

# Large Vestibular Aqueduct Syndrome: A Tutorial and Three Case Studies

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## Abstract

The large vestibular aqueduct (LVA) syndrome is a congenital malformation that predisposes the patient ultimately to a loss of hearing and possible continuing vestibular disorder. If the LVA patient is diagnosed, it typically is not until later life, when he/she exhibits profound sensorineural hearing loss. To better understand this disorder and to bring it to the attention of audiologists, we provide a brief tutorial of LVA and present three case studies that illustrate the syndrome.

**Key Words:** Congenital malformation, large vestibular aqueduct (LVA), sensorineural hearing loss

**T**he occurrence of a large vestibular aqueduct (LVA) is a relatively rare event that affects both cochlear and vestibular systems. Because the syndrome lacks unique characteristics, it may be undetected or not diagnosed until adulthood. As a group, audiologists seem to be minimally aware of LVA, which is unfortunate, because they can be key players in diagnosis and follow-up care. To alert our fellow audiologists to the occurrence of LVA, we provide a brief tutorial and present three case studies.

## HISTORY

**L**V A was named in 1978 by Valvassori and Clemis after retrospectively reviewing 3700 patient polytomographic studies. The authors described the characteristics of LVA in the 50 patients identified with the syndrome. In a subsequent article that provided additional insight, Emmett (1985) found 26 LVA cases after reviewing 2683 patient computed tomography (CT) or polytomographic studies. The data from both studies were from highly selected populations, that is, patients who had presented with auditory or vestibular complaints and had CT or polytomography completed, which unfortunately precludes general estimates of prevalence.

## KEY CHARACTERISTICS

**T**he LVA syndrome is a congenital malformation of the temporal bone that predisposes the affected person to early onset hearing loss and vestibular disorders (Arcand et al, 1991). While the criteria for determining LVA is somewhat vague, a vestibular aqueduct diameter larger than 1.5 mm (Valvassori and Clemis, 1978) to 2.0 mm (Swartz et al, 1985) is generally considered to be the defining characteristic. Figure 1 illustrates the dramatic difference between a normal vestibular aqueduct and LVA, as seen by axial CT scan.

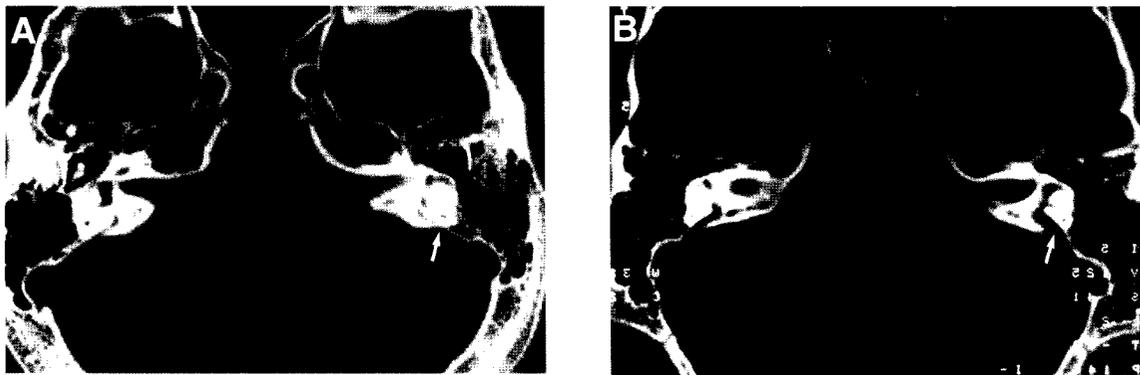
The pathophysiology of hearing loss associated with LVA is not fully understood. There are several theories that suggest various mechanisms such as concomitant cochlear malformations. Lacombe et al (1989) suggest that LVA occurs during the seventh week of embryogenesis and represents a variant of Mondini dysplasia. Levenson et al (1989) disagree with this notion since hearing loss associated with LVA usually occurs well after birth.

Another theory suggests that LVA hearing loss results from cerebrospinal fluid (CSF) pressure fluctuations transmitted by the endolymphatic duct to the scala media, or through the lateral wall of the internal auditory canal or cochlear aqueduct (Jackler and De La Cruz, 1989; Hirsch et al, 1992).

