

Pediatric Clinical Support: Enlarged Vestibular Aqueduct Syndrome (EVAS)

Also known as Large Vestibular Aqueduct Syndrome, EVAS is a non-congenital, syndromic form of hearing loss caused by an enlarged vestibular aqueduct*, usually with a diameter larger than 1.5mm (Valvassori & Clemis, 1978). In EVAS, the endolymphatic duct and sac are also enlarged (see Figure 1), which are believed to cause hearing and balance problems. Approximately 1-1.5% of individuals with SNHL or vestibular problems have EVAS. Five to fifteen percent of children with sensorineural hearing loss have EVAS (<http://www.nidcd.nih.gov/health/hearing/Pages/eva.aspx>).

Characterized by: Bouts of vertigo with hearing loss, usually precipitated by a head injury, changes in barometric pressure, or after physical exertion. Hearing loss can be congenital and/or of unknown causes, with no cochlear deformities.

Anatomical and Physiological Development:

During gestational development, the vestibular aqueduct is short, straight, and broad. Following birth, it continues to elongate and grow into an inverted “J” shape, reaching an adult length of 10mm by 3 to 4 years of age, with an average diameter of 0.8mm (Madden et al, 2003; Dewan, Wippold, & Lieu, 2009). Since it is one of the last inner-ear structures to mature, it is highly vulnerable to traumatic insults, usually resulting from abnormal postnatal or during early childhood development.

Manifestation: May result in sudden, fluctuating, or progressive SNHL with a pseudo-conductive hearing loss (Merchant et al, 2007). A “step-wise” pattern in hearing loss has been observed in EVAS patients, where a drop in hearing may occur following a mild head injury. This pattern can occur either gradually or suddenly, and thresholds normally do not recover to previous levels. Research (Abe, et al, 1999; Madden, et al, 2007) has indicated that genetic defects related to chromosome 7q31 (inherited recessive trait) and SLC26A4 overlap with chromosomal markers for Pendred Syndrome, and have suggested that individuals with EVAS are predisposed to developing hearing loss, but may not have it at birth (Arjmand & Weber, 2004; Madden et al, 2003).

Differential Diagnosis: Pendred Syndrome, Meniere’s disease, Connexin 26, and Connexin 30. Cardiac, thyroid, and renal function should be evaluated, as EVAS can be associated with Branchiootorenal (BOR) syndrome. EVAS was originally thought to be a variation of Mondini’s cochlea and is now considered a separate

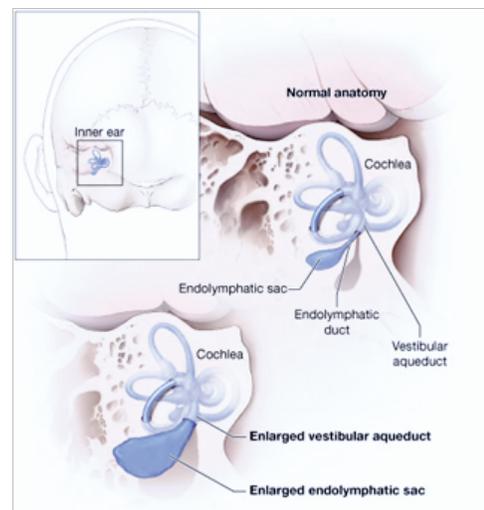


Figure 1. Graphic depicting normal endolymphatic sac and duct, and vestibular aqueduct, and an enlarged vestibular aqueduct and endolymphatic sac. <http://www.nidcd.nih.gov/health/hearing/pages/eva.aspx>

condition; however, EVAS can be associated with Mondini’s cochlea.

Diagnosis & Prognosis: Usually done by positively identifying EVAS on a CT-scan (preferred diagnostic tool) or confirming an enlarged endolymphatic duct and sac on high-resolution MRI following diagnosis of SNHL.

There is no cure for EVAS, although early diagnosis can help with making appropriate educational and therapeutic decisions. Precautions should be considered to avoid further hearing loss, including avoiding possible causes of head trauma (e.g., avoiding contact sports), or abrupt or jarring movements.

The vestibular aqueduct is a narrow bony “canal” that runs between the inner ear and cranial cavity, with a normal diameter ranging from 0.5 to 1.4mm. The endolymphatic duct is located in the vestibular aqueduct. When enlarged, endolymph may travel back to the endolymphatic sac in the inner ear, causing abnormal or delayed cochlear development.

