

# USE OF WIDEBAND ABSORBANCE MEASUREMENT TO ASSESS LARGE VESTIBULAR AQUEDUCT SYNDROME: A CASE STUDY

## Contributions:

A Study design/planning  
B Data collection/entry  
C Data analysis/statistics  
D Data interpretation  
E Preparation of manuscript  
F Literature analysis/search  
G Funds collection

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## Abstract

There is a limited number of diagnostic procedures to verify the presence of large vestibular aqueduct syndrome (LVAS) in humans. The wideband absorbance (WBA) measurement technique is a non-invasive and objective tool for assessing the amount of sound energy absorbed by the tympanic membrane over a wide frequency range. We suggest that WBA could be an aid for detecting LVAS in some hearing-impaired subjects. We report use of WBA on a patient who had bilateral sensorineural hearing loss associated with LVAS and cochlear abnormalities in both ears.

**Key words:** sensorineural hearing loss • large vestibular aqueduct syndrome • sudden hearing loss

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## EL USO DE LA MEDICIÓN DE LA ABSORBANCIA EN EL DIAGNÓSTICO DEL ACUEDUCTO VESTIBULAR DILATADO: ESTUDIO DEL CASO

### Resumen

Existe un número limitado de procedimientos diagnósticos que sirven para confirmar o descartar el síndrome del acueducto vestibular dilatado (inglés: *Large Vestibular Aqueduct Syndrome*, LVAS) en humanos. La técnica de medición de la absorbancia (inglés: *wideband absorbance*, WBA) permite de modo no invasivo y objetivo determinar la cantidad de energía acústica absorbida por el tímpano en banda ancha de frecuencias. Según los autores, el método WBA puede resultar útil en la detección de LVAS en algunas personas con hipoacusia. En el presente trabajo se describe el uso del método WBA en un paciente con hipoacusia neurosensorial bilateral en el desarrollo del síndrome acueducto vestibular dilatado (LVAS) y la presencia de anomalías cocleares en ambos oídos.

**Palabras clave:** hipoacusia neurosensorial • acueducto vestibular dilatado • hipoacusia súbita

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## ИСПОЛЬЗОВАНИЕ ТИМПАНОМЕТРИИ В ДИАГНОСТИКЕ ШИРОКОГО ВОДОПРОВОДНОГО ПРЕДДВЕРИЯ: ОПИСАНИЕ СЛУЧАЯ

### Изложение

Существует ограниченное количество диагностических процедур, служащих для подтверждения или исключения синдрома широкого водопроводного преддверия (англ. *Large Vestibular Aqueduct Syndrome*, LVAS) у человека. Техника измерения широкополосной тимпанометрии (англ. *wideband absorbance*, WBA) позволяет неинвазивным и объективным способом определить количество акустической энергии, поглощаемой барабанной перепонкой в широкой полосе частот. По мнению авторов, WBA может оказаться полезным при диагностировании LVAS у некоторых лиц с тугоухостью. В настоящей работе описано использование WBA у пациента с двусторонней нейросенсорной тугоухостью при широком водопроводном преддверии (LVAS) и наличии аномалии улитки в обоих ушах.

**Ключевые слова:** нейросенсорная тугоухость • широкое водопроводное преддверие • внезапная тугоухость

## ZASTOSOWANIE POMIARU ABSORBANCJI W OCENIE POSZERZONEGO WODOCIĄGU PRZEDSIONKA: OPIS PRZYPADKU

### Streszczenie

Istnieje ograniczona liczba procedur diagnostycznych, służących do potwierdzenia bądź wykluczenia poszerzonego wodociągu przedSIONKA (ang. *Large Vestibular Aqueduct Syndrome, LVAS*) u ludzi. Technika pomiaru absorbancji (ang. *wideband absorbance, WBA*) pozwala w sposób nieinwazyjny i obiektywny określić ilość energii akustycznej pochłanianej przez błonę bębenkową w szerokim paśmie częstotliwości. Zdaniem autorów, WBA może okazać się pomocne w wykrywaniu LVAS u niektórych osób z niedosłuchem. W niniejszej pracy opisano wykorzystanie WBA u pacjenta z obustronnym niedosłuchem czuciowo-nerwowym w przebiegu LVAS i obecności anomalii ślimaka w obu uszach.