Another possible cause of LVA hearing loss has been suggested by Belenky et al (1993).

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**Figure 1** High resolution, axial CT scans of the temporal bones. A, normal vestibular aqueducts (arrow); B, large vestibular aqueducts of patient AG.

They report a high incidence of round window abnormalities that may predispose the LVA patient to perilymph fistulas, which could be the cause of sensorineural hearing loss.

The degree of hearing loss reported with LVA is variable. Typically, the patient progresses to a profound sensorineural loss bilaterally. The course associated with the progression of hearing loss is quite variable. Some patients suffer a total loss of hearing within the first few months of life, while others do not experience decrements in hearing until later in childhood (Arcand et al, 1991). The audiometric configuration of LVA is not distinct, but Jackler and De La Cruz (1989) report that hearing does not fluctuate, but rather drops in a stepwise fashion. Furthermore, events like minor head trauma have been reported in the literature as possible triggers for decrements in hearing (Jackler and De La Cruz, 1989; Hirsch et al, 1992).

It is not uncommon for LVA to be misdiagnosed initially, or perhaps remain undiagnosed throughout the life of the patient. The typical scenario for LVA is early childhood sensorineural hearing loss that usually is attributed to a viral insult or some unknown etiology. Later in life, the diagnosis of LVA is made when hearing deteriorates further, or when help is sought for vestibular problems.

Although LVA is relatively rare, there is general consensus on the actual occurrence of the disorder. This agreement is in contrast to the literature on the occurrence of the enlarged cochlear aqueduct syndrome, which has been controversial (Jackler and Hwang, 1993).

The prevalence of LVA in the general population is not known. In the two previously cited retrospective studies of highly selected populations (Valvassori and Clemis, 1978; Emmett, 1985), the incidence was 1 percent in

the population studied. While LVA appears to be somewhat rare, it does occur more frequently than the Mondini dysplasia. Jackler and De La Cruz (1989) reported that LVA is common in children with sensorineural hearing loss, but suggests that it is underdiagnosed.

LVA has historically been considered a bilateral disorder. Recently, however, Belenky et al (1993) documented two cases of unilateral LVA. In a population of eight children with LVA, these authors identified two children with unilateral occurrence.

Treatments for LVA remain palliative and controversial. Efforts to stabilize hearing through endolymphatic sac surgery have resulted in further decrements in hearing or have been subject to complications (Hirsch et al, 1992; Belenky et al, 1993).

Vestibular findings may or may not be present with LVA. In some cases, vestibular symptoms may persist even after the hearing loss has progressed to the profound category (Hirsch et al, 1992). In the literature, while vestibular findings are frequently reported, data detailing specific vestibular test results are scarce (Schesel and Nedzelski, 1992).

## CASE STUDIES

The problem of correctly identifying LVA is complex, as are the issues related to treatment. When identified, the typical LVA patient has a profound sensorineural hearing loss, which allows audiometric data to provide minimal insight into diagnosis. We present three patients with LVA who have sufficient residual hearing to enable evaluation with not only basic audiometric tests, but also auditory brainstem response (ABR), electrocochleography (ECoG), electronystagmyography (ENG), and vestibular

ocular reflex (VOR) tests. We detail case history and audiometric and vestibular data on each patient to provide insight into this unique disorder.

**Patient MV**

A 23-year-old female was seen for routine follow-up for unilateral hearing loss (Fig. 2). The patient reported right-sided profound hearing loss occurring early in childhood of unknown etiology. The patient denied tinnitus, except on one occasion at age 13, which resolved spontaneously. She also reported no dizziness or balance problems. A CT scan was ordered at the time she presented to our clinic and LVA was diagnosed. ABR and ECoG testing of the good ear produced normal data. ENG data were normal, except for caloric responses. Closed loop irrigation of the right ear produced no nystagmus with warm or cool water, while stimulation of the left ear produced weak responses. VOR testing produced normal gain, phase and symmetry data. In view of the hypofunctional right labyrinth, the VOR data suggested that this patient was exhibiting central compensation.

This healthy, 23-year-old female has been informed of her diagnosis of LVA. Counseling regarding restrictions on activities (no skydiving, bungee jumping, SCUBA diving, etc.) and periodic exams will hopefully prolong useful hearing in the left ear.

