

Large Vestibular Aqueduct Syndrome

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Summary

Large Vestibular Aqueduct Syndrome is a congenital malformation of the temporal bone characterised by early onset of sensorineural hearing loss and vestibular disturbance. Familial large vestibular aqueduct syndrome suggests autosomal recessive or X-linked inheritance and accounts for non-syndromic sensorineural hearing loss in these patients.

Key Words: Sensorineural hearing impairment, Congenital, Computed Tomography (CT)

Case Report

A 6 years old Chinese female presented to the ENT Outpatient Department with right sided hearing loss. At age of 4 years, she had a sudden hearing loss in her right ear, which recovered spontaneously after one month. Similar attacks occurred frequently, more so during attacks of common cold. She gave no history of vertigo or symptoms suggestive of vestibular disturbance. Physical examination revealed no abnormalities. Craniofacial structures were normally developed. The tympanic membranes were intact and mobile with no evidence of ossicular malformation. The cranial nerves were intact bilaterally; cerebellar testing and Romberg's test were normal.

The audiogram at her first visit showed a right profound sensorineural hearing loss. One month later, her hearing had improved, particularly in the lower frequencies (Figure 1).

A temporal bone Magnetic Resonance Imaging (MRI) scan was performed and demonstrated a right large vestibular aqueduct (Figure 2). Measurement of the aqueduct diameter was performed halfway between the common crus (superior and posterior semicircular canals) and the external aperture (posterior fossa). The

vestibular aqueduct measured 4mm on the right as compared to 1mm on the left. (In normal temporal bones, the size of vestibular aqueduct ranged from 0.4 to 1.0mm diameter).

Discussion

Large vestibular aqueduct is characterized by bony dilatation within the posterior petrous bone and is thought to be an abnormal enlargement of the endolymphatic sac and duct. It has been reported in association with other inner ear anomalies or as a separate radiologic entity in some patients¹. Valvassori and Clemis² first described Enlarged Vestibular Aqueduct Syndrome (EVAS) as congenital malformation of the temporal bone predisposing the affected person to early onset hearing loss and vestibular disturbance.

Although enlarged vestibular aqueduct syndrome is a congenital condition, hearing loss may not be present or apparent from birth. Many children with enlarged vestibular aqueduct syndrome demonstrate a post lingual onset of deafness. Clinical findings include fluctuating and sometimes progressive sensorineural hearing loss and vestibular symptoms. A conductive or mixed component can be observed on audiometric

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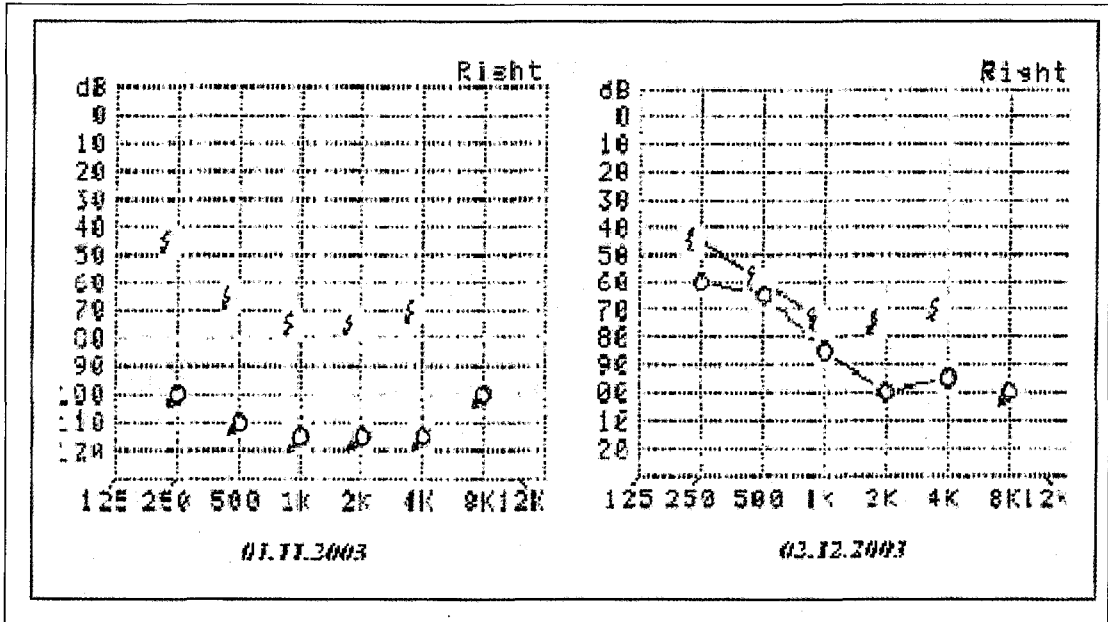


