Large Vestibular Aqueduct Syndrome: An Overlooked Etiology for Progressive Childhood Hearing Loss

Diana M. Callison
Karl L. Horn

Abstract

If a drastic change in hearing has occurred in a child following a minor head trauma, change in barometric pressure, or physical exertion, large vestibular aqueduct syndrome (LVAS) should be considered. Most audiologists are unaware of LVAS or do not suspect it, in part due to the presence of a conductive component. LVAS can be seen in conjunction with Mondini's dysplasia or may appear by itself and is easily identified by a computed tomography scan. We present five cases of LVAS and discuss the natural history, audiologic and imaging findings, and relevant literature.

Key Words: Children, computed tomography scans, fluctuating hearing loss, large vestibular aqueduct syndrome

Abbreviations: ABG = air-bone gap, ABR = auditory brainstem response, CSF = cerebrospinal fluid, CT = computed tomography, FM = frequency modulation, FSE = fast spin echo, LVA = large vestibular aqueduct, LVAS = large vestibular aqueduct syndrome, SNHL = sensorineural hearing loss

More than 200 years ago, temporal bone dissection by Carlo Mondini revealed wide vestibular aqueducts in patients with cochlear dysplasia (Mondini's dysplasia). In 1978, Valvassori and Clemis were the first to use imaging to identify 50 patients with large vestibular aqueducts (LVAs). In their report, they noted an association between enlargement of the vestibular aqueduct and sensorineural hearing loss (SNHL). The association of this anatomic anomaly with SNHL led them to coin the term "large vestibular aqueduct syndrome" (LVAS) (Phelps, 1996). Jackler and De La Cruz (1989) have stated that LVAS should be diagnosed when a LVA is the only "anomaly of the inner ear evident on radiographic studies."

The prevalence of an enlarged vestibular aqueduct is thought to be relatively uncommon. A review by Levenson et al (1989) of 1700 computed tomography (CT) scans revealed 12 children (0.64%) who were found to have a LVA occurring as an isolated temporal bone anomaly. The anomaly was bilateral in 10 cases and unilateral in 2 cases. Within this group of individuals, eight ears with normal hearing or varying degrees of SNHL (mild and moderate hearing losses) were followed longitudinally and a progression in hearing loss was noted in six children. A sudden shift in hearing accounted for 5 of the 12 children with hearing loss. Three of the changes in auditory function were documented after relatively minor head trauma. The fourth loss occurred while forcefully playing a wind instrument, and the fifth loss occurred immediately after an airplane flight.

A high prevalence of LVAS was reported by Okumura et al (1995), who reviewed 181 patients with SNHL of unknown etiology and found 13 patients (7%) who had LVAs. High-frequency hearing was noted to be involved more commonly than low-frequency hearing. Additionally, half of these individuals presented with a history of sudden hearing loss. Most of the patients experienced minor head trauma, a common cold, or exercise before a decrease in hearing was noticed. Because LVAS can occur without a cochlear anomaly present, we suggest that...
hearing loss associated with LVAS could be related to or caused by the LVA and not a cochlear anomaly.

In a recent retrospective review of pediatric patients examined in our practice between January 1, 1989 and December 31, 1997, we found 146 children with an ICD-9 diagnosis code of mixed hearing loss, SNHL, sensorineural-combined-type hearing loss, or total hearing loss. Of the 146 hearing-impaired children, 51 of these children received a CT scan and the results are as follows: 37 patients were normal, 7 had LVAS, 3 CT scans were lost and could not be reviewed, 2 patients had Mondini's dysplasia, 1 had a middle ear anomaly, and 1 had a short cochlea. In addition, a CT scan was ordered on one patient; however, the patient deferred this evaluation. Of those patients in whom CT scanning was obtained, 14 percent were noted to meet the criteria of Wilson et al (1997) for LVAS. Of the total number of charts reviewed (146), 5 percent were found to have LVAS.

Our overall prevalence of 5 percent of LVAS in hearing-impaired children is very similar to the 7 percent reported by Okumura et al (1995). LVAS is now a recognized disorder that may result in sudden, fluctuating, or progressive SNHL. However, this disorder has received little recognition in the audiology literature. Because of the lack of recognition that LVAS has received in the past, as well as the varying degrees in which LVAS presents itself audiologically, we present the following cases demonstrating some of the children seen in our practice with LVAS.