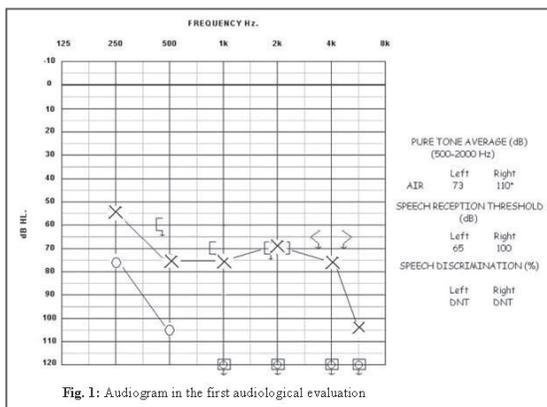


Fig. 1: Audiogram in the first audiological evaluation

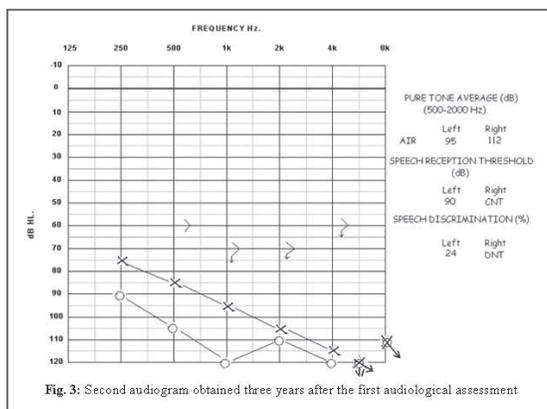


Fig. 3: Second audiogram obtained three years after the first audiological assessment

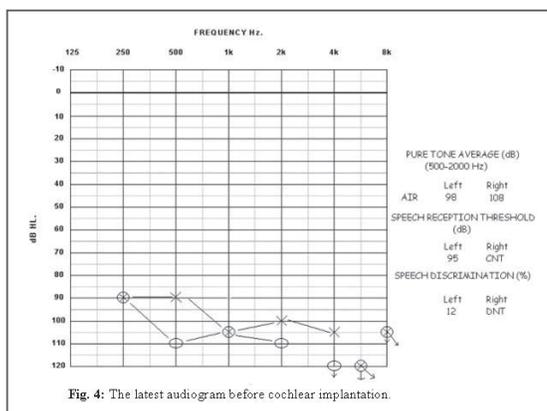


Fig. 4: The latest audiogram before cochlear implantation.

Figures 2-4. Progression of EVAS in a child.

<http://www.marmaramedicaljournal.org/text.php3?id=235>

Online Support Sources and References:

<http://health.groups.yahoo.com/group/LVAS/>
<http://www.parentcenternetwork.org/parentcenterlisting.html>
<http://hearinglosshelp.com/weblog/category/ear-problems/large-vestibular-aqueduct-syndrome>
<http://evahelp.org/othersites.html>
<http://www.ncbi.nlm.nih.gov/sites/ga?disorder=Enlarged%20vestibular%20aqueduct%20syndrome>
<http://www.nidcd.nih.gov/health/hearing/pages/eva.aspx>
<http://vestibular.org/enlarged-vestibular-aqueduct-syndrome-evas>
<http://www.hearinglosshelp.com/articles/lvas.htm>
<http://www.marmaramedicaljournal.org/text.php3?id=235>

Audiological Findings: Characteristic pattern, where hearing is normal for the first years of life, then hearing loss can develop and progressively worsen. Figures 2 through 4 provide serial audiograms for a child diagnosed with EVAS. Head trauma can exacerbate the hearing loss, as can anything that can increase intra-cranial pressure (e.g., congestion related to the common cold, rigorous exercise, etc.). One-third of EVAS patients report some degree of vertigo (www.hearinglosshelp.com/articles/lvas.htm).

Treatment and Rehabilitation: Steroid use for sudden hearing loss has not been proven effective for EVAS. Surgical shunting or removal of the endolymphatic sac is ineffective and considered harmful and can further progress hearing loss or destroy hearing altogether. Hearing aids, preferably with a large range of gain, are beneficial in the event of fluctuating or progressive hearing loss. Cochlear implants are a viable option if hearing loss becomes profound and has stabilized. FM systems or other ALDs should be considered when in environments where SNR is poor.

Professional Considerations: Quality of life considerations may have to be addressed if the child struggles with vertigo. A physical therapist may be helpful with improving the child's balance. SLPs, Audiologists, and child psychologist may all interact with the child and assist in different aspects of habilitation, including speech/language therapy, use of hearing aid/cochlear implant and FM use, and coping with stress.

Other References:

Abe S, Usami S, Hoover DM, Cohn E, Shinkawa H, Kimberling WJ. (1999). Fluctuating sensorineural hearing loss associated with enlarged vestibular aqueduct maps to 7q31, the region containing the Pendred gene. *Am J Med Genetics*, 82:322-- 328.

Arjmand, E.M., Webber, A. (2004). Audiometric findings in children with a large vestibular aqueduct. *Arch Otolaryngol Head Neck Surg.*, 130:1169-74.

Dewan, K., Wippold, F.J., Lieu, J.E. (2009). Enlarged Vestibular Aqueduct in Pediatric SNHL. *Otolaryngology Head and Neck Surgery*, 140(4): 552-558.

Madden, C., Halsted, M., Benton, C., Greinwald, J., Choo, D. (2003). Enlarged vestibular aqueduct syndrome in the pediatric population. *Otol Neurotol*, 24:625-632.

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Merchant, S.N., Nakajima, H.H., Halpin, C., et al (2007). Clinical Investigation and Mechanism of Air-Bone Gaps in Large Vestibular Aqueduct Syndrome. *Annals of Otolology Rhinology and Laryngology*, 116(7): 532-541

Valvassori, G.E., Clemis, J.D. (1978). The Large Vestibular Aqueduct Syndrome. *Laryngoscope*, 88(5): 723-728.