Fig. 1: Audiograms of patient at first visit (left) and at 1 month later (right)

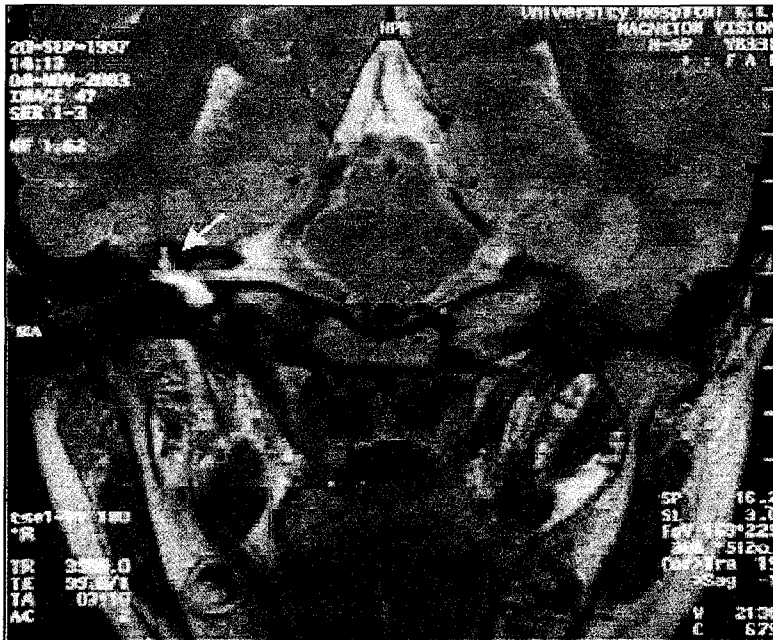


Fig. 2: Magnetic Resonance Imaging (MRI) Scan demonstrating a Large Vestibular Aqueduct (arrow)

evaluation. The average conductive gap ranged from 10 to 50 dB and conductive hearing loss was most prominent at 250 to 500 Hz (low frequencies) despite a clinically normal ear and a normal tympanogram in 28% of the patients¹. Vestibular symptoms range from severe episodic vertigo to occasional unsteadiness in adults, where as incoordination and imbalance predominate in children³. The age of diagnosis of the hearing loss has been found to be variable ranging from infancy to adulthood. However the progression of sensorineural hearing loss varies between studies from 12 to 65%³.

Enlarged vestibular aqueduct is the most common form of congenital inner ear abnormality seen on radiologic assesment. The prevalence of enlarged vestibular aqueduct syndrome has been estimated to range from 1-12% in population with sensorineural hearing loss¹. More recently improved CT scan and heightened clinical awareness of enlarged vestibular aqueduct syndrome has resulted in vestibular aqueduct anomalies being identified in up to 14% paediatric sensorineural hearing loss patient. A female to male ratio of 3:2 and a bilateral versus unilateral ratio of 2:1 has been reported¹.

Associated inner ear anomalies such as an enlarged lateral semi circular canal were found in 60 to 80% of the patient's CT scans¹. Other ear anomalies were an abnormally enlarged vestibule and a dysplastic cochlear.

Parents of children with large vestibular aqueduct syndrome clearly demonstrate no abnormality either

based upon clinical, audiologic, vestibular, cytogenetic or radiographic analysis. This suggests either autosomal recessive inheritance or X-linked inheritance with mother being a carrier. Modifying genes and stochastic factors are likely to account for the variations observed within the family and in general population.

Enlarged vestibular aqueduct syndrome has been associated with a range of congenital disorder such as CHARGE Syndrome, Alagille Syndrome, Pendred Syndrome and Brachio otorenal Syndrome¹. High resolution, thin section CT scanning of the temporal bone has been the mainstay of inner ear imaging in children with sensorineural hearing loss because of its high spatial resolution and contrast. The criteria for determining an enlarged vestibular aqueduct are somewhat vague. A vestibular aqueduct diameter larger than 1.5 mm at the midpoint (half way point between the operculum and the posterior wall of the crus commune of vestibule) or an opercular measurement of greater than 2mm are generally considered to be defining characteristics of this syndrome¹.

The ability to identify children with progressive hearing loss would significantly improve the clinical, communicative and educational planning and management of children at risk¹. Early auditory amplification, speech and language therapy and counseling regarding the risk of head injury are the mainstay of treatment eg. avoidance of contact sport (football), restriction of activities, routine use of seat belts in the car and a helmet during bicycling, skateboarding and rollerblading¹.

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