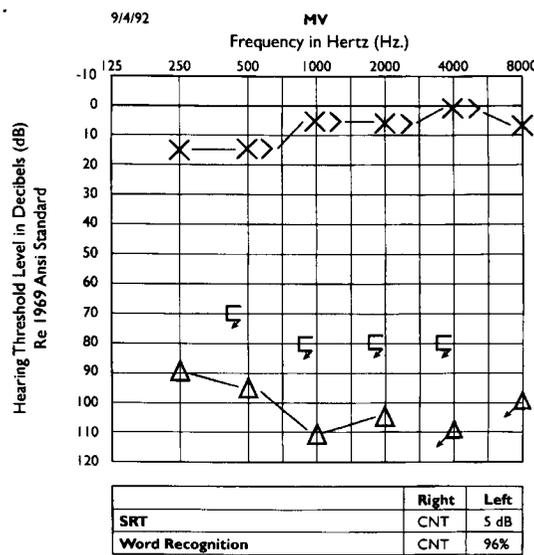


Figure 2 Audiometric data of patient MV.

**Patient DM**

This 31-year-old female was seen upon referral for problems related to recent auditory and vestibular changes. The patient reported that bilateral hearing loss was first discovered at 5 or 6 years of age. By age 10, she exhibited a severe to profound sensorineural hearing loss (Fig. 3) bilaterally with the left ear being somewhat better than the right. Her hearing remained stable for almost 20 years, when suddenly her hearing and balance began to deteriorate. Over a 1-year period, pure-tone and speech thresholds in her left ear dropped substantially. Right ear pure-tone thresholds remained relatively stable, while speech reception thresholds dropped approximately 20 dB (Fig. 4). During the 18 months prior to her coming to our clinic, she reported marked dizziness and vertigo. At the time of her initial evaluation at this clinic, she exhibited true rotatory vertigo, which occurred on a daily basis. This patient had been diagnosed with various disorders over the previous 18 months, that is, autoimmune disorder, Meniere's disease, delayed endolymphatic hydrops, congenital sensorineural hearing loss, and psychiatric problems. High-dose prednisone had been prescribed at the time of the sudden loss and had been maintained for over 14 months, resulting in excessive weight gain and other side effects. A CT scan obtained during her evaluation at our clinic revealed the presence of LVA. The steroids were tapered and discontinued with no additional

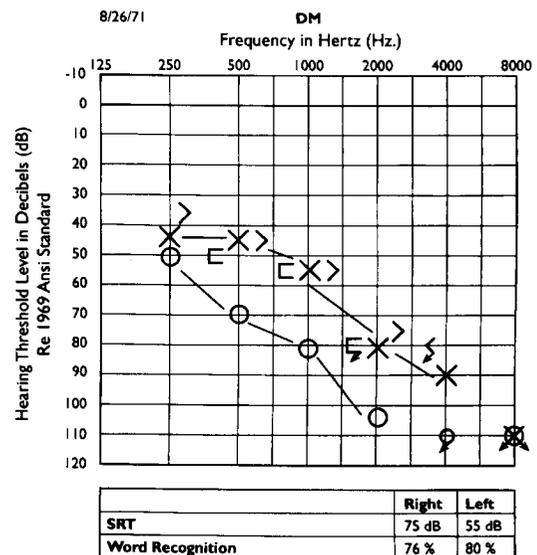


Figure 3 Audiometric data of DM at age 10.

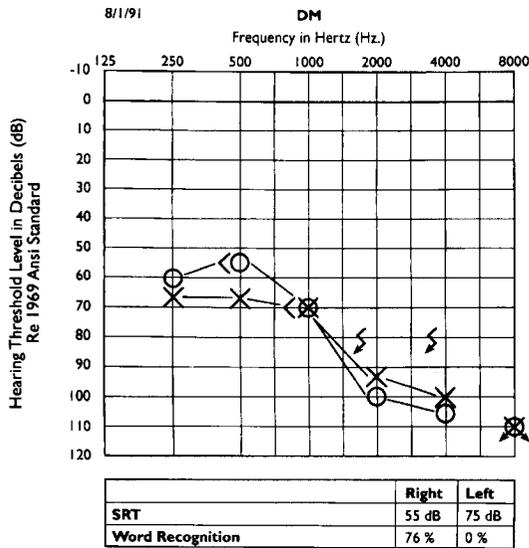


Figure 4 Audiometric data of DM following onset of symptoms.

otologic consequences, and resolution of the side effects. Balance problems, however, persisted.

