



A Brief Review on Large Vestibular Aqueduct

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Large vestibular aqueduct, is a structural deformity of the inner ear. Enlargement of this duct is one of the most common inner ear deformities and is commonly associated with hearing loss during childhood[1]. The term was first discovered in 1791 by Mondini when he was completing a temporal bone dissection. It was then defined by Valvassori and Clemis as a vestibular aqueduct that is greater than or equal to 2.0 mm at the operculum and/or greater than or equal to 1.0 mm at the midpoint. Some use the term enlarged vestibular aqueduct syndrome but this is felt by others to be erroneous as it is a clinical finding which can occur in several syndromes[2].

Although large vestibular aqueducts are a congenital condition, hearing loss may not be present from birth. Age of diagnosis ranges from infancy to adulthood, and symptoms include fluctuating and sometimes progressive sensorineural hearing loss and disequilibrium. It is possible that a small portion of those with large vestibular aqueducts will not exhibit auditory symptoms such as hearing loss. While this is uncommon, hearing loss can progress later in life especially after head trauma. Hearing loss can be unilateral or bilateral and can be sensorineural, conductive, or mixed. Vestibular symptoms, such as vertigo and imbalance, occur in adult patients 45% of the time.

Enlarged vestibular aqueducts are commonly picked up after newborn hearing screen when a child is identified as having a hearing loss. The hearing loss is commonly mixed and can be of any degree when first identified. The conductive component is due to a third window effect caused by the widened vestibular aqueduct. During an audiologic assessment, an air-bone gap in the low frequencies can occur due to the enlarged vestibular

aqueduct acting as a third mobile window[3,4]. Hearing loss in the contralateral ear to the enlarged vestibular aqueduct can also occur due to the chemical composition on both sides. Torticollis has also been found to be an indicator of an enlarged vestibular aqueduct in children. Identification of the enlarged vestibular aqueduct in a child is usually by MRI scan which identifies the fluid within the endolymphatic duct and sac. CT scan may be needed to see the vestibular aqueduct clearly. In adults CT scan may be the first investigation. In order to diagnose the cause of the enlarged vestibular aqueduct the physician will need a detailed family history, full examination to include vestibular examination and, if a bilateral finding, molecular genetic testing as appropriate. With unilateral enlarged vestibular aqueducts molecular genetic testing is currently not recommended.

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