Persistence of perilymph fistula mechanism in a completely paretic posterior semicircular canal

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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) slow phases with a duration of two to three seconds. Except for the external ear canal there was a nystagmus with upward and counterclockwise 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: mean, 0.90; SD, 0.15). 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: mean, 0.90; SD, 0.15).

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: mean, 0.90; SD, 0.15). 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: mean, 0.90; SD, 0.15).
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 fundoscopy 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 and anterior canals, but also for the posterior canal. 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, gain 0.43) or posterior canal (gain 0.3°), or did not provide canal specific VOR gain values. Thus, a partially paretic canal is still capable of producing signs of PLF or other canal related disorders, such as horizontal benign paroxysmal positioning vertigo. 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. 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, 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 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
fact that the large PLF in our patient was adjacent to the ampulla. Thus, PLF nystagmus arises from the canal, but it may not necessarily require a functioning semicircular canal.

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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.

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