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Cystic Cochleovestibular Malformation (Incomplete Partition Type 1)

A 5-year-old female with bilateral profound hearing loss underwent computerized tomographic imaging of the temporal bone as part of the work-up to determine the etiology of her deafness and to delineate middle and inner ear anatomy prior to cochlear implantation. The examination revealed an inner ear malformation which based on the newest classification of cochleovestibular malformations by Sennaroglu and Saatci, is called an ***incomplete partition type I (IP-1)*** or ***cystic cochleovestibular malformation***. This condition is characterized by: (1) a cochlea that is lacking the entire modiolus and cribriform area resulting in a cystic appearance; and (2) a large cystic vestibule.[1]

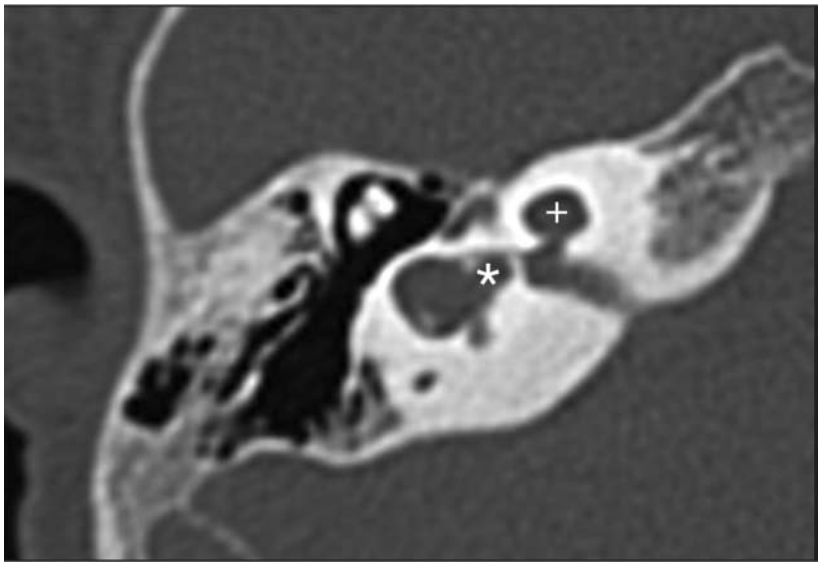

Figure 1. Axial view of the abnormal inner ear showing the cystic cochlea (+) and the cystic vestibule (*) without the central bony island usually found in the horizontal semicircular canal.

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Temporal bone imaging is among the most useful examinations in the etiological investigation of idiopathic sensorineural hearing loss in children, with up to 30%[2] of the imaging studies showing an abnormality. The detection of inner ear malformations is important as some abnormalities are associated with an increased risk of meningitis or progressive hearing loss following head trauma.[3] Likewise, the approach to cochlear implantation may be influenced by the type of malformation. In this particular patient, the use of a cochlear implant with a full-band electrode design may be more appropriate, as the location of the neural elements within the cystic cochlea is not definitely known.

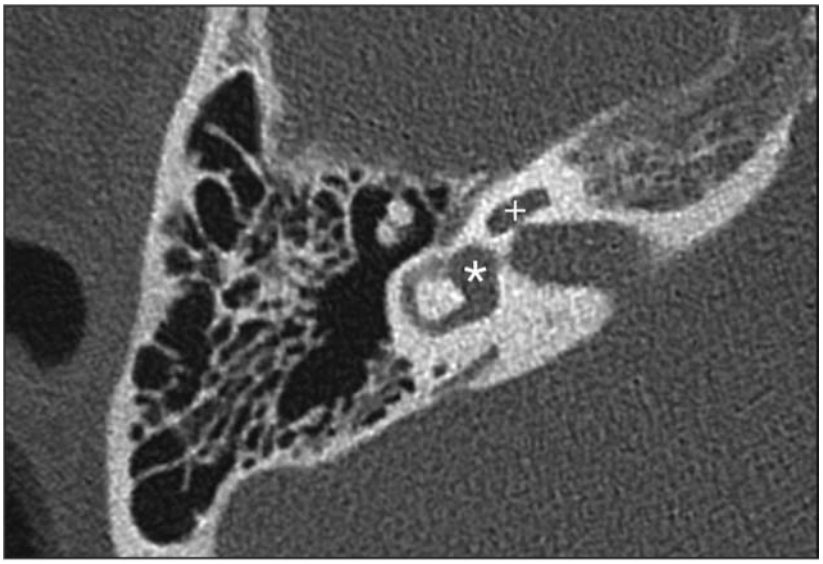

Figure 2. Axial view of a normal inner ear at the same level as Figure 1, showing the "signet ring" configuration of the horizontal semicircular canal and vestibule (*) and the first turn of the cochlea (+).

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