Unilateral Mondini dysplasia

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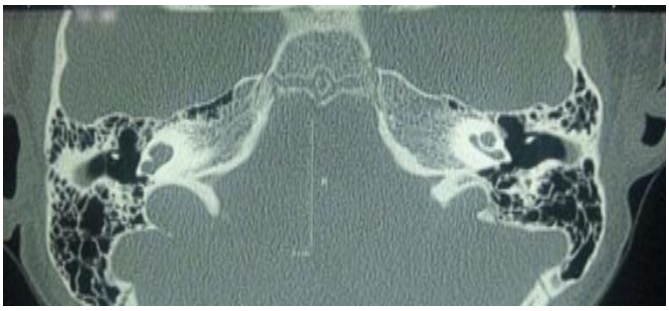
Abstract:



Incomplete Partition (Mondini)

Arrest at the seventh week of gestation yields a cochlea that has only 1.5 turns. This is the most common type of cochlear malformation, accounting for more than 50% of all such deformities. Radiographically, the cochlea is smaller than normal and partially or completely lacks an interscalar septum although the cochlea usually measures 8 to 10 mm vertically; it is typically in the 5- to 6-mm range in incomplete partition deformity. Care must be exercised in counting the number of cochlear turns radiographically because this may be difficult to determine even using high-resolution CT. The radiographic diagnosis depends more on cochlear size and the absence of a scalar septum than on the number of cochlear turns perceived. Histologically, incomplete partition appears to be the radiographic correlate of classical Mondini's dysplasia .In numerous reported cases, a small cochlea with 1.5 turns possessing an apical scala communis secondary to deficiency in the osseous spiral lamina has been described . Sennaroglu and Saatci have subtyped the incomplete partition deformity into three variants. Type I lacks the entire modiolus and interscalar septa and demonstrates a cystic appearance. Type II has a normal base turn but a cystic apex ("Mondini type"). Recently Sennaroglu has proposed a type III variant with deficient modiolus and partial interscalar septation at the cochlea's periphery. Organ of Corti development is variable, as is the auditory neural population. As might be expected, auditory function also is variable, ranging from normal to profound SNHL.SCC deformities accompany incomplete partition of the cochlea in approximately 20% of cases. Mondini deformity is mostly bilateral finding. I reported a case of unilateral Mondini deformity of a male child 3 years old presented with severe to profound hearing loss (Figure).

Biography

Dr.Mahmoud farouk, MSc. Otology and Audiology-Egypt.He is manager and owner at Hearing Care Clinic otologist and Vestibular Medicine at Egypt Air Hospital and Otologist at Wadi El-Nile Hospital. He **Studied Otorhinolaryngology-Head & Neck Surgery Board at faculty of medicine Ain_shams university.he lives in Cairo ,Egypt**

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