Vestibular perception of angular velocity in normal subjects and in patients with congenital nystagmus

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Abstract
A technique is described for the assessment of vestibular sensation. The two main goals of the study were (i) to compare the perception of angular velocity with the eye velocity output of the vestibulo-ocular reflex and (ii) to study vestibular function in patients with congenital nystagmus; this was needed since most previous studies, based on eye movement recordings, have been inconclusive. Subjects indicated their perceived angular velocity by turning by hand a wheel connected to a tachometer. The vestibular stimuli used consisted of sudden deceleration from rotation at a constant horizontal velocity of 90°/s (‘stopping’ responses). Eye movements were recorded simultaneously with electro-oculography. In normal subjects the perceived angular velocity decayed from the moment of deceleration in an exponential fashion. The mean time constant of sensation decay was ~16 s. Eye movement velocity decayed with a similar exponential trajectory (time constant 16 s). Congenital nystagmus patients showed markedly shortened vestibular sensation (mean time constant 7 s). The following conclusions can be drawn: (i) the similarity of the eye velocity and perceptual responses suggests that these two systems receive a vestibular signal which has been similarly processed; (ii) the time constant of the responses indicates that this vestibular signal probably originates in the same brainstem ‘velocity storage’ integrator; (iii) the technique described is useful for clinical assessment of vestibular function, particularly in patients with ocular motility disorders; (iv) patients with congenital nystagmus have short vestibular time constants, which is probably due to changes induced in velocity storage processing by the persistent retinal image motion present in these patients.

Keywords: velocity storage; vestibular perception; congenital nystagmus; nystagmus; motion perception

Abbreviation: EOG = electro-oculography

Introduction
There has been a recent surge of interest in the subject of vestibular perception (Dai et al., 1989; Mergner et al., 1991; Brandt et al., 1994; Bohmer and Rickenmann, 1995; Kanayama et al., 1995; Nakamura and Bronstein, 1995; Bisdorff et al., 1996). This interest has been prompted partly by the poor correlation found between vestibulo-ocular test results and symptoms reported by vestibular patients (Kanayama et al., 1995) and partly by recent work which has identified cortical areas presumably mediating conscious vestibular sensations (Mergner et al., 1985; Grusser et al., 1990; Bottini et al., 1994; Brandt et al., 1994; Vitte et al., 1996).

The perception of tilt has been investigated extensively both in normal subjects and in patients with CNS and labyrinthine disorders (Freidmann, 1971; Dai et al., 1989; Dieterich and Brandt, 1992, 1993; Brandt et al., 1994; Bohmer and Rickenmann, 1995; Bisdorff et al., 1996; Bronstein et al., 1996; Anastasopoulos et al., 1997a, b). This percept, however, depends not only on vestibular otolith input but also on somatosensory cues (Mittelstaedt, 1992; Bisdorff et al., 1996) and, in the case of the perception of visual verticality, on ocular torsional position (Curthoys et al., 1991). The perception of rotation, as opposed to static tilt, has also been investigated, but most studies have been based on the ability of subjects to estimate angular displacements (Groen and Jongkees, 1948; Benson, 1968; Bloomberg et al., 1988, 1991; Mergner et al., 1991; Metcalfe and Gresty, 1992; Nakamura and Bronstein, 1995). Since the vestibular nerve codes not head displacements but head velocity (Goldberg and Fernandez, 1971a, b; Buttner and Waespe, 1981), estimates of angular displacements require further CNS processing, namely integration [mathematically, the process of transforming a velocity signal into a displacement (or position) signal is called integration]. In contrast, most
Oculographic studies of vestibular function are based on measurements of the slow-phase velocity of the eye (e.g. see reviews by Precht, 1979; Barnes, 1980). Therefore, there is some inconsistency between velocity-based ocular studies and displacement-based perceptual studies, and we felt that this inconsistency had to be tackled directly by simultaneously measuring ocular and perceptual (subjective) velocity. A specific question was whether the ‘velocity storage’ mechanism of the vestibulo-ocular reflex (Raphan et al., 1979) is present at a perceptual, presumably cortical, level.

The concept of velocity storage, pivotal to the understanding of central vestibular processing, emerged from observations made during rotations at constant angular velocity. These rotational stimuli, often called velocity steps, are the most frequently used for clinical assessment of the vestibular system. If a monkey or man is suddenly rotated at constant velocity in the dark, the velocity of the slow phase of the nystagmus decays exponentially with a time constant of ~15–20 s (Benson, 1968; Buttner and Waespe, 1981; Cohen et al., 1981). Direct recordings of the vestibular nerve in monkeys have shown, however, that the head velocity signal transmitted by the vestibular nerve has a time constant of decay of only 7–10 s (Buttner and Waespe, 1981). Thus, the head velocity signal appears to be stored in the brain and then released onto ocular motor neurons for the generation of nystagmus. This velocity storage process effectively carries out a partial mathematical integration of the head velocity signal travelling in the vestibular nerve so that the low-frequency content of the afferent signal is partly recovered. Brainstem circuits in the vicinity of the vestibular nuclei, behaving as mathematical integrators, are thought to mediate this storage process (Buettner et al., 1978; Blair and Gavin, 1981; Galiana and Outerbridge, 1984; Cannon and Robinson, 1985). To our knowledge, questions as to whether this velocity storage process is also present in cortical areas and whether it is involved in the perception of angular velocity have not been directly investigated. More specifically, the question of whether the dynamic characteristics (time constants) of vestibulo-ocular and vestibuloperceptual processes are essentially similar, as found by some authors (Cohen et al., 1981; Mergner et al., 1983, 1991) but not others (Benson, 1968; Jongkees, 1974; Hulk and Jongkees, 1948; Melvill Jones et al., 1964), was re-examined.

