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Unilateral Horizontal Semicircular Canal Malformation Causing Recurrent Vertigo

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A 62-year-old man consulted for recurrent episodes of vertigo lasting from seconds to several minutes. The vertigo was variably described as spinning, lateral swaying and a feeling of being "unsure of his position in space." These episodes were noted to have begun when the patient was still in his 20's. Standard pure tone audiometry revealed a mild-to-moderate downsloping mixed hearing loss in the left ear. Bithermal caloric testing indicated the presence of a significant left-sided peripheral vestibular loss. Due to the fact that the vertigo episodes presented relatively early in life, the possibility of a congenital inner ear malformation was considered as a cause for his symptoms. Computerized tomographic (CT) imaging of the temporal bone was performed. This clearly showed the left horizontal semicircular canal lacking a central bony island. *(Figure 1 and 2)* The cochlea, superior and posterior semicircular canals, vestibular and cochlear aqueducts and ossicular chain were grossly normal.

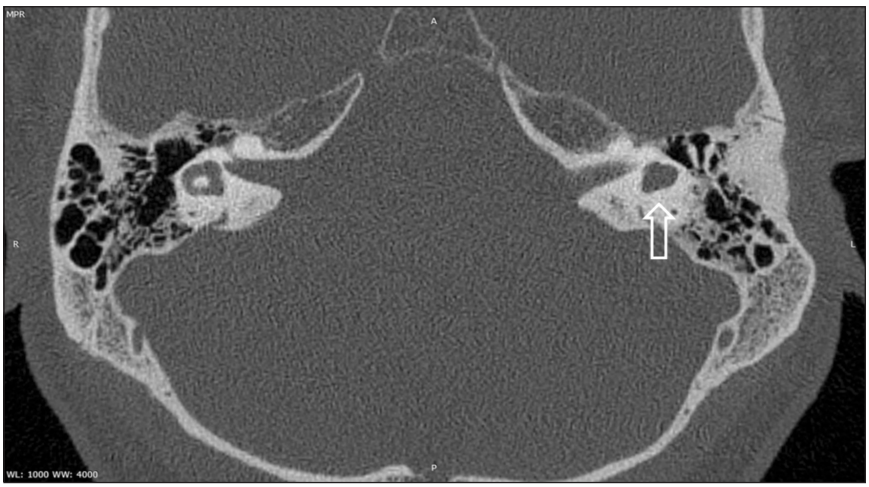

Figure 1. Computerized tomographic imaging of the temporal bone in the axial view at the level of the horizontal semicircular canal. The arrow points to the left horizontal semicircular canal which lacks a central bony island and has a cystic appearance. In comparison, the right horizontal semicircular canal has the classic "signet ring" appearance.

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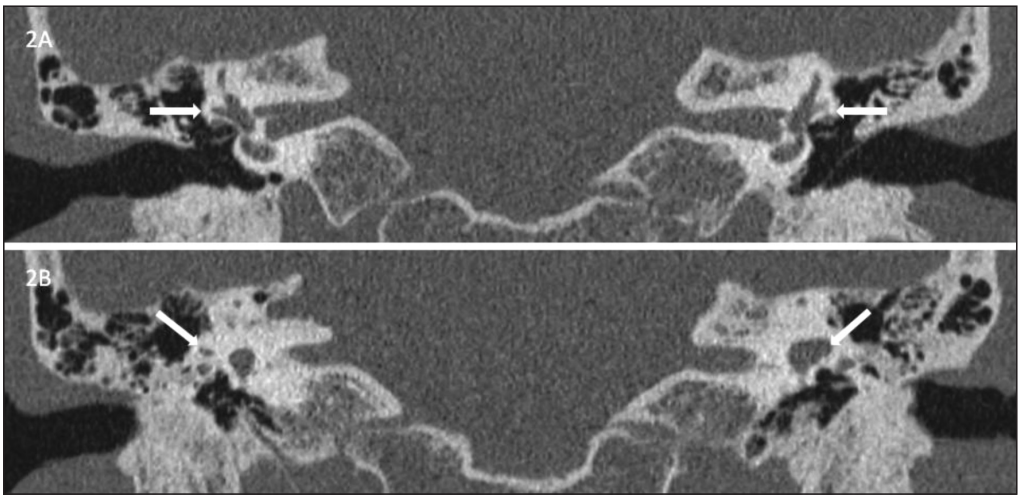

Figure 2. Computerized tomographic imaging of the temporal bone in the coronal view showing the horizontal semicircular canals at two different levels. **A.** is at the level of the ampullated end of the canal, where the canals look similar (white arrows). **B.** is at the midpoint of the canal where the right side shows a small ovoid lumen separated from the vestibule by bone; whereas the left side shows an enlarged lumen representing the combined vestibule and semicircular canal without any intervening bone (angled white arrows). These images illustrate the difficulty in identifying the abnormality on coronal view as compared to the axial view.

A malformation of the horizontal or lateral semicircular canal is one of the most common inner ear malformations since it is the last vestibular structure to be formed during inner ear embryogenesis. As such, it may occur in isolation or may be associated with other vestibular, cochlear, or middle ear malformations.[1,2] Although vertigo and dizziness are symptoms to be expected in such a condition, existing data indicates that it may be totally asymptomatic or it may also present as a sensorineural, conductive or mixed type of hearing loss.[1,3] Radiologic imaging is of prime importance in diagnosing such conditions especially when auditory and/or vestibular symptoms manifest early in life. This case perfectly illustrates the need for such studies as the patient went undiagnosed for more than forty years!

No definitive statements can be gleaned from existing medical literature with respect to treatment. However, in patients with debilitating vestibular symptoms, management with modalities that selectively target the vestibular system, but spare the auditory system, such as vestibular neurectomy and trans-tympanic aminoglycoside therapy appear to be reasonable options.

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