The patient currently has lost all hearing in what was her better ear (Fig. 5). She currently communicates via auditory input from her right ear and lipreading. The loss of residual hearing in the left ear in concert with her continuing vestibular problems has been most difficult. It has changed her lifestyle dramatically, with prognosis of improvement being very poor.

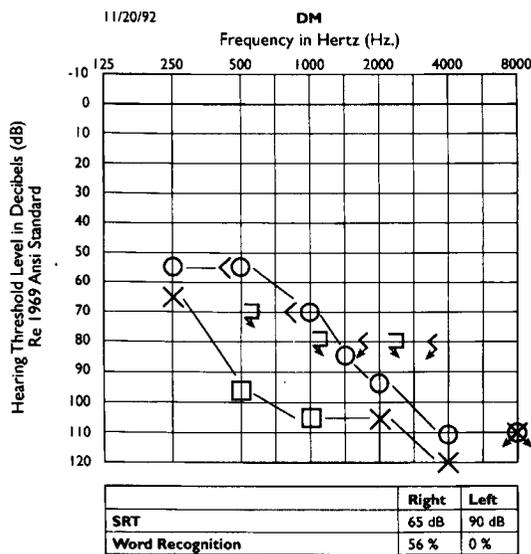


Figure 5 Present audiometric data of DM.

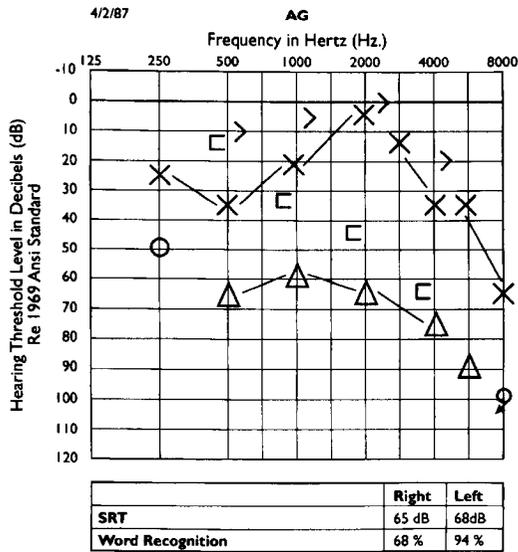


Figure 6 Audiometric data of AG at age six.

### Patient AG

A 16-year-old male was evaluated for sudden hearing loss of the left ear with no imbalance or vertigo. Two days prior to the loss, he was involved in an altercation, resulting in trauma to the head with no loss of consciousness, otologic complaint, or significant injury. A pretraumatic hearing loss documented at age 11 was attributed to "congenital stapes fixation superimposed on viral cochleitis" (Fig. 6). A hearing test performed at the time AG was seen in our office revealed a severe hearing loss in the right ear and a profound loss in the left ear (Fig. 7). The patient was admitted to the hospital and placed on bed rest and 80 mg of prednisone. All investigations were negative except a CT scan, which demonstrated bilateral LVA. After 8 days of medical management, there was no improvement. An exploratory tympanotomy failed to demonstrate an active perilymphatic fistula, but revealed minor middle ear anomalies, that is, incudo-stapedial joint and stapes footplate irregularities, and the oval window niche was unusually shallow with a groove extending toward the fissula ante fenestrum. Although no active fistula was seen, as a precautionary measure, the round and oval window niches were packed with small free grafts of temporalis muscle to seal the most common areas of perilymphatic fistula. Low-frequency hearing in the left ear eventually did improve to the point where it approximated the pretraumatic level by day 30 (Fig. 8). At that point, the patient was

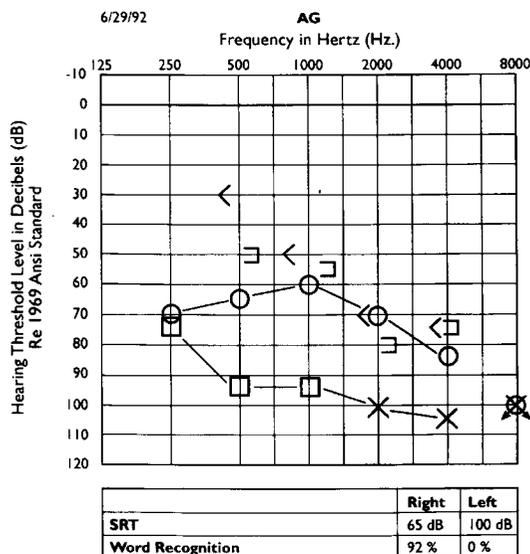


Figure 7 Audiometric data of AG following minor head trauma.

counseled to return urgently for any audiometric or otologic changes.