**Słowa kluczowe:** niedosłuch czuciowo-nerwowy • poszerzenie wodociągu przedSIONKA • nagły niedosłuch

### Introduction

Large vestibular aqueduct syndrome (LVAS) is considered the most common cause of hearing loss in cases of congenital malformation of the temporal bone in humans [1]. Its incidence is probably underestimated because of diagnostic limitations of radiological examination at an early age [1]. Currently, the most appropriate tool for detection and confirmation of this type of inner ear malformation is computerised tomography (CT scan).

The onset of hearing loss in LVAS is nonspecific; however hearing loss from LVAS is often associated with sudden changes in pressure in the middle ear cavity (barotrauma) and head trauma [2]. Among other co-factors, LVAS is characterized by increased endolymphatic pressure in the vestibule of the cochlea while retaining an intact tympanic membrane and aerated middle ear [3]. Some authors report only cases of LVAS in patients with sensorineural hearing loss, while others also present cases with mixed hearing loss [4]. Some authors claim the conductive component of the hearing loss is a pure cochlear conductive loss and that this may be pathognomonic for the disease [5], an opinion in line with recent model and experimental findings [6].

Wideband absorbance (WBA) measurement is a relatively new non-invasive method of assessing the state of the middle ear which has now become clinically available. The principle behind the technique is to measure, over a wide frequency range, the amount of sound energy absorbed by the tympanic membrane [7].

Models of how the hearing organ functions suggest that higher than normal fluid pressure in the cochlea may increase impedance at the oval window and result in decreased mobility of the ossicles [6]; in turn, the increased impedance reduces the amount of energy absorbed by the eardrum. In theory, the physical connection of the eardrum to the inner ear through the ossicular chain and oval window might allow an indirect assessment to be made of inner ear status.

Another reason for interest in WBA is that fluctuations in hearing associated with LVAS, and its usually progressive nature, generate the risk of misdiagnosis and inappropriate methods of treatment.

In some abnormalities of the middle or inner ear – such as ossicular discontinuity, otosclerosis, or superior semicircular canal dehiscence – measurements of absorbance combined with pure-tone audiometry (air–bone gap) have been shown to be clinically valuable in assessing each of these disorders and differentiating between them [8]. We therefore hypothesized that WBA measurement might be a relatively sensitive tool for assessing LVAS as well.

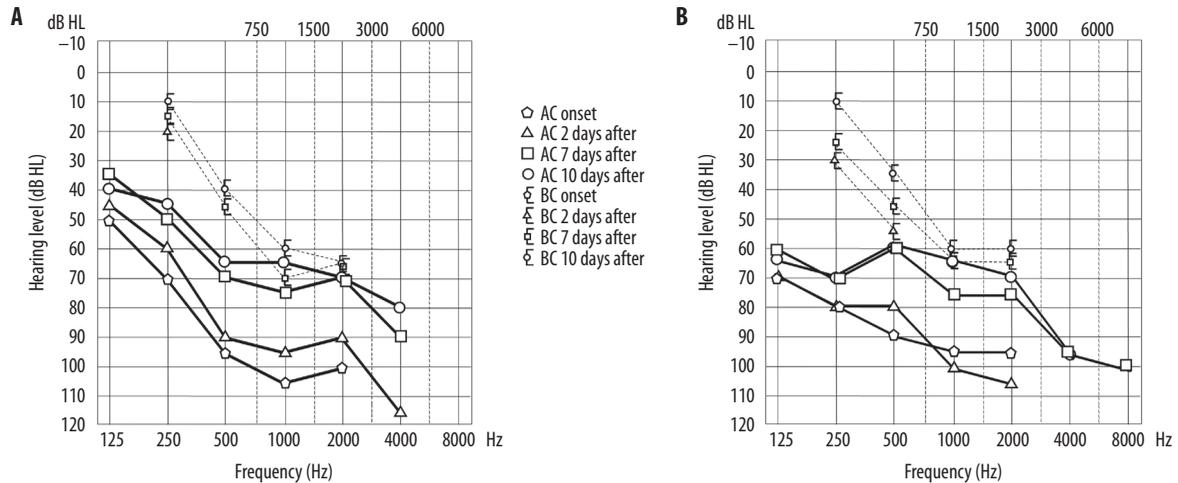
The aim of this case report is to illustrate the WBA characteristics of a patient with confirmed bilateral LVAS and profound hearing loss.