CASE HISTORIES

Case 1

Case 1 presented as a 7-year-old child with prior history of fluctuating asymmetric hearing loss. Audiologic evaluation in our office revealed a moderate to profound mixed hearing loss that was worse in the right ear (Fig. 1). Due to inconsistent responses and asymmetry in hearing, an auditory brainstem response (ABR) evaluation was performed with wave V identified at 60 dB nHL in the right ear and 40 dB nHL in the left ear. Blood work for an immune-related hearing loss was negative except for a speckled pattern on ANA testing for lupus erythematosus. A high-resolution temporal bone CT scan demonstrated LVAs, relatively large internal auditory canals, and no cochlear anomaly bilaterally (Fig. 2). Avoidance of sports activities involving potential head trauma, hearing aid evaluation, and audiologic monitoring at 3-month intervals during the first year after diagnosis and then every 6 months was recommended. This patient has been followed for the past 7½ years with no significant decrease in hearing.

Case 2

Case 2 presented as a 6-year-old child with a hearing loss in the left ear. The hearing loss was first identified during a school hearing test after the child complained of difficulty hearing from the left ear. Prior to the decrease in hearing, it was reported that the left side of the child's head and ear had been struck on a goal post while playing soccer. A magnetic resonance imaging (MRI) scan, ordered by the child's pediatrician, revealed no cerebellopontine angle or internal auditory canal mass. An audiologic evaluation in our office revealed normal hearing in the right ear and a
severe to profound mixed hearing loss in the left ear (Fig. 3). Preferential classroom seating, no amplification, and a follow-up in 1 year were recommended. Three years later, after mild trauma to the head while on the school playground, the child reported an immediate hearing loss in the right ear, tinnitus, and dizziness. An audiogram performed that same day by another facility revealed a severe to profound hearing loss in the right ear and a profound hearing loss in the left ear (Fig. 4). A CT scan of the temporal bone demonstrated LVAs bilaterally with no cochlear anomaly. Over the next month, hearing fluctuated in the right ear ranging from moderate to severe. The child was treated with amplification in the right ear and referred back to our office for a cochlear implant evaluation. At that time, chief complaints included decreased benefit from amplification and severe tinnitus. An audiolologic evaluation in our office demonstrated a severe to profound mixed hearing loss in the right ear and a profound mixed hearing loss in the left ear (Fig. 5). Due to the mixed nature of the hearing loss, the patient was not a candidate for a cochlear implant. The cochlear implant team did, however, recommend avoidance of all contact sports, continued use of amplification, use of an FM system at school, and an audiolologic evaluation at 3-month intervals in order to closely monitor any further progression in hearing loss.

Case 3

Case 3 presented as a 2-year-old child with a history of recurrent acute otitis media. Sound-field testing revealed a mild hearing loss in the better hearing ear. After bilateral ventilation tube insertion, sound-field testing was normal. Eighteen months later, parental concern regarding possible hearing loss and difficulty testing in the sound field led to diagnostic ABR testing. This study revealed a threshold of 60 dB nHL in the right ear and 20 dB nHL in the left ear. Hearing amplification was suggested in the right ear due to a significant speech and language delay. Speech and language therapy was initiated. Several months later, audiologic testing revealed a mild to moderate conductive hearing loss (PTA of 58 dB HL) with an air–bone gap (ABG) of 30 to 50 dB in the right ear (Fig. 6). The left ear remained within normal limits. Six months later, a significant shift was noted in air and bone conduction in the right ear (Fig. 7). A CT scan demonstrated a large right vestibular aqueduct and no cochlear anomaly in either ear (Fig. 8). There was no documented incident of head trauma. Final recommendations for this
Case 4

Case 4 presented as an 18-month-old child with a history of acute otitis media and parental concern regarding hearing. Soundfield testing revealed no response at the limits of the equipment. ABR testing 2 days later revealed a bilateral profound hearing loss and no response at the limits of the equipment (85 dB nHL). At that time, it was recommended that the patient be fit with binaural amplification, receive immediate speech therapy, and have an audiologic evaluation every 3 months during the first year after diagnosis and then every 6 months thereafter. At age 4, behavioral audiometry under headphones revealed a severe to profound bilateral hearing loss. Six months later, a significant shift in hearing was noted in the left ear. A CT scan demonstrated no cochlear anomaly and LVAs bilaterally. At that time, audiologic testing revealed a profound mixed hearing loss in the right ear and a severe to profound mixed hearing loss in the left ear (Fig. 9). Recommendations were close audiologic monitoring, continued use of amplification and FM system, and consideration for cochlear implantation.

