent with this, observations of a stable fixation point and moving background were more frequent (7/16) for the lowest luminance slides (Fig. 2), where the luminance ratio between the fixation target and the surround was 4430:1. The luminance ratios for the other three backgrounds and the centre are 164:1, 48.1:1 and 3.8:1, respectively. Thus, when these CN subjects fixated the LED during any of the three sized slides at the dimmest luminance, the lower luminance portion of the uniformly moving stimulus (background) would always arrive later in visual cortex than the higher luminance portion (LED) because of the intensity-dependant visual latency differences in the retina (Allik & Kreegipuu, 1998). Uniform subtraction of efference copy across the visual field as the eyes oscillate would result in retinal image motion in different regions of the visual field being corrected with variable effectiveness, creating a phase lag between the perception of the bright

and dim portions of the visual stimulus. This explanation would also be plausible for the 164:1 and 18.1:1 luminance ratios, but less so for the 3.8:1 luminance ratio. Such an explanation would not a priori favour seeing either the LED or background as oscillating, but normal experience might predispose subjects to expect to see a small, attended object as moving against a stable background. This explanation would also be consistent with the infrequent observation of LED motion in an otherwise dark room, as with a dark background rather than a dim one there is no longer a delayed retinal slip signal from the periphery. Furthermore, since low contrast also increases latency (van der Tweel, Estevez, & Cavonius, 1979) and our lowest luminance background was also the lowest in contrast, only experiments where luminance and contrast are disambiguated will identify which parameter contributes most to perceptual stability.

In this first effort at evaluating how background size, brightness and luminance differences in visual stimuli could affect the perceptual stability of subjects with CN, we have found an apparent association between the presence of a bright central target against a dim background and the onset of spatially inhomogeneous oscillopsia. Much more remains to be determined regarding the stability of such perceptions, their susceptibility to changes in attention and their relationship with changes in nystagmus waveform. However, the fact that these phenomena occur at all imposes additional constraints on those mechanisms that maintain perceptual stability for most CN patients under most viewing conditions.

Acknowledgements

We thank Dr. James Elder who willingly searched through his private practice records to identify suitable participants for the study. Thanks to Dr. Louis Dell'Osso and Dr. Jonathan Jacobs for providing us with their modified Zoomtool and NFF foveation analysis software packages. This research was supported by the Jack Brockhoff Foundation and the Eye, Ear, Nose and Throat Research Institute.

Appendix A. Responses to the oscillopsia questionnaire

- (1) Have you ever experienced the world/objects to move?
Yes—75%
No—25%
- (2) If yes, can you describe this/these experience(s) by the use of an example(s):
Digital clock radio display—17%
Words on computer screen—8%

Words whilst reading—17%
Toys in bright lights—8%
People in bright lights—8%
Book shelves—8%
Entire room—17%
The thing being watched—25%
Things on TV that are not supposed to be moving—8%
Oncoming traffic—8%

- (3) When was your first experience? Days/months/years ago?
Years ago—100%
- (4) Do you experience it rarely/sometimes/frequently?
Rarely—8%
Sometimes—42%
Frequently—50%
- (5) Does the oscillopsia last long? Seconds/minutes/hours?
Seconds—17%
Minutes—25%
“How ever long looking at particular object”—58%
- (6) Can you voluntarily stop it? If yes, how?
Yes—42%
“Turn head”—40%
“Close both eyes for few seconds”—20%
“Close one eye”—20%
“Look at something different”—20%
No—58%
- (7) Does it only occur in a certain gaze position? E.g. when you look in the position where your nystagmus is maximal. Or does it occur whilst you look straight ahead?
Straight ahead—67%
Where nystagmus is maximal “when turning head laterally”—17%
Lying down—8%
During times of “visual effort”—8%
- (8) Does it occur in very well illuminated conditions or in dim lighting?
Well illuminated—25%
Dim lighting—42%
Any lighting condition—33%
- (9) Is it possible that only a certain portion of your visual field moves, e.g. the central portion moves whilst the peripheral surround remains still? Or vice versa
Entire portion—83%
Central portion moves—17%
- (10) What is the speed of the oscillopsia? Slow/moderate/fast?
Slow—8%
Moderate—58%
Fast—33%
- (11) Is it horizontal or vertical oscillopsia? Or both?
Horizontal—83%

Vertical—0%

“Can be either” horizontal or vertical—17%

- (12) Does anyone in your family have CN/LN/MLN?
Do you know if they complain of oscillopsia?

Yes—8% (x 1 older brother with oculocutaneous albinism—complains of oscillopsia)

No—92%

- (13) Have you been recommended some form of treatment to decrease the oscillopsia? E.g. baclofen

No—100%

- (14) Is there anything that triggers your oscillopsia?
E.g., fatigue/headaches/smoking/alcohol/sports?

No—42%

Fatigue—50%

Illness (cold/flu)—8%

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