Anterior canal failure: ocular torsion without perceptual tilt due to preserved otolith function

M Strupp, S Glasauer, E Schneider, T Eggert, M Glaser, K Jahn, T Brandt

A patient with anterior semicircular canal dehiscence syndrome underwent surgical patching that caused an isolated dysfunction of the left anterior semicircular canal postoperatively. He exhibited significant ocular torsion toward the side of the affected labyrinth (17° excyclotropia of the ipsilateral eye), but no displacement of the subjective visual vertical. This dissociation suggests that an isolated ocular torsion may occur after an anterior semicircular canal lesion. A combined ocular torsion and subjective visual vertical tilt, which is usually seen with vestibular lesions, requires an associated otolith dysfunction.

Healthy subjects are able to adjust their subjective visual vertical (SVV) within a precision of ±2.5° when placed in front of an unstructured background providing no cues for visual orientation. This ability is attributed to the vestibular system (namely the otolith organs) and to some extent also to the somatosensory system. A direction specific tilt of the SVV is a typical and sensitive sign of peripheral and central vestibular disorders. Psychophysical studies of patients before and after unilateral vestibular neuronecrtomy provide support for this interpretation. A tilt of the SVV has, however, also been observed in non-vestibular disorders: third and fourth cranial nerve palsies, for instance, cause monocular SVV tilts, but they are only minor and unpredictable.

Vestibular disorders not only involve a tilt of SVV but also an associated ocular torsion (OT). The tilt of SVV increases with larger angles of OT, both in the same direction. This raises the following questions: (1) Are the tilts of SVV and OT two independent signs of a vestibular tone imbalance or is the tilt of the SVV the perceptual correlate of the OT? (2) What are the differential effects of isolated vertical SCC and otolith dysfunction on both signs? These questions cannot be addressed in animal experiments (determination of the SVV is not possible) or in subjects with lesions of the eighth nerve (which also to the somatosensory system). A direction specific tilt of the SVV is a typical and sensitive sign of peripheral and central vestibular disorders.

For measurement of the SVV, the patient remained sitting in an upright position and looked into a hemispheric dome 60 cm in diameter. The surface of the dome extended to the limits of the observer’s visual field and was covered with a random pattern of coloured dots; 30 cm in front of the observer was a circular target of 14° visual angle with a straight line through the centre. The patient had to adjust the central test edge to the vertical. SVV was determined by means of 10 adjustments of the target disk from a random offset position to the SV (normal range (2SD): ±2.5°).

Eye movement recordings

For 3D analysis of the eye movements and measurement of the gain of the vestibular ocular reflex (VOR), a soft annulus containing two fine coils (Scalar, Delft, Netherlands) was placed on the eye after topical anaesthesia and on the head. Eye position and head position were measured simultaneously by the voltage induced by three orthogonal magnetic fields (Remmel, Ashland, MA, USA) on the two coils. The signals were digitised with a 12 bit analogue-digital converter at a sampling rate of 1 kHz. The average 3D direction of the eye movement was evaluated by the first principal component of the instantaneous eye velocities. Average slow phase velocity was evaluated using the trimmed mean algorithm. This was recorded by a search-coil 3D analysis of the vector of the nystagmus and measurements of VOR gain by the head-impulse test in the planes of each SCC.

CASE REPORT

A 53 year old ophthalmologist had had recurrent attacks of vertigo and oscillopsia induced by coughing and Valsalva manoeuvres for many years. High resolution temporal bone computed tomography showed a defect of the bone overlying the left aSCC. Three dimensional eye movement recordings with the search-coil technique and subsequent vector analysis demonstrated that the eye movements were caused by excitation of the left aSCC. A superior (also called anterior) canal dehiscence syndrome was diagnosed, and the patient underwent surgical patching of the left aSCC (fig 1). A polydioxanone synthetic folio was placed together with fascia above the plugged canal to prevent transmission of changes in intracranial pressure. The intervention caused rotatory vertigo and spontaneous torsional-vertical nystagmus (last phase; from the viewpoint of the patient: torsional clockwise and upward)

Abbreviations: OT, ocular torsion; aSCC, anterior semicircular canal; SVV, subjective visual vertical; VOR, vestibular ocular reflex

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due to an isolated dysfunction of the left aSCC, leading to a vestibular tone imbalance between the left aSCC and the right posterior SCC. This was recorded by (a) a search-coil 3D analysis of the slow phase vector of the nystagmus, which pointed in the direction of the aSCC, and (b) plane specific measurements of VOR gain, which showed that the gain of the VOR in the direction of the left aSCC was 0.43, for all other SCC >0.75.

The patient had not had either an OT or a displacement of the SVV before surgery (table 1). Five days after surgery, when the spontaneous nystagmus had ceased, we found a significant OT toward the side of the affected labyrinth (17° to the left (excyclotropia—that is, torsion of the papilla-fovea line clockwise from the viewpoint of the observer toward the left) and the right eye, a 3° excyclotropia. (C) Schematic drawing of torsional eye positions (indicated by crosses within the eyes) before surgery and SVV (dashed line), as measured in the patient. (D) Data measured in the patient five days after surgery. Despite constant rotation of the retina the adjustment of the SVV was correct. This may be explained by the occurrence of central adaptation to the inappropriate OT by virtue of the visual input within the five postoperative days and/or by a space constancy mechanism used as an efference copy of the ocular motor signal, as suggested for compensation of OT during head-centric visual localisation. The lesion of the left aSCC is indicated by an X. (E) Expected SVV setting if the patient would have adjusted the line according to a subjective tilt corresponding to his ocular torsion. (F) Expected SVV if ocular torsion and subjective tilt are added (no compensation of ocular torsion).