Approximately 4 weeks later, AG returned with a sudden profound hearing loss in the right ear. On this occasion, the hearing loss was not associated with any physical trauma or event. Bedrest and systemic steroid therapy resulted in a recovery of hearing that approximated to the previous level noted in Figure 8. A full complement of audiometric and vestibular tests (ABR, ECoG, ENG, VOR, etc.) produced normal data.

The initial decrease in hearing was associated with minor head trauma. The LVA literature

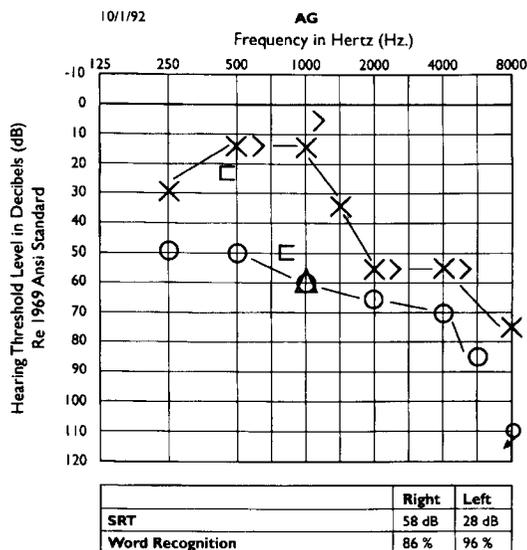


Figure 8 Present audiometric data of AG.

documents similar cases where hearing also dropped as a result of head trauma. In this case, intervention, for the short term, may have resulted in recovery. It may be possible with the diagnosis of LVA that aggressive and urgent intervention when hearing begins to drop may prolong or perhaps prevent what has been the inevitable profound hearing loss. At age 19, this patient has reported only one minor episode of hearing fluctuation over a 3-year period. He has not experienced any head traumas and his hearing has remained stable.

### SUMMARY AND THOUGHTS ON LVA

The three cases of LVA that we reviewed represent different case histories and audiometric/vestibular configurations. After looking at their individual results, we could not characterize LVA based solely on the data. It would be fortunate if there were audiometric/vestibular findings suggesting that the CT be performed for confirmation. We, however, found no specific findings common to all three patients that were unique to LVA.

As a point of curiosity, we performed ECoG with MV and AG sitting and in a supine position with their feet elevated. It was postulated that if, in fact, hearing loss occurs due to CSF pressure fluctuations to the cochlea, slight changes that occur in CSF pressure with postural changes might be reflected in the ECoG summating potential/action potential (SP/AP) ratios. Unfortunately, we did not see differences between sitting and supine SP/AP ratios. While postural ECoG is not a standard test at present, Campbell and Abbas (1993) have reported different SP/AP ratios with changes in posture for fistula patients.

While it appears from the literature that LVA is rare, actual prevalence may be higher than expected because of frequent nondiagnosis. As a result, there needs to be vigilance for patients who may have LVA. Audiologists should be aware that children or adolescents who present with sensorineural hearing loss of unknown etiology, with or without vestibular symptoms, may have the syndrome. These patients should be evaluated for possible LVA. If patients do not get a CT, they should be monitored audiometrically on a regular basis for further decrements, which, if present, suggest LVA.

The value of early diagnosis of LVA is couched in the assumption that awareness on the part of the parents, child, physician, audiologist, etc. may change or delay the inevitable profound

hearing loss. Theoretically, changes in patient behavior at an early age, which reduce head traumas or repeated bumps to the head, could delay the onset of the hearing loss. This proactive approach could be beneficial for young children who are still in the stages of language development. If profound hearing loss can be delayed for these children so that they can develop language, the impact on their education and lifetime of communication might be dramatic.

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