In addition, vestibular sensation was assessed in patients with congenital nystagmus. Congenital nystagmus is a common disorder but its precise incidence has not been fully established (Casteels et al., 1992). Even though patients with congenital nystagmus are usually asymptomatic, there are several reasons why they come to be seen by neurologists and why neurologists are interested in congenital nystagmus. When a patient is clinically examined for a neurological symptom, the finding of an unexpected nystagmus is usually baffling. If the long-standing presence of the congenital nystagmus is not known to the patient or if the nystagmus emerges late in life (Gresty et al., 1991), eye movement recordings and imaging procedures are warranted in order to identify pathognomonic nystagmus waveforms (Dell’Osso and Daroff, 1975; Yee et al., 1976) and to rule out structural CNS disease (Barton and Sharpe, 1993).

The presence of nystagmus in congenital nystagmus patients also makes the assessment of vestibular function difficult. In clinical practice, examination of vestibular function in a patient with balance problems or vertigo and congenital nystagmus is often restricted to the subjective assessment of the vertigo provoked by caloric irrigation of the external auditory meati. Modulation of the spontaneous nystagmus by rotational (Gresty et al., 1985) and caloric (Forssman, 1964a) procedures can occur and, based on oculographic investigations, a number of abnormalities in vestibular function have been reported. The first significant study in a large sample of patients undergoing caloric and electro-oculographic evaluation was conducted by Forssman (Forssman, 1964a), who concluded that ~50% of patients had no vestibulo-ocular responses. A second study by Forssman (Forssman, 1964b) concluded that caloric-spinal responses in those congenital nystagmus patients with absent caloric-ocular responses were normal but that the vertigo induced by the caloric stimulus was significantly less than that present in normal subjects. However, the assessment of the induced vertigo was based only on a subjective scaling of the symptom. Further caloric abnormalities in the form of canal pareses have also been reported (Yamazaki, 1979). Using rotational stimuli, Yee and colleagues (Yee et al., 1981) reported abnormal patterns of vestibulo-ocular response in 45% of congenital nystagmus patients, but these abnormalities were thought to depend mainly on the superimposed nystagmus and on deficient fast-phase generation. These authors therefore concluded that ‘the vestibulo-ocular response was basically normal in patients with CN [congenital nystagmus]’. Demer and Zee (Demer and Zee, 1984) also studied three albino–congenital nystagmus patients who had no ice-water caloric response. On rotational tests these patients had time constants of vestibulo-ocular responses of only 1–2 s, which is many times shorter than that of normal vestibular nystagmus and vestibular nerve activity. Thus, these authors suggested the possibility that the velocity storage process in congenital nystagmus patients could be inverted (i.e. that the time constant of the incoming vestibular nerve activity was shortened instead of lengthened), perhaps due to abnormal vestibular decussation, as occurs for the visual projections in albinos (Demer and Zee, 1984). Alternatively, the deficit may lie elsewhere, either in the peripheral vestibular system (a possibility rejected by all authors) or in the final velocity-to-position ocular integrator responsible for gaze-holding (Demer and Zee, 1984).

In order to overcome the limitations imposed by the strong spontaneous nystagmus upon quantitative vestibular assessment, Faldon and colleagues (Faldon et al., 1997) recently conducted a study in congenital nystagmus patients using a vestibular perceptual test. In this study, subjects seated on a rotating chair were exposed to discrete angular displacements in the dark. Their task was to drive themselves,
on the rotating chair, back to the starting position in the dark. It was found that patients with congenital nystagmus without a history of vestibular disease were normal or mildly abnormal. These results were thus not compatible with the earlier reports of either absent or extremely short vestibulo-ocular responses (Forssman, 1964a; Yamazaki, 1979; Demer and Zee, 1984). The relative normality in the perceptual test may have been due either to the task chosen, arguably a position-driven ‘navigational’ task rather than an instant, on-line assessment of their perception of angular velocity (Israel et al., 1996), or to the frequency content of the stimulus (0.17–0.5 Hz), which was not low enough to detect dysfunction in the velocity storage mechanism. In view of the discrepancies surrounding vestibular function in congenital nystagmus and the impossibility of settling the issue with eye movement recordings, we investigated a group of congenital nystagmus patients without labyrinthine symptoms using the vestibular perceptual test described below.

