

## SHORT REPORT

## Persistence of perilymph fistula mechanism in a completely parietic posterior semicircular canal

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The three dimensional eye movements (search coil technique) of a patient with a completely parietic left posterior semicircular canal as a result of a perilymph fistula (PLF) were studied. The patient still exhibited pressure induced nystagmus that obeyed Ewald's first law. This finding cannot be explained by otolith stimulation, but might indicate that PLF mechanisms either persist in canal plugging or act on the ampulla by directly deflecting the cupula.

Perilymph fistulas (PLFs) are pathological connections between the perilymphatic space of the labyrinth and the middle ear (outer fistula) or the cerebrospinal fluid (inner fistula). These abnormal connections transmit pressure to excite the affected semicircular canal, which clinically leads to paroxysmal vertigo of short duration. PLF nystagmus enables the validity of Ewald's first law to be investigated,<sup>1,2</sup> which helps the clinician to differentiate between a peripheral and a central vestibular lesion. This law states that the axis of the elicited nystagmus should reflect the anatomical orientation of the canal that generates it. However, previous PLF studies have not focused on the relation between canal function and the mechanism that elicits PLF nystagmus. Specifically, how much canal and otolith function is required to still elicit PLF nystagmus?

We report on the three dimensional eye movements of an exceptional patient who suffered from a PLF of the left posterior canal. This is the first report showing that PLF nystagmus of the posterior canal can still be elicited in a completely parietic canal. This finding indicates that PLF might also act directly on the cupula, without using endolymphatic flow in the semicircular canal.

## PATIENT AND METHODS

## Case report

A 32 year old truck driver had a nine month history of recurrent paroxysmal spells of rotational vertigo, oscillopsia, and nausea lasting for seconds. He noticed that the spells were typically elicited by coughing, sneezing, swallowing, hiccupping, or by any kind of physical pressure applied on his left ear, but not by loud sounds. On clinical examination and additional video recordings, after finger tip pressure on the left dorsal aspect of the external ear canal there was a nystagmus with upward and counterclockwise (direction from the patient's point of view) fast phases with a duration of two to three seconds. Except for a left hypacusis, the remaining neurological and orthoptic examination was unremarkable.

High resolution computed tomography revealed a mass on the left temporal bone. The tumour extended from the mastoid into the petrous pyramid, infiltrated the posterior semicircular canal (PC) of the bony labyrinth, and destroyed its medial and lateral bony wall adjacent to the vestibule (fig 1). It did not affect the horizontal or anterior semicircular canal. In two surgical attempts to biopsy (first operation) and to remove the tumour from the mastoid and the medial

aspects of the temporal bone (second operation, three months later) the PLF of the posterior canal could not be closed properly and PLF symptoms persisted. After the second operation he noticed dizziness and severe oscillopsia on particular head movements when turning backward and to the left. Histology from the second operation revealed an eosinophilic granuloma. Search coil recordings of the three dimensional eye nystagmus components were obtained twice, after the first and the second operation.