### Case report

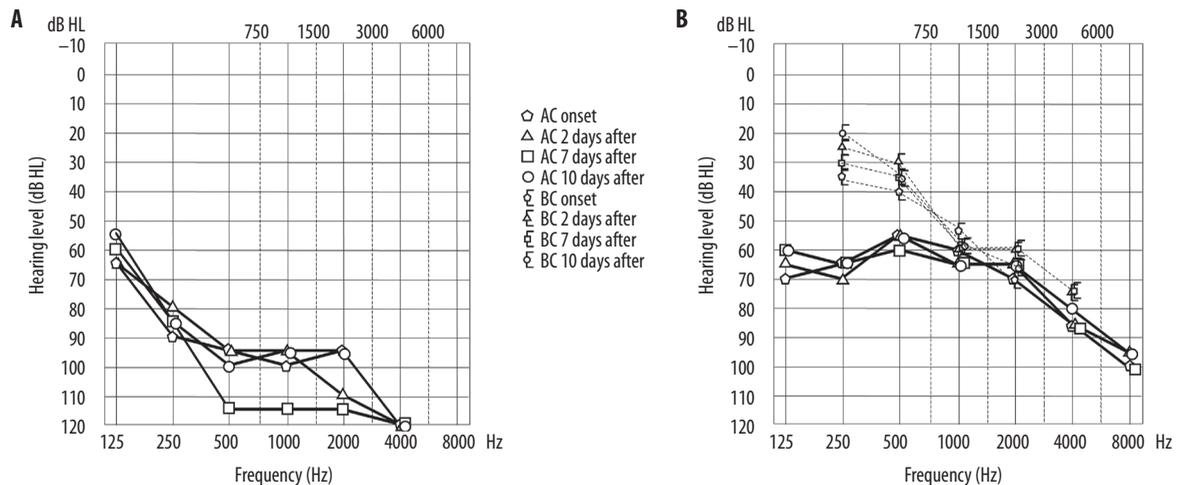
We present the case of a 16-year-old female with LVAS and mixed hearing loss in both ears who came to the clinic because of repeated sudden worsening of hearing. She had no confirmed vestibular dysfunction.

Pure-tone audiometry was performed according to standard procedures in a sound booth. WBA measurements were performed using the Interacoustics Titan device. Each recording session consisted of two consecutive measurements on each ear, taking out and refitting the probe to confirm accuracy. The measurements were subsequently repeated within two days during the course of the patient's stay in the clinic.

The patient had worn behind-the-ear hearing aids bilaterally from early childhood, with satisfactory results. She had experienced four documented episodes of sudden deafness in the past. The first two were probably induced by head traumas which occurred some time before 2014, and no precise details are available. Her third episode of sudden deafness in November 2015 was observed and treated in our clinic. Clinical notes suggest that immediate corticosteroid therapy was helpful in that it was followed by hearing recovery in both ears (Figure 1). Unfortunately, 3 months later sudden deafness in her right ear occurred again. This fourth episode was probably evoked by Valsalva maneuvers and inhalations under increased pressure which were used to try and treat problems with transient dysfunction of the Eustachian tube and an upper respiratory tract infection. This time the same steroid treatment as earlier was of no benefit, and no recovery of hearing was observed after 14 days in hospital (Figure 2).



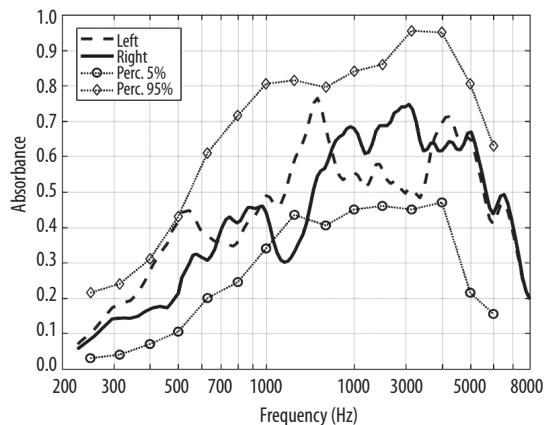
**Figure 1.** Fluctuation of air conduction (AC) and bone conduction (BC) hearing thresholds at the third episode of sudden hearing loss, and for various intervals after corticosteroid treatment was applied. Right ear audiogram (A) and left ear (B). Symbols represent observations at onset and 2, 7, and 10 days after drug treatment



