For horizontal canal benign paroxysmal positional vertigo, determination of the pathologic side is difficult and based on many physiological assumptions. This article reports findings on a patient who had one dysfunctional inner ear and who presented with horizontal canal benign paroxysmal positional vertigo, giving us a relatively pure model for observing nystagmus arising in a subject in whom the affected side is known a priori. It is an interesting human model corroborating theories of nystagmus generation in this pathology and also serves to validate Ewald’s second law in a living human subject.

**Key Words:** Benign paroxysmal positional vertigo; lateral canal; horizontal canal; vertigo; positional vertigo; Ewald; human model; in vivo; ageotropic.

**INTRODUCTION**

We present a rare case combining horizontal canal benign paroxysmal positional vertigo (HCBPPV) with a dead ear on the contralateral side, therefore arising from a known side. This case acts as a human model to corroborate theories of nystagmus generation in HCBPPV, which are used to pick the side of pathology, and also serves to again validate Ewald’s second law in a living human subject.

Benign paroxysmal positional vertigo is a very common cause of episodic vertigo, with the posterior semicircular canal (SCC) being affected most frequently. The associated nystagmus seems to follow nystagmus characteristics that would be predicted by particles (presumed to be otoconia) acting under gravity in a fluid medium in the posterior SCC, and the affected side is usually easy to determine, as the nystagmus typically only arises with the affected ear down, nearly always has a latency, and follows a stereotypical pattern, being geotropic (i.e., superior pole of eye fast phase toward ground).

HCBPPV has also been described. It is much less common and usually more difficult to deal with, both in diagnosing the side of origin and in its response to therapy. Identifying the pathologic side in HCBPPV is a challenge for a number of reasons. First, when performing the diagnostic maneuver (the supine head roll test), unlike the posterior canal, one horizontal canal (HC) cannot be isolated from the other, and thus both test positions always affect both HCs. Thus, despite only one ear being affected by HCBPPV, nystagmus is always seen with either ear down. Furthermore, HCBPPV causes direction-changing nystagmus that can be geotropic or ageotropic (i.e., fast phase beating either toward or away from the dependent ear, respectively), depending on the specific pathology. In either case, however, for a given pathology, the direction of the nystagmus relative to the ground is the same in both positions of the supine head roll test (i.e., geotropic on both sides or ageotropic on both sides).

Various theories, outlined briefly below, are used to explain the nystagmus seen according to specific pathology. Geotropic nystagmus is generally ascribed to canalolithiasis in the posterior arm of the HC. The theory is that with the affected ear down, as the canaliths float under gravity, they deflect the cupula toward the utricle, thus exciting the HC and resulting in geotropic nystagmus (Fig. 1A). When the subject is rolled so that the same canaliths in the affected ear now float away from the cupula, deflecting it away from the vestibule, inhibiting the afferent resting firing rate, and resulting in nystagmus away from the affected ear, which is now up, causing geotropic nystagmus once again (Fig. 1B). The ageotropic type is thought to be caused by either canaliths in the anterior arm of the canal (Fig. 1B) or by cupulolithiasis (Fig. 1C) (otooliths attached to the cupula). With the affected ear down, either will move the cupula under gravity away from the vestibule and inhibit the resting firing rate in the ampullary nerve, resulting in nystagmus beating away from the affected ear (i.e., ageotropic). Turning the patient so that the
affected ear is up will result in movement of the cupula toward the vestibule, thus exciting the affected HC and resulting in nystagmus directed toward the affected ear, thus ageotropic once again. Canalolithiasis and cupulolithiasis are theoretically distinguished by the latency of onset and duration of nystagmus, with canalolithiasis having a latency as particles move and a relatively short duration as they settle to the most dependent portion of the canal, whereas cupulolithiasis converts the cupula into a gravity sensor, with no latency in onset and prolonged duration of nystagmus, as the cupula stays deflected as long as gravity acts on the higher density mass attached to it.

Therefore, identifying laterality of HCBPPV is challenging for the following reasons: canalolithiasis of the posterior arm in either lateral SCC will cause geotropic horizontal nystagmus on both sides when performing the supine head roll test, and canalolithiasis of the anterior arm of the lateral SCC and cupulolithiasis of either lateral SCC produces ageotropic nystagmus in both test positions. Furthermore, there is no way to isolate one HC from the other during the supine head roll test. In the face of these laterization challenges, Ewald’s second law (i.e., nystagmus amplitude is larger with excitatory cupula movement vs. inhibitory) is usually invoked to select the affected side by identifying the side with the greater nystagmus amplitude and working backward through the theories described above. In geotropic HCBPPV, the nystagmus amplitude is greater with the affected side down. In ageotropic HCBPPV, the opposite is true.