Material and methods

Experimental apparatus

Subjects were seated on a friction wheel-driven motorized rotating chair fitted with foot, arm and head rests. The latter was a padded, rigid U-shaped occipitoparietal support; the absence of head movements during the experiments was monitored by experienced observers with a chair-mounted infra-red camera. Attached to the chair in front of the subject was a 16-cm diameter low-friction wheel connected to a tachogenerator (Fig. 1A). The tachogenerator was made up of an optical encoder (British Encoder Products, Wrexham, UK) and a frequency-to-voltage converter with a bipolar option; the output signal was resistor–capacitor-filtered (time constant 0.5 s) to smooth the jerkiness of the hand movement. The subject’s task was to turn the wheel with a 7-cm high handle to indicate their own perceived turning speed in total darkness. Eye movements were simultaneously recorded with DC electro-oculography (EOG, bandwidth 0–70 Hz). Bitemporal electrodes were used since all subjects tested had either conjugate eye movements or slight comitant squints.

Experimental protocol

The chair was rapidly accelerated (~0.5 s) to a constant velocity of 90°/s. This velocity was maintained until the nystagmus (normal subjects) and turning sensation (normal and congenital nystagmus subjects) had stopped. The absence of the perception of turning was indicated verbally and by the absence of tachometer signal; the rotational stimulus was always maintained for no less than 60 s. A total of four rotations, two in each direction, and four stopping responses (deceleration ~0.5 s) were delivered.

Instructions

The subjects were instructed to turn the tachometer wheel with their preferred hand (all were right-handed), according to the sensation of their own velocity of rotation. They were informed that under this test condition the maximum speed was usually felt immediately after the chair started or stopped. They were asked to turn the wheel quickly, ensuring that the movement was smooth and comfortable, and then told that this was how they should indicate their own perceived speed when the chair suddenly started or stopped. They were then instructed that if the sense of rotation diminished they should indicate this by turning the wheel more slowly. If they no longer felt any rotation they should stop turning the wheel and shout ‘stopped’; this verbal report was timed separately by the experimenter.
Table 1 Clinical status—congenital nystagmus patients

<table>
<thead>
<tr>
<th>Subjects case no</th>
<th>Age (years)</th>
<th>Sex</th>
<th>Snellen acuity corrected</th>
<th>Ocular alignment</th>
<th>Clinical diagnosis/ wave form</th>
<th>Stereopsis (Titmus)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>28</td>
<td>M</td>
<td>0.3; 0.3</td>
<td>Esotropia</td>
<td>Idiopathic/PAN</td>
<td>Nil</td>
</tr>
<tr>
<td>2</td>
<td>26</td>
<td>M</td>
<td>1.2; 1.0</td>
<td>Orthotropia</td>
<td>Idiopathic/PAN</td>
<td>40°</td>
</tr>
<tr>
<td>3</td>
<td>27</td>
<td>M</td>
<td>0.5; 0.5</td>
<td>Orthotropia</td>
<td>Idiopathic/ bidirectional jerk</td>
<td>50°</td>
</tr>
<tr>
<td>4</td>
<td>40</td>
<td>M</td>
<td>0.17; 0.17</td>
<td>Esotropia</td>
<td>Inherited optic atrophy/bidirectional jerk</td>
<td>Nil</td>
</tr>
<tr>
<td>5</td>
<td>45</td>
<td>M</td>
<td>0.3; 0.3</td>
<td>Orthotropia</td>
<td>OA/X chrom/PAN</td>
<td>800°</td>
</tr>
<tr>
<td>6</td>
<td>40</td>
<td>M</td>
<td>0.67; 0.67</td>
<td>Orthotropia</td>
<td>Idiopathic/ left jerk</td>
<td>140°</td>
</tr>
<tr>
<td>7</td>
<td>47</td>
<td>M</td>
<td>0.33; 0.33</td>
<td>Orthotropia</td>
<td>Idiopathic/PAN</td>
<td>120°</td>
</tr>
<tr>
<td>8</td>
<td>29</td>
<td>M</td>
<td>0.7; 0.7</td>
<td>Orthotropia</td>
<td>Idiopathic/ bidirectional jerk</td>
<td>140°</td>
</tr>
<tr>
<td>9</td>
<td>33</td>
<td>M</td>
<td>0.7; 0.7</td>
<td>Orthotropia</td>
<td>Idiopathic/ bidirectional jerk</td>
<td>480°</td>
</tr>
<tr>
<td>10</td>
<td>34</td>
<td>F</td>
<td>0.1; 0.1</td>
<td>Orthotropia</td>
<td>occ albimism/ bidirectional + left jerk</td>
<td>800°</td>
</tr>
<tr>
<td>11</td>
<td>34</td>
<td>M</td>
<td>0.16; 0.16</td>
<td>Exotropia</td>
<td>occ albimism/ pendular + torsion</td>
<td>Nil</td>
</tr>
<tr>
<td>12</td>
<td>55</td>
<td>M</td>
<td>0.67; 0.5</td>
<td>Orthotropia</td>
<td>X-chromosomal recessive</td>
<td>100°</td>
</tr>
<tr>
<td>Controls</td>
<td>24–49</td>
<td>8 M, 6 F</td>
<td>1.0; 1.0</td>
<td>Orthotropia</td>
<td>NA</td>
<td>40–50°</td>
</tr>
</tbody>
</table>

PAN = periodic alternating nystagmus; OA/X chrom = ocular albinism, X chromosomal; occ = occutaneous; NA = not applicable.