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**Table 1** OT and displacement of the SVV two weeks before surgery and five days, 20 days, and six months afterwards

<table>
<thead>
<tr>
<th></th>
<th>Two weeks before surgery</th>
<th>Five days after surgery</th>
<th>Twenty days after surgery</th>
<th>Six months after surgery</th>
</tr>
</thead>
<tbody>
<tr>
<td>OT left eye (°)</td>
<td>4 Exc</td>
<td>17 Exc</td>
<td>6 Exc</td>
<td>5 Exc</td>
</tr>
<tr>
<td>OT right eye (°)</td>
<td>8 Exc</td>
<td>3 Exc</td>
<td>8 Exc</td>
<td>9 Exc</td>
</tr>
<tr>
<td>SVV binocular (°)</td>
<td>+0.8 (1.6)</td>
<td>-1.1 (1.6)</td>
<td>+0.4 (1.1)</td>
<td>+2.3 (0.9)</td>
</tr>
<tr>
<td>SVV left eye (°)</td>
<td>-1.8 (2.2)</td>
<td>-2.4 (1.3)</td>
<td>+0.9 (1.2)</td>
<td>+0.6 (1.4)</td>
</tr>
<tr>
<td>SVV right eye (°)</td>
<td>+0.1 (2.4)</td>
<td>+0.6 (2.2)</td>
<td>+2.8 (2.0)</td>
<td>+3.1 (2.6)</td>
</tr>
</tbody>
</table>

Exc, excyclotropia; +, displacement of the SVV to the right from the patient’s view; −, displacement of the SVV to the left; values are given as mean or as mean (SD).
function, which was tested by brief, high acceleration lateral translations (“heaves”), also appeared to be intact (a quantitative test of utricle function was not performed). Testing of the saccular function by vestibular evoked potentials showed that the threshold for clicks was reduced in the left ear (75 dB) before surgery but was normal after surgery (90 dB). Twenty days after surgery OT had disappeared, and the patient was symptom free except for a dynamic deficit of the left aSCC, which manifested as oscillopsia during rapid head movements in the plane and the on-direction of the left aSCC. No vertigo or oscillopsia was observed during coughing or Valsalva manoeuvres (follow up time six months). Audiological testing was also normal.

DISCUSSION
The patient posed an intriguing puzzle: the binocular torsion of the eyes was not associated with a tilt of the perceived vertical. This finding suggests that the ocular motor and perceptual systems in the roll plane may operate independently under exceptional conditions. The case is unique in that dysfunction of the aSCC was isolated, and the receptors of the labyrinth, which indicate the direction of gravity (the otolith organs), were not affected.

How can these signs and symptoms be explained? The patient underwent canal plugging, which caused damage to the neuroepithelium/vestibular hair cells of the aSCC. This led to a:

1. static deficit of the aSCC VOR, indicated by the torsion of the eyes and the transient torsional downbeat nystagmus. As canals do not respond to static positional changes, it has to be assumed that OT and nystagmus are caused by an isolated canal lesion. As recently shown, static OT can be elicited by SCC stimulation if resetting torsional quick phases are suppressed. Nystagmus suppression could be one of the first steps in the process of central vestibular compensation, which would lead to a static OT as demonstrated by our patient.

2. a dynamic deficit of the aSCC VOR, indicated by the decreased gain of the VOR that was measured by the search-coil technique in the plane of this canal; but

3. no dysfunction of the otolith organs, namely the utricle. The “head-heave test” (which has its limitations) was used to test the function of the utricle in our patient and vestibular evoked potentials, to test the saccular.

These findings suggest that an intrasensory vestibular mismatch occurred between the canal and otolith signals (SCC indicating self rotation in the plane of the affected canal compared with otolithic indicating true verticality). This mismatch was resolved differentially for ocular motor function and perception. There was also an intersensory mismatch between the visual and vestibular signals (rotation of the retina compared with true perceived verticality). The adjustment of the SVV was, nevertheless, correct despite constant rotation of the retina. At least two causative mechanisms are conceivable: (1) central adaptation to the inappropriate OT occurred by virtue of the visual input within the five postoperative days and/or (2) a space constancy mechanism used an efference copy of the ocular motor signal, as suggested for compensation of OT during head-centric visual localisation. It must also be assumed that the isolated canal dysfunction (otolith function was preserved) caused no perceptual tilt as illustrated in figure 1C–F. This case has clinical and experimental implications, particularly for the modelling of ocular motor and perceptual functions. Current mathematical models of otolith-canal and visual-vestibular interaction must also be revised, for they assume that an ocular motor response and a perceptual response share the same computational structures and, thus, are directly linked.

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REFERENCES