**Figure 2.** Fluctuation of air conduction (AC) and bone conduction (BC) hearing thresholds during corticosteroid treatment following the fourth episode of sudden hearing loss. Audiogram for right ear (A) and left ear (B); symbols as per Figure 1

As a consequence of consecutive steroid therapies, steroid-induced diabetes developed. During the patient’s stay in the clinic, new approaches to measuring changes in her hearing became available. Figure 2 shows the standard audiogram, and Figure 3 shows WBA measurements. The WBA measurements were compared to normative absorbance values obtained by Shahnaz et al. [9]. The mean absorbance levels (ALs) (averaged over 0.6–1 kHz), in comparison with the air–bone gap (ABG, averaged over 0.25–1 kHz), were as follows: AL=−0.51 dB for the right ear (at ABG ≤10 dB); AL=−0.4 dB for the left ear (at ABG ≤10 dB). The results for the left ear were within normative

**Figure 3.** Mean wideband absorbance values in right ear (solid black line) and left ear (dashed grey line) for the presented subject. The dotted and dashed-dotted lines represent respectively the 5<sup>th</sup> and 95<sup>th</sup> percentile of wideband absorbance values for normal hearing subjects according to Shahnaz et al. [9]



data while the absorbance for the right ear exceeded normative data around 1200 Hz.

Magnetic resonance imaging and a CT-scan were performed to confirm the pathologies of the inner ear and to assess the possibility of a cochlear implant in the future. Even though LVAS is characterized by a high rate of spontaneous recovery after sudden deafness, the poor psychological state of the patient and rapidly increasing problems with communication indicate that a CI may be beneficial. If her hearing does not recover in the near future, she will qualify for a CI procedure.

## Discussion

From the characteristics of the audiograms one might come to the conclusion that the patient had a mixed hearing loss in both ears before hearing suddenly worsened. However, there is now strong evidence that the air-bone gap depends not only on the status of the middle ear but also on malfunction of the inner ear [8]. The CT-scan and standard audiometric battery tools confirmed there was no middle ear disease. If one concludes that sudden deafness affected only the cochlea, it is of interest to consider whether the newly available WBA measurements might also be able to show, even indirectly, these inner ear changes.

The WBA measurements were conducted during the fourth episode of sudden deafness. Figure 3 shows that, in comparison to normal results for adults, absorbance values across the frequency range were somewhat abnormal. The values of WBA in the affected right ear showed a large negative peak near 1200 Hz which was below the 5<sup>th</sup> percentile. In the left ear, the WBA was inside the normative range; however, unusual peaks near 500 Hz and 1500 Hz were present. We believe that since the middle ears in this

case were apparently normal, the differences evident on WBA measurement suggest that the indicated anomalies may be due to some abnormality in the cochlea. For example, these values are similar to those obtained for superior semicircular canal dehiscence disorder [8].

The presented case of a patient with LVAS syndrome confirms the complexity of the issues relating to the diagnosis and treatment of this disease. There are still no available diagnostic methods other than CT-scan to unequivocally confirm LVAS syndrome. Therefore, in a number of individuals this disease is detected relatively late. Lack of knowledge by patients and specialists about the cause of the hearing loss may in particular cases lead to sudden progression of hearing loss or even deafness, a loss which might have been avoided or postponed if preventive measures had been taken. For example, this case documents how a Valsalva maneuver can lead to sudden deafness in patients with LVAS, and so the patient should be advised that all exertions which increase intracranial pressure must be avoided. The limited number of ways of treating the disease is still a challenge for modern medicine.

## Conclusions

The WBA measurement technique may play a useful role in diagnosing possible LVAS or other inner ear diseases that can cause abnormal fluid pressure in the cochlea. However, it is worth noting that a full range of