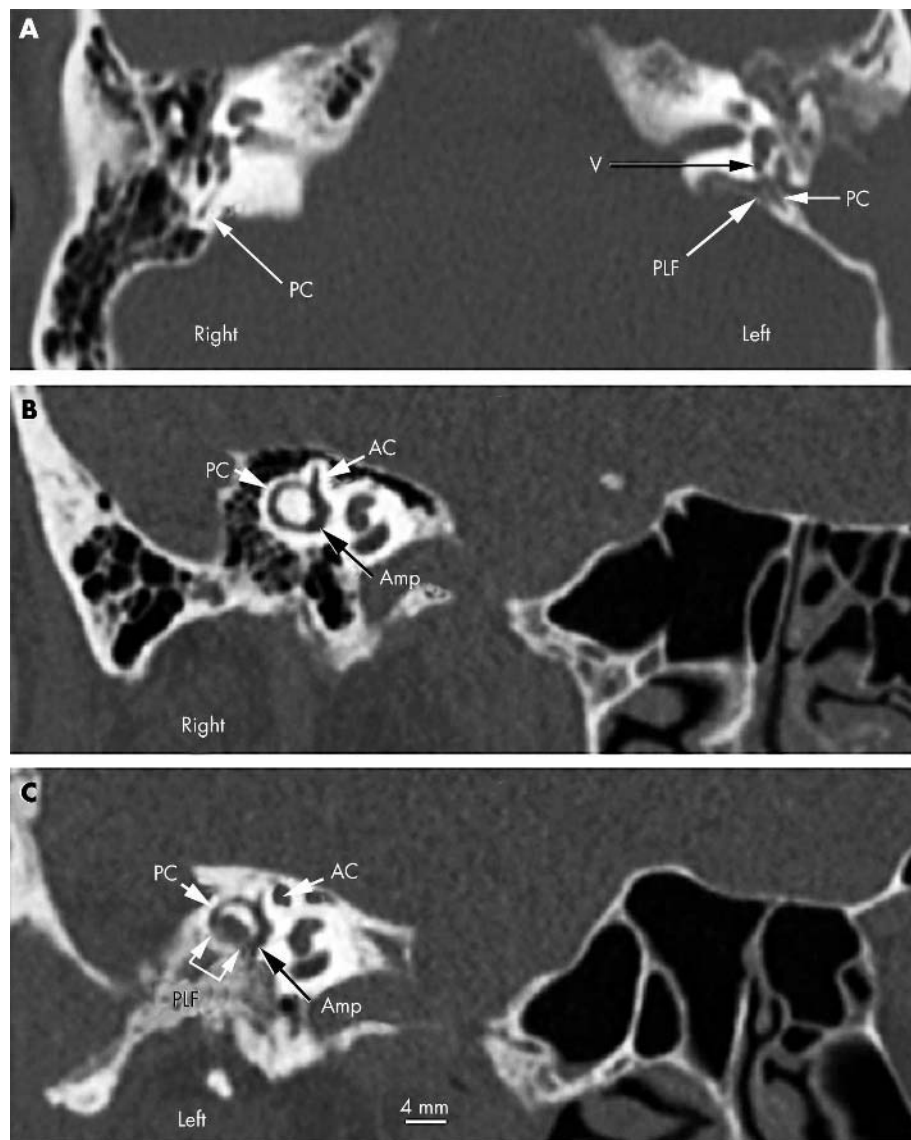
## Methods

Written informed consent was obtained from the patient according to the Declaration of Helsinki. Cochlear function was assessed by audiogram and brainstem evoked response audiometry. Otolith function was obtained by click evoked myogenic potentials (CEMP), funduscopy, and by the subjective visual vertical (SSV).<sup>1,3,4</sup> Semicircular canal function was analysed by head thrusts using scleral search coil recordings (Remmel Laboratories, Ashland, Maryland, USA) with three dimensional coils (Skalar, Delft, the Netherlands).<sup>1,3,4</sup> Eye and head position calibration was performed using a combined offline in vitro and in vivo technique.<sup>3,5</sup> The "eye in space" and "head in space" positions were used to calculate the "eye in head" position.<sup>6</sup> Positive values indicate clockwise torsional, downward, and leftward movements. High acceleration, small amplitude head thrusts in the vertical and horizontal canal planes were used to analyse individual semicircular canal function.<sup>3,7</sup> The gains of the vestibulo-ocular reflex (VOR) were compared with five age matched control subjects (mean age, 29.4; SD, 5.6 years). The median slow phase eye velocity of the spontaneous nystagmus was compared with the orientation of the semicircular canals obtained from anatomical data,<sup>8</sup> as described previously.<sup>1,6</sup>

## RESULTS

There was no spontaneous nystagmus or gaze evoked nystagmus in light or darkness. Physical pressure on the left external ear canal elicited a vigorous upward and counterclockwise (ipsilesionally) beating nystagmus. Its mean (SD) slow phase velocity was 36.5 (17.8) deg/s for the vertical, 27.5 (10.2) deg/s for the torsional, and 1.7 (5.87) deg/s for the horizontal component. The velocity vector orientation of the PLF nystagmus closely aligned with the left PC and did not change after the second operation (fig 2A). Head thrust tests in the canal planes revealed a reduced VOR gain (mean, 0.32; SD, 0.07) in the left posterior canal after the first operation (fig 2B,D) and virtually absent responses (fig 2C,D) to head rotations after the second operation, indicating a loss of left PC function. The left anterior (AC) and left horizontal (HC) semicircular canals were normal (AC: mean, 0.82; SD, 0.15; HC: mean, 0.95; SD, 0.05) and did not differ from the age matched control subjects (AC: mean, 0.79; SD, 0.10; HC:

**Abbreviations:** AC, anterior semicircular canal; CEMP, click evoked myogenic potentials; HC, horizontal semicircular canal; PC, posterior semicircular canal; PLF, perilymph fistula; SVV, subjective visual vertical; VOR, vestibulo-ocular reflex



**Figure 1** (A) Axial computed tomography (CT) scans of the temporal bones showing a large deficit (perilymph fistula; PLF) of the left posterior semicircular canal (PC), which is adjacent to the left vestibule (V). (B, C) Oblique sagittal CT sections of the temporal bone within the plane of the (B) right and (C) left posterior semicircular canal show the proximity of the left PC deficit (double arrow) to the ampulla (Amp). Note the destruction of the bony wall of the inferior limb of the PC adjacent to the ampulla. AC, anterior semicircular canal.

mean, 0.90; SD, 0.02; PC: mean, 0.87; SD, 0.11) and they were also normal on the healthy right side.

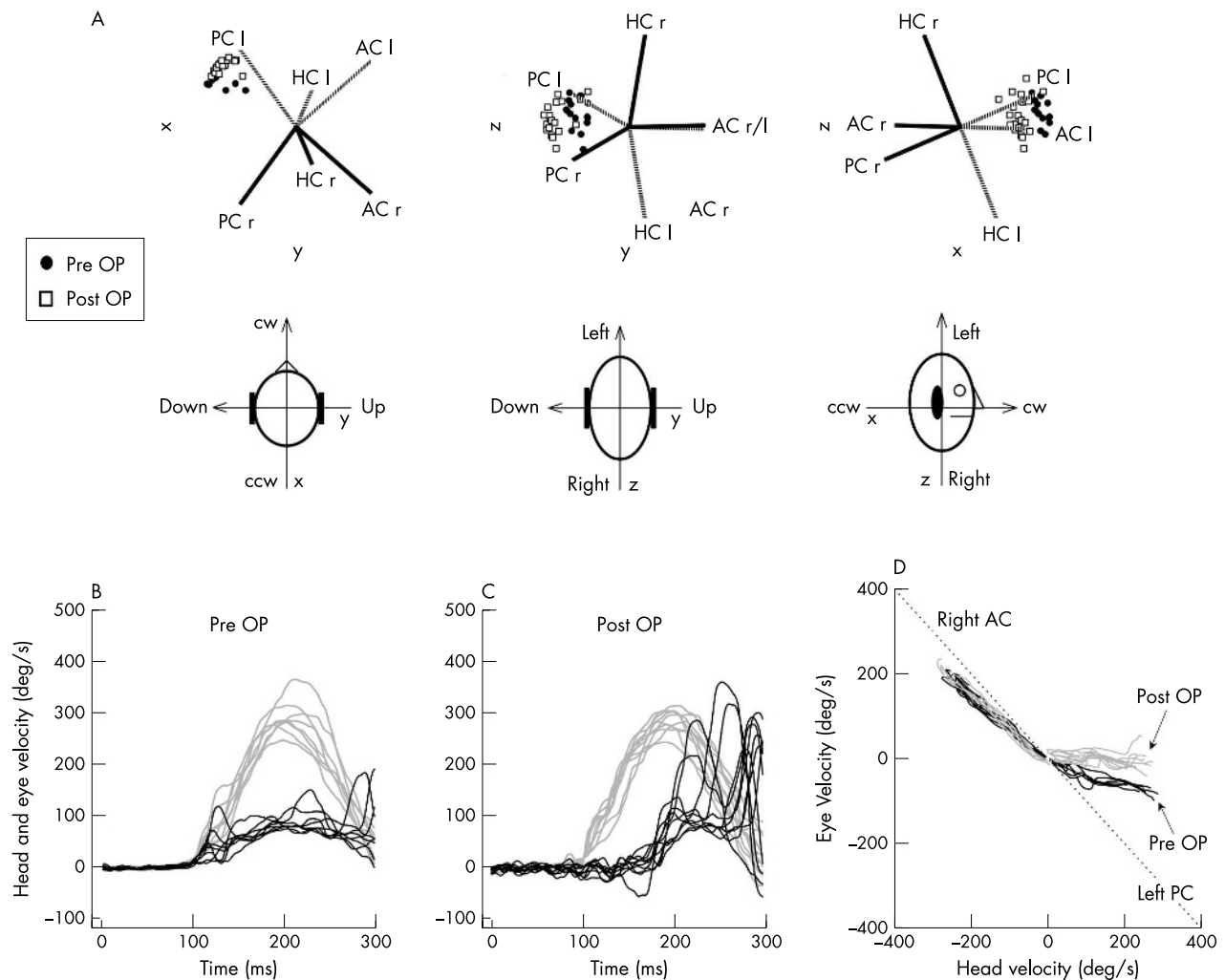
Fundoscopy (right eye: excyclo 2°, left eye: excyclo 7°) and the SVV were normal (2°) at the time of complete canal paresis. SVV was also not pathologically tilted during the provocative pressure application but funduscopy was only measured in the interval. CEMP elicited by 120 dB sound pressure level click stimuli revealed a pathological amplitude ratio of 2.9, with the larger amplitude on the left side (34.5% side difference). Caloric irrigations were normal. Pure tone audiometry revealed a left hypacusis with a conductive hearing reduction of at least 35 dB of the normal hearing level in the low and medium frequencies.