Case 5

Case 5 presented as a 10-year-old child with a history of left-sided progressive hearing loss for 2 to 3 years and a history of otitis media. Audiologic evaluation in our office revealed normal hearing in the right ear and a mild to profound mixed hearing loss in the left ear with an ABG of 20 to 25 dB. At that time, a CT scan demonstrated a large left vestibular aqueduct and normal cochlear development. There was no documented incident of head trauma when the hearing loss was first noted. Recommendations included preferential classroom seating, audiologic
Figure 10  Audiogram demonstrating hearing loss in case 5.

evaluation every 6 months, and avoidance of any contact sport that has the potential for head trauma. When tested 6 years later, hearing in the right ear was within normal limits and testing in the left ear revealed a moderate to profound mixed hearing loss (ABG of 20–35 dB) (Fig. 10).

**PATHOPHYSIOLOGY**

Early in the development of the inner ear, the vestibular duct is short, straight, and proportionally broader than at maturity. During fetal development, the duct narrows, elongates, and assumes the typical J-shaped form of that found in adult life. The vestibular aqueduct and sac do not reach full maturity until 4 years of age. Insult during the early stage of development is thought to result in the vestibular duct remaining in its fetal (large) form (Jackler and De La Cruz, 1989).

While the mechanism of hearing loss associated with LVAS is unclear, some have speculated that it could be the enlarged vestibular aqueduct, cochlear deformity, mixing of intracochlear fluids, or pressure from cerebrospinal fluid (CSF) rushing past the endolymphatic sac that causes the hearing loss. Jackler and De La Cruz (1989) speculated that hearing loss associated with LVAS may be caused after relatively minor head trauma due to a rupture in an intracochlear membrane with resulting mixing of endolymph and perilymph. Abnormal transmission of CSF pressure to the inner ear via a LVA is speculated to cause a defect or tear in an area of congenital weakness in the basilar membrane or Reissner’s membrane. It was once thought that endolymphatic hydrops played a role in LVAS. However, as pointed out by Tomndorf (1975), endolymphatic hydrops causes damage to the apical region of the cochlea resulting in a low-frequency SNHL as opposed to a high-frequency SNHL seen in LVAS.

**IMAGING OF THE VESTIBULAR AQUEDUCT**

Various forms of imaging may be used to detect an enlarged vestibular aqueduct. A high-resolution CT scan of the temporal bone offers high spatial resolution and bony contrast, which facilitates the identification of inner ear anomalies. An alternative to the traditional use of CT scanning is use of high-resolution MRI imaging with thin-section fast spin echo (FSE) pulse sequence. Harnsberger et al (1995) found that visualization of soft tissues and endolymph and perilymph in the labyrinth and the endolymphatic duct and sac is superior with this method when compared to traditional CT imaging. Unfortunately, FSE MRI machines are not readily available. Several methods of measuring the vestibular aqueduct using CT scans have been employed to determine enlargement of the vestibular aqueduct. Reussner et al (1995) suggested measurement of the aqueduct in its midstation, halfway between the common crus and the endolymphatic duct and sac. A diameter greater than 1.5 mm is considered to be enlarged. Wilson et al (1997) determined the diameter of the vestibular aqueduct by measuring midway between the common crus and the external aperture. They considered the vestibular aqueduct to be enlarged when the diameter was greater than twice the diameter of the adjacent posterior semicircular canal. Okumura et al (1995) considered the vestibular aqueduct to be enlarged when the aperture was greater than 4 mm and the distance between vestibule and traceable part of the vestibular aqueduct is shorter than 1 mm.