It must be emphasized that these are theoretical models and have never been validated with direct dynamic observations of movements in the canals under gravity. The latter would be impossible in humans with current imaging technologies. We were fortunate to observe a subject in whom one inner ear was dysfunctional by disease, with HCBPPV, giving us a relatively pure model for observing nystagmus arising in a subject in whom the affected side is known a priori. This is an unusual situation and can help us validate current theories of HCBPPV.

**CASE REPORT**

An 87-year-old male presented with HCPPV and a history of invasive cholesteatoma of his left ear 10 years prior. His cholesteatoma had fistulized into the HC, and preoperatively he manifested a dead ear audiologically, as well as a positive head thrust to the diseased ear. He had undergone a canal wall down tympanomastoidectomy, including removal of disease from a large fistula in the lateral canal. The fistula was quite extensive, with granulation tissue extending to within the canal’s lumen. Lack of caloric responses in the left lateral SCC was confirmed postoperatively, thus leaving only the right horizontal SCC functioning.

A number of years after his surgery, the patient presented with complaints of chronic dysequilibrium and slight nausea that had been ongoing for some months, particularly associated with head movements. He did not have any other neurologic or new otologic complaints. On physical exam, the patient had a normal cranial nerve and cerebellar exam, and once

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again strongly positive head thrust to the left. Although Dix-Hallpike did not evoke torsional nystagmus, a supine head roll test demonstrated bilateral ageotropic purely horizontal nystagmus, being more marked with the head turned to the left (i.e., toward the nonfunctioning ear with the presumed affected ear up). The nystagmus began immediately in each position (no latency) and persisted throughout the position (not fatiguable).

The same positioning tests were done under video nystagmography (VNG) for quantification and to confirm the clinical impression. Figure 2 demonstrates the bilaterally ageotropic nystagmus that begins immediately, once the lateral head position is reached. VNG also demonstrated the increased slow phase velocity of the nystagmus when the head is turned to the left, that is, away from the presumed affected ear, which is consistent with that which would be predicted using Ewald’s second law. Figure 2 clearly demonstrate Ewald’s second law. Figure 2 demonstrates bilaterally ageotropic nystagmus that begins immediately, once the lateral head position is reached. VNG also demonstrated the increased slow phase velocity of the nystagmus when the head is turned to the left, that is, away from the presumed affected ear, which is consistent with that which would be predicted using Ewald’s second law. Figure 2 clearly demonstrate Ewald’s second law. Figure 2 clearly demonstrate Ewald’s second law. Figure 2 clearly demonstrate Ewald’s second law. Figure 2 clearly demonstrate Ewald’s second law.

DISCUSSION

This clinical capsule is interesting in that it represents a human model corroborating the lateralization hypotheses for the ageotropic type of HCBPPV. As mentioned above, Ewald’s second law has been previously used to rationalize the identification of the affected ear. Calorics test mostly HC function at low frequencies, and cupulolithiasis, being a static deflection, is a very low-frequency stimulus. Therefore, the absence of left caloric responses could be argued to effectively rule out the left ear as a cause for the HCBPPV in this case. Other factors pointing to this ear being disabled are the strongly positive horizontal head thrust to the left (an HC test), and the presence of the HC fistula at surgery, which is likely to have significantly damaged the HC and the entire inner ear, as all hearing was also lost in the left ear. We can thus be reasonably certain that any dynamic pathology being manifest in this patient is coming from his right ear, but of course we cannot be completely sure.

He manifests the ageotropic type of HCBPPV on supine head roll testing. As mentioned above, this would suggest either canalolithiasis in the anterior arm of the HC, or cupulolithiasis in the HC. The very short latency and prolonged duration of the nystagmus in this case suggests cupulolithiasis. In keeping with theory, this did produce ageotropic nystagmus, and as predicted by Ewald’s second law, the amplitude of the nystagmus was higher with the affected (right) ear up.

CONCLUSION

For HCBPPV, determination of the pathologic side is difficult and based on many physiological assumptions. With the exclusion of one horizontal SCC, this case allows us to verify that, for cupulolithiasis of the HC, our assumptions are valid in
picking the correct ear affected, and so serves as a useful in vivo model.

BIBLIOGRAPHY