In pilot studies, subjects often reported perceiving slight rotation for unusually prolonged periods (>2 min) during the actual rotation. This may have been due to the following factors, alone or in combination: cutaneous (‘wind in the face’), acoustic or proprioceptive (vibration of the turntable) cues. For this reason only the data collected during the stopping responses, when none of the above artefacts was present, were used in this study.

Data analysis
EOG, chair motion and tachometer wheel signals were recorded on a chart recorder and stored on a computer at a sampling rate of 250 Hz. The vestibular sensation data from the tachometer from the four stopping responses were normalized for direction (since no clear asymmetries were present on visual inspection) and averaged on the computer from the offset of chair velocity. As the resulting curve followed an approximately exponentially decaying trajectory, the data were fitted with an exponential. This was done by log-converting the data and following a linear fit with a least squares procedure against time. From the fitted curve the time constant of decay was determined; this represents the time taken for the signal to decay to 36.8% of its initial value (Fig. 1B). The total duration of the sensation was measured independently of the verbal report and the cessation of wheel output. EOG signals were filtered electronically (70 Hz), acquired on a computer and smoothed with a five-point moving average. On digitally differentiated data, saccades were automatically identified using an acceleration criterion and removed; the resulting data gaps were interpolated with mean pre-/post-gap values obtained with a 50-sample moving average (in-house software by Mr D. Buckwell). In patients with congenital nystagmus this technique could occasionally remove some accelerating slow phases of nystagmus, but it would always remove unwanted quick phases of nystagmus. Right and left responses were combined since no clear asymmetries were found on visual inspection. The four responses for each subject were then averaged from the offset of the chair velocity signal and fitted with an exponential curve. Raw eye recordings were visually inspected for classification of the nystagmus waveform and beating direction in the congenital nystagmus subjects. Additional grand averages (across normal and congenital nystagmus subjects) for eye velocity and sensation data were also obtained. Eye velocity was expressed in actual angular velocity units (°/s) but perceptual responses were expressed in arbitrary units and were normalized by their maximal amplitude. In order to quantify the overall envelope of activity in the perceptual and eye velocity responses, the area under the curve (velocity against time) was also computed. For sensation responses the amplitude of the curves was normalized, and the units of area measured were therefore also arbitrary; for eye velocity responses the units obtained in area measurements were degrees. In summary, the measurements taken were as follows: duration, time constant of decay and area under the curve for both eye and perceptual velocity response. All three measurements do not actually assess different aspects of the vestibular function, but essentially reflect the high-pass characteristic of vestibular
responses. However, duration and area make no assumptions about the rate of decay (exponential or not) of the response.

**Experimental subjects**

Twelve congenital nystagmus patients were tested, 11 males and one female (mean age 36 years, range 26–55 years). The diagnosis was established by a history of nystagmus since childhood, the absence of labyrinthine/neurological disease and eye movement recordings with typical waveforms. These recordings were obtained with DC EOG or infra-red reflection oculography (IRIS, Skalar, Delft, Netherlands), most patients having both recordings taken. A clinical summary of the patients can be found in Table 1; three patients had ocular albinism. No patient suffered from head-nodding; this could have influenced the outcome of the vestibular responses.

Fourteen normal subjects, age-matched to the congenital nystagmus patients and with no history of labyrinthine, neurological or visual medical history (except refraction defects), were also tested (eight males and six females, mean age 34 years, range 24–49 years). The latter constituted the normal control group for the congenital nystagmus patients, but the results for a larger group of normal subjects (n = 31), collected as normal control data for future clinical studies, will also be presented here (19 males, 12 females, mean age 31 years, range 21–62 years). To validate further the vestibular specificity of the test, four male patients with severe reduction of vestibular function on conventional caloric and rotational tests were investigated. Mean age was 58 years (range 41–70 years). The aetiology of the vestibular loss was idiopathic in three patients (it was suspected to be viral in one of these) and antibiotic ototoxicity in the fourth. Statistical comparisons between normal and congenital nystagmus subject groups were carried out with the Mann–Whitney U test. Consent was obtained from all subjects and patients, and the study was approved by the National Hospital for Neurology and Neurosurgery and the Institute of Neurology Joint Medical Ethics Committee.

**Results**

**General characteristics of the normal sensation and ocular response**

As the chair stopped, all normal subjects indicated that they felt rotation and turned the wheel accordingly. The maximal output from the tachometer was usually recorded within 2–3 s (range 0.48–3.8 s) of the chair stopping. Although subjects were given the option to turn the wheel in either direction (in order to avoid possible delays or uncertainty), all normal subjects turned the wheel in the direction of their perceived direction of rotation, i.e. clockwise on stopping from a leftward rotation and vice versa. The four stopping responses were averaged and normalized as if they were from a rightward turning sensation, i.e. upwards deflection of the trace. The envelope of both the tachogenerator (sensation) and the eye velocity signals approximated an exponential waveform (Fig. 2).

The data for individual subjects (sensation and eye velocity) were in most cases well fitted by the exponential curve, with a correlation coefficient (r) having a mean of 0.92 and ranging from 0.55 to 0.99. Only three subjects out of 31 showed poorly fitting curves, with a value of <0.80. There was considerable scatter in the time-constant perceptual data (mean 15.78 s, SD 7.73). The mean time constant of the ocular response was 15.74 s (SD 6.83). The duration of the sensation response from the offset of the chair velocity signal was 29.0 s (SD 8.2) from the verbal report and 32.6 s (SD 9.0) from the complete cessation of tachowheel output. This small discrepancy arose because the subjects said ‘stopped’ as they completed their last turn on the tachogenerator wheel and the tachowheel had a time delay of 1.5 s. Thus, the shape and time constant of the ocular and sensation responses are similar, although the total duration of the former is some 5 s longer than that of the latter. At the time when the subjects reported the end of the turning sensation, mean eye velocity was ~4°/s. The values of perceptual and eye velocity were positively but weakly correlated within subjects (linear regression; one-tailed significance values; n = 31); for time constant data r = 0.306, P = 0.047; for response duration r = 0.620, P < 0.005; for area under the curve data, r = 0.337, P = 0.036.

Four labyrinthine-defective patients were tested in order to confirm that the sensation transduced by the tachowheel was primarily mediated by the vestibular system. The responses of one such patient are presented in Fig. 2; the tachowheel output indicating turning sensation lasted ~3 s (at the same time the patient had two beats of nystagmus, lasting 2–3 s; not shown). The mean duration of sensation for the labyrinthine-defective patients was 3.5 s (range 1–8 s); eye movement activity lasted 5 s (range 0–8 s).
caused by the spontaneous (congenital nystagmus) nystagmus, normalization and averaging processes reduced the noise in congenital nystagmus patients was unexpected. It is likely that the approximately exponential eye velocity curve in the congenital nystagmus patients was not associated with the slight asymmetries noted in the vestibular sensation task. He was not able to tolerate the procedure and was dismissed from the study. The data presented here are therefore from 11 congenital nystagmus patients. All patients felt turning when the rotation started and stopped. One of the patients always turned the wheel in the clockwise direction but he later reported correctly his perceived directions of turning. The main result was a shortening of vestibular sensation in congenital nystagmus patients compared with normal controls (data for individual subjects are in Fig. 2; grand average data are in Fig. 3). The sensation curve had an approximately exponentially decaying profile (mean $r = 0.93$, range 0.89–0.99) but the time constant was significantly shorter than in normal controls (14 age-matched normal subjects, 18.56 s, SD 8.89; congenital nystagmus patients, 7.18 s, SD 3.81; $P < 0.01$). Nine out of 11 congenital nystagmus subjects had a time constant of <10 s. Figure 4 shows that the mean duration and area under the curve of the turning sensation were also significantly smaller in congenital nystagmus patients than in age-matched normal controls (both $P < 0.01$). These findings were independent of the nystagmus waveform. The beat direction of the nystagmus in the congenital nystagmus patients was not associated with the slight asymmetries noted in the vestibular sensation task.

Figure 3 also shows grand averages of slow-phase eye velocity during the stopping responses for both groups. It can be seen that the eye velocity and sensation profiles look similar in both the normal group and the congenital nystagmus group. The time constant of the grand average eye velocity curve was 16.04 s ($r = 0.99$) in the normal subjects and 6.84 s ($r = 0.95$) in the congenital nystagmus patients. The well-shaped, approximately exponential eye velocity curve in the congenital nystagmus patients was unexpected. It is likely that the normalization and averaging processes reduced the noise caused by the spontaneous (congenital nystagmus) nystagmus, thereby allowing the vestibulo-ocular response to come through. Individual eye-velocity recordings in congenital nystagmus patients were confounded by the spontaneous congenital nystagmus, and for this reason grand average data were considered a more appropriate representation of the vestibulo-ocular reflex response. However, post hoc inspection of the individual averages showed that this velocity profile was apparent, to varying degrees, in eight patients. The grand average eye velocity curve of the congenital nystagmus patients (Fig. 3) had a secondary decay some 30 s after peak velocity. Inspection of the individual EOG recordings suggested that this was probably due to a non-specific increase in spontaneous nystagmus in some congenital nystagmus patients with left-beating jerk congenital nystagmus, outlasting the initial specific vestibular effect. Admittedly, vestibulo-ocular data in the congenital nystagmus patient group are problematic, but still supportive of the psychophysical findings.

**Vestibular sensation in congenital nystagmus patients**

Patient 12 (Table 1) underwent only one rotation in the sensation task. The data presented here are therefore from 11 congenital nystagmus patients. All patients felt turning when the rotation started and stopped. One of the patients always turned the wheel in the clockwise direction but he later reported correctly his perceived directions of turning. The main result was a shortening of vestibular sensation in congenital nystagmus patients compared with normal controls (data for individual subjects are in Fig. 2; grand average data are in Fig. 3). The sensation curve had an approximately exponentially decaying profile (mean $r = 0.93$, range 0.89–0.99) but the time constant was significantly shorter than in normal controls (14 age-matched normal subjects, 18.56 s, SD 8.89; congenital nystagmus patients, 7.18 s, SD 3.81; $P < 0.01$). Nine out of 11 congenital nystagmus subjects had a time constant of <10 s. Figure 4 shows that the mean duration and area under the curve of the turning sensation were also significantly smaller in congenital nystagmus patients than in age-matched normal controls (both $P < 0.01$). These findings were independent of the nystagmus waveform. The beat direction of the nystagmus in the congenital nystagmus patients was not associated with the slight asymmetries noted in the vestibular sensation task.