**AUDIOLOGIC FINDINGS**

Harke and Eiten (1995) listed several characteristics in a child’s case history and audiologic findings that may indicate LVAS. These include late identification of hearing loss, speech and language skills better than expected based on severity of loss, documented audiologic findings of progressive or fluctuating hearing loss, sudden drops in hearing following minor head trauma, especially in young toddlers, and a conductive component with normal tympanometry. Fluctuating hearing loss and a conductive component often accompany LVAS and are important audiologic findings when diagnosing this syndrome. The conductive component may represent stapes fixation, which has been noted in at least one case (Shirazi, 1994). The
mechanism for fluctuation in auditory function remains obscure at this time. Although a conductive component may occur in some individuals with LVAS, a SNHL is the most predominant audiologic finding. Often, a sudden decrease in sensorineural auditory function is associated with a specific trauma.

A prospective study of vestibular aqueduct enlargement and the progression and degree of hearing impairment has not been performed. When a LVA is identified on an imaging study in a child or an adult with normal hearing, the only prognosis that can be drawn from this is that a hearing loss of some degree may be expected in the future, especially after head trauma. In a retrospective review, Jackler and De La Cruz (1989) identified 17 individuals with a LVA as the only labyrinthine anomaly. Of 33 abnormal ears, 31 ears had associated hearing loss. Eight of the abnormal ears (27%) displayed a mixed hearing loss with the most predominant component being sensorineural. Over time, a progression in hearing loss was found to occur in 65 percent (15/23) of the ears. Complaints of dizziness were present in 29 percent of the cases. They suggest that vestibular aqueduct enlargement has a reasonably frequent association with SNHL in children. Average hearing levels in ears with LVAS were better when a cochlear anomaly was present than not.

**TREATMENT**

Although some authors have advocated endolymphatic sac surgery for fluctuating or progressive-SNHL-associated congenital malformations of the inner ear, it is now well recognized that shunting the endolymphatic sac does not halt progression of hearing loss and indeed is associated with a significant likelihood of profound deafness. Jackler and De La Cruz (1989) found an immediate decrease in hearing in four of seven ears after endolymphatic shunt surgery.

Hirsch et al (1992) reported on a patient with deafness attributed to a viral infection at 3 months of age. CT imaging demonstrated an enlarged vestibular aqueduct. It was thought that an endolymphatic sac shunt would be helpful in dissipating her tinnitus and dizziness. A right-sided endolymphatic sac exploration was performed and revealed an enlarged cavity that continued to reaccumulate CSF. After continued attempts at suctioning out the cavity, the sac was sutured closed and the area packed with fat to obliterate the endolymphatic sac. Two years postoperatively, the patient was reported as being asymptomatic for tinnitus and dizziness.

Wilson et al (1997) have recently published on a series of patients with LVAS who underwent obliteration of the endolymphatic sac in an attempt to stabilize auditory function. All six of their patients had progressive SNHL preoperatively. Progression in hearing loss was often noticed after minor head trauma. Hearing remained stable in four of the seven ears and improved in two ears, and the hearing loss continued to progress in one ear. While this study may be flawed in design, the concept of interrupting the transmission of increased intracranial pressure into the cochlea is novel and will certainly remain an area of continued investigation.

**CONCLUSION**

Although the term LVAS was coined approximately 2 decades ago, it is apparent that its application and diagnosis in a clinical setting has been latent. An 8-year retrospective review of hearing-impaired children in our practice revealed a 5 percent incidence of LVAS. Children with LVAS may present with different clinical courses. Because of the various manners of presentation, and the often confusing presence of a conductive component, it is essential that audiologists and otologists work together in diagnosing LVAS. A crucial recommendation, especially in bilateral cases of LVAS, is avoidance of all contact sports that may involve potential head trauma or extreme changes in barometric pressure such as scuba diving. Restriction from these activities will not necessarily prevent a progression in hearing loss; however, it may decrease its likelihood. We believe that the use of CT scanning in cases with a fluctuating sensorineural or mixed hearing loss of unknown etiology will increase the identification of LVAS in children.

**Acknowledgment.** This work was supported in part by funds from Presbyterian Ear Institute, Albuquerque, NM.

**REFERENCES**