## DISCUSSION

The close alignment of the slow phase velocity vectors of the PLF nystagmus of our patient with the left posterior semicircular canal reflects a pathological stimulation of the left PC and confirms that Ewald's first law is not only valid for the horizontal<sup>1</sup> and anterior canals,<sup>9</sup> but also for the posterior canal.<sup>2</sup> In addition, our patient was unique in that PLF nystagmus of the PC was elicited in the absence of left posterior semicircular canal function. Previous studies on PLFs reported on partially deficient functions of the anterior (gain 0.47,<sup>9</sup> gain 0.43<sup>10</sup>) or posterior canal (gain 0.3<sup>7</sup>), or did

not provide canal specific VOR gain values.<sup>1, 2</sup> Thus, a partially paretic canal is still capable of producing signs of PLF<sup>7, 9, 10</sup> or other canal related disorders, such as horizontal benign paroxysmal positioning vertigo.<sup>3</sup> The exceptional situation of an absent PC function in our patient may shed light on a special pathomechanism of PLF nystagmus.

Compression of the membranous PC by dura may impede endolymphatic flow.<sup>9</sup> Thus, pressure induced PLF nystagmus may come from excitation of the vestibular hair cells of the affected semicircular canal or the otoliths. PLF nystagmus is thought to arise from the semicircular canal, not the utricle,<sup>9, 10</sup> which is in accordance with the absence of utricular dysfunction in our patient (no ocular torsion, SVV, or skew deviation). With a partially deficient canal, negative pressure (for example, finger removal from the tragus) on the PLF causes an ampullofugal deflection of the cupula<sup>2</sup> via endolymphatic flow, which elicits the nystagmus. The PLF nystagmus in a completely paretic canal in our patient might indicate: (1) that the pressure could be transmitted by fluids other than endolymphatic flow acting directly on the cupula; (2) PC plugging, which prevented endolymphatic flow with head impulses, but still elicited endolymphatic flow over a small distance in the proximity of the cupula; (3) hyperexcitability of the cupula hair cells as derived from sensory deprivation; or (4) an intact cupula organ. This is also in line with the



**Figure 2** (A) The normalised rotation axes of the three dimensional slow phase components of PLF nystagmus in the dark are shown after the first (closed circles) and second (open squares) operation in three projections as indicated schematically in the lower traces. For comparison, the axes of the left sided (dashed lines) and right sided (bold lines) canals are given. The nystagmus axes align close to the left posterior canal. x, torsional; y, vertical; z, horizontal; cw, clockwise; ccw, counterclockwise; AC, anterior canal; HC, horizontal canal; l, left; r, right. (B–D) Head thrust tests for the left posterior canal (B) before and (C) after the operation showing partial (B,D) and complete (C,D) canal paresis. Single eye (black lines, inverted for better comparison) and head (grey lines) velocity traces are plotted over time. The delayed high eye velocity responses in (C) reflect refixating saccades. In (D) eye versus head velocity is shown for the first 80 ms after head thrust onset for the right anterior–left posterior canal plane (dashed line indicates a gain of 1; black lines, before; grey lines, after the second operation). There is virtually no eye velocity signal with left posterior canal stimulation.

fact that the large PLF in our patient was adjacent to the ampulla. Thus, PLF nystagmus arises from the canal,<sup>9</sup> but it may not necessarily require a functioning semicircular canal.

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