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**Discussion**

The aims of the study reported here were to (i) compare the vestibularly mediated perception of turning with vestibulo-ocular reflex velocity and (ii) assess vestibular function in patients with congenital nystagmus with a non-eye movement-based technique.

**Vestibularly mediated perception of angular velocity in man**

Our study showed that eye velocity and the perception of turning in response to an angular velocity step were remarkably similar. Both functions increased rapidly after the chair stopped and then decayed exponentially with similar time constants of ~16 s. This similarity indicates that the ‘velocity storage’ process is also present at a perceptual, presumably cortical level. The only difference is that the duration of the eye velocity curve (i.e. the nystagmic response) is slightly longer, outlasting the sensation curve by 4–5 s. This difference reflects the fact that perceptual thresholds are higher than nystagmic thresholds, particularly after velocity steps (Benson, 1968; Guedry, 1974; Hood, 1984). We ensured that the response obtained was based solely on vestibular input by assessing the turning sensation during the stopping response, when no acoustic or somatosensory stimuli could mediate the sensation and, further, by showing that the sensation was drastically shortened in patients with reduced vestibular function.

Although recent work has suggested agreement between body displacement perception and vestibulo-ocular reflex data (Mergner et al., 1983, 1991), the extent of such agreement between perceptual velocity and ocular-motor velocity during velocity steps could not be predicted. This is partly due to the fact that simultaneous measurements of eye and perceptual responses had been obtained only sporadically (Cohen et al., 1981; Baloh et al., 1982) and that the results indicated only small differences between the two (Cohen et al., 1981). Also,
Vestibular perception in congenital nystagmus

Fig. 4 Mean duration, time constant and area under the curve of turning sensation in congenital nystagmus patients (n = 14) and age-matched normal controls (n = 14).

cortical neurons in the cat (Mergner et al., 1981, 1985) and monkey (Grusser et al., 1990) show large phase variability and different discharge patterns that can be related to angular acceleration, velocity, displacement or intermediate functions. The existence of different types of vestibulocortical neurons can be expected since they presumably mediate cognitive or motor responses to different dynamic components of a vestibular stimulus. This diversity, however, was one of the reasons why the precise dynamics of vestibular sensation could not be predicted.

A number of earlier studies in the literature indicated lack of agreement between vestibular perceptual and ocular responses (Van Egmond et al., 1948; Melvill Jones et al., 1964; Benson, 1968; Jongkees, 1974). Most of these disagreements can be explained as technical and task-dependent factors. Before the advent of electro-oculography, it was accepted that the results of sensation and nystagmus ‘cupulograms’ were different (Jongkees, 1974). This technique (cupulometry) was based on measurements of the duration of either nystagmus or turning sensation in response to sudden deceleration from velocity steps of different sizes. With this technique, nystagmus thresholds were higher than sensation thresholds, which effectively resulted in nystagmus and sensation cupulograms of different slopes (i.e. different time constants). It is likely that poor detection of the nystagmus at low rotational velocities, by direct observation with Frenzel’s glasses, led to the unusual finding that nystagmic thresholds were higher than sensation thresholds. Even when EOG was used to measure the nystagmus time constant, as in the current study, the sensation responses were still measured by cupulometry (Melvill Jones et al., 1964). The result was an even larger discrepancy between the two measures, e.g. a time constant of 15 s for nystagmic responses to velocity steps measured with oculography and a sensation time constant of 10 s with cupulometry (Melvill Jones et al., 1964). It should be noted that time constants measured by sensation cupulograms always yielded short vestibular time constants [e.g. 8 s by Van Egmond and colleagues (Van Egmond et al., 1948)]. This suggests that the time constants determined by traditional cupulometry might reflect true cupular deflection, without prolongation by the velocity storage integrator. Thus, the employment of different experimental procedures and recording techniques was a likely cause of observed discrepancies and emphasized the need for studies to employ simultaneous ocular and sensation recordings.

Our results, based on simultaneous recording of subjective (perceptual) and objective (ocular) velocity, provide evidence that the perception of angular velocity is based on signals subserved by the brainstem velocity storage system. The agreement between our ocular-motor and perceptual responses was clear on direct inspection of grand average data and from the correlation of the two data sets across individual subjects. Although the latter showed a weak statistical correlation, it must be noted that the correlation between sensation and ocular data was better for raw measurements (duration and area under the curve) than for exponentially fitted data (time constants). Taking all this into consideration, it would seem that, in our experiments, part of the intra-individual disagreement between perceptual and ocular-motor data relates to limitations in the number of recordings available (averaging factor) and imperfect exponential fitting of the responses. However, some degree of genuine dissociation is likely since different pathways and processing must be required for gaze stability and motion perception functions.

For instance, the amount of voluntary control over ocular and perceptual responses is likely to differ. This would produce
additional dissociation, even during simultaneous ocular and perceptual measures. By way of example, vestibulo-ocular reflex responses can be drastically enhanced or suppressed according to mental task or attentional set (Barr et al., 1976).

Such fluctuations, which are common during rotational/caloric testing, could weaken the intra-individual correlation between perceptual and ocular velocity.

Differences in mental set or task may also influence vestibulo-perceptual tasks, although few studies have investigated this. In fact, most studies concerned with vestibular sensation have emphasized the use of external spatial cues in the task. This has taken the form of either pointing or returning to an imaginary/remembered part of the room (Bloomberg et al., 1988, 1991; Metcalfe and Gresty, 1992; Nakamura and Bronstein 1995; Israel et al., 1996) or estimating the angle travelled during rotation (Groen and Jongkees, 1948; Bekesy, 1955; Benson, 1968; Cohen et al., 1981; Baloh et al., 1982). The findings by Benson (Benson, 1968) strongly suggest that such cues may have impressing effects on vestibular perception. In this study (Benson, 1968), subjects rotating at constant speed indicated each time they felt they had travelled 90°. When subjective velocity was calculated from this subjective displacement estimate, it was found that subjects grossly overestimated their own angular velocity and that the time constant of decay of the vestibular sensation was of the order of 35–40 s. These perceptual time constants are twice as long as those we report here, despite the fact that the time constant of nystagmus was identical to our findings. Thus, it appears that velocity estimates based on tasks requiring a mental reconstruction of external space can give results different from velocity estimates that are based on direct feeling of the subjective speed. In the latter case, as in our task, subjective estimates seem to be provided directly by velocity signals carried by post-velocity-storage vestibular pathways. In the former case, when the task is referenced to external space, further processing of the vestibular velocity signals seems to take place. The overestimation of subjective velocity and the unusually long time constants reported in externally referenced tasks during velocity steps (e.g. Benson, 1968; for review, see Guedry, 1974) suggest that the additional spatial processing changes the dynamic characteristics of the perceptual vestibular signal. A recent study comparing slow-phase eye velocity with verbal reports of subjective velocity during velocity steps also showed slightly longer time constants for perception (24 s) than for eye velocity (15 s) (Howard et al., 1998). These findings could suggest that cognitive processes involved in velocity estimation or verbal communication of the estimate may also underlie the reported dissociation between ocular and subjective velocity. More natural, shorter-duration stimuli seem to be sensed correctly by normal subjects, both in terms of peak velocity and total displacement (Mergner et al., 1996).

In summary, our study shows good agreement between the grand average vestibular perceptual and ocular responses. This is to be expected since the common central processing of vestibular signals allows functional congruency between the various behavioural outputs, i.e. gaze stabilization by the vestibulo-ocular reflex and the perception of self-motion in the external world. The weak intra-individual correlation observed in the present study may relate to fluctuations in mental set as well as to technical limitations. On the other hand, some degree of genuine dissociation between the two is likely on the basis that different neural pathways must be involved for perceptual and gaze stability functions (Peterka and Benolken, 1992). A number of tests for the assessment of perceptual vestibular function have been described, many from the days before oculography became established (Groen, 1948; Hulk and Jongkees, 1948; Van Egmond et al., 1948; Bekesy, 1955; Guedry, 1974; Mergner et al., 1981; Metcalfe and Gresty, 1992; Nakamura and Bronstein, 1995). From a clinical perspective, advantages of the test reported here are that it directly assesses velocity perception (as opposed to displacement), it is easily understood by subjects, it does not significantly lengthen the examination and, perhaps more importantly, it can run simultaneously with conventional oculographic evaluation of vestibular function. The method also allows measurement of vestibular function in patients in whom the eye movement disorder is not relevant to a complaint of dizziness or vertigo (e.g. congenital nystagmus; see below). A disadvantage, shared with other techniques based on manual pointing or apparatus-driving (Mergner et al., 1981, 1985; Metcalfe and Gresty, 1992), is that it requires reasonably good hand control and may thus be unsuitable for some neurological patients.

**Vestibular function in congenital nystagmus**

The assessment of vestibular function by means of perceptual tests is particularly useful for patients with eye movement disorders. Faldon et al. (Faldon et al., 1997) recently examined vestibular perception in congenital nystagmus patients by instructing them to steer the rotating chair back to the starting position, a task predominantly based on a displacement estimate of the stimulus (Bloomberg et al., 1988, 1991; Israel et al., 1996). The study showed that congenital nystagmus patients without cochlear–vestibular symptoms produced either normal or mildly hypoactive responses but that those patients with additional suspected vestibular disease were significantly abnormal (>2 SD from normal controls). These results were most encouraging since it was the first reliable study using a non-eye movement-based, quantitative method of measuring vestibular function in congenital nystagmus patients.

On the basis of the findings in the congenital nystagmus subjects with no vestibular symptoms, a model of the vestibular system in congenital nystagmus was proposed which included a shortened time constant of the velocity storage system (Faldon et al., 1997). Our results, based on velocity perception estimates, now provide direct evidence of a significantly shortened vestibular time constant. The time constant of decay of vestibular sensation in congenital nystagmus patients (7–8 s) was half the duration of the time constant found in normal subjects. These values are very similar to the time constant of
the activity in the vestibular nerve (Buttner and Waespe, 1981), indicating that congenital nystagmus patients have a minimal or absent velocity storage mechanism. In the study by Faldon et al., the degree of dysfunction of the velocity storage mechanisms could not be measured since the frequencies tested (0.17–0.5 Hz) were not low enough to require significant intervention of the velocity storage integrator.

Neither the study of Faldon et al. (Faldon et al., 1997) nor the current study, however, lend support to the suggestion by Demer and Zee (Demer and Zee, 1984) that in congenital nystagmus there may be an ‘inverted storage’ mechanism which would explain the ultra-short time constant of vestibular nystagmus (~2 s) present in their patients. The findings of ultra-short vestibulo-ocular time constants may be due to added dysfunction in gaze-holding mechanisms (the velocity-to-position integrator), an alternative possibility suggested by those authors.

The mechanism of the reduction in vestibular time constant in congenital nystagmus patients is intriguing. We postulate that the shortening of the vestibular time constant is secondary to the continuous movement of retinal images (retinal slip) in these patients. Indeed, retinal image slippage can have profound effects on the velocity storage mechanism, e.g. the shortening of the time constant of the post-rotatory nystagmus by visual fixation, usually called visual ‘dumping’ of the velocity storage (Cohen et al., 1981; Waespe and Schwarz, 1986). This means, essentially, that vestibular responses in the dark can be shortened by as much as 50% by a period of fixation (i.e. retinal slippage) as brief as 2–3 s. Vestibulo-ocular (Young and Henn, 1974), spinal (Kato et al., 1977) and vestibuloperceptual responses (our unpublished observations) are also reduced in normal subjects by prolonged optokinetic stimulation. These findings support the view that prolonged visual motion stimulation can shorten the velocity storage time constant and may thus explain the findings in congenital nystagmus patients.

The functional consequences of the reduction of the time constant of vestibular sensation in congenital nystagmus patients are not clear. We may ask what potential disadvantages could the congenital nystagmus patients experience as a result of a shortening of vestibular time constants. In normal animals and in man, the partial integration of the vestibular afferent signal produced by the velocity storage mechanism helps to increase the low-frequency response (<0.1 Hz) of the vestibular system. The loss of this low-frequency component of the response would have very little effect in a normal subject, since gaze stability at such low frequencies of head motion can be achieved by optokinetic reflexes. Congenital nystagmus patients are continuously exposed to a large amount of retinal image motion. The small additional retinal slippage should have a negligible effect, particularly since congenital nystagmus patients have raised thresholds of visual motion detection (Dieterich and Brandt, 1987; Shallo-Hoffman et al., 1998).

The shortened time constant of vestibular perception, however, can account for the small reduction in ‘vestibular navigation’ capabilities observed in congenital nystagmus patients (Faldon et al., 1997). These observations, however, were made during purely vestibular guided re-orienting movements in the dark, without participation of the somatosensory or locomotor system (by remote control steering of the rotating chair). Such conditions are rarely encountered outside the experimental scenario and, in agreement, there are no reports of symptoms in congenital nystagmus patients which could be correlated with the small decrement in vestibular navigation capacity. Thus, handicap does not appear to arise, for congenital nystagmus patients, as the result of shortened vestibular time constants. The few situations in which short vestibular time constants are observed in normal subjects relate to highly skilled physical activities, for instance those performed by ballet dancers, figure skaters and aircraft pilots (Aschan, 1954; Fukada et al., 1967; Dix and Hood, 1969) or in experimental laboratory conditions with repetitive exposure to vestibular and/or optokinetic stimuli (Brand, 1968) and sensation (our unpublished observations). From these observations it would appear that, in certain circumstances, a shortening of the vestibular time constant may be desirable in order to suppress long-lasting unwanted vestibular nystagmus, disorienting turning sensations and motion sickness-like symptoms (Young and Henn, 1974). Against this background, the finding of reduced vestibular velocity storage in congenital nystagmus could be viewed as an adaptive phenomenon attempting to reduce visuovestibular disorientation in patients with excessive retinal instability. In support of this view are the findings of a recent study in congenital nystagmus patients by Eggert and colleagues (Eggert et al., 1997). These authors found that, whilst visual detection of object motion was impaired in congenital nystagmus, visual functions mediating self-motion perception were spared. They concluded that the sparing of visually mediated self-motion functions indicates that congenital nystagmus patients adapt to their nystagmus centrally, at the level of visuovestibular interactions. It is likely that such central adaptation can involve changes in vestibularly mediated self-motion perception, as we report here.

In summary, this study describes a strong similarity in the vestibularly mediated perception of body angular velocity and the slow-phase velocity nystagmic response. The brainstem velocity storage integrator seems to provide a similar input to the brainstem ocular-motor plant and to the cortical structures mediating the conscious perception of rotation. Congenital nystagmus patients exhibited a shortening of the vestibular sensation time constant, indicative of a severe reduction in velocity storage function. It is postulated that this reduction is secondary to the continuous retinal image motion induced by the nystagmus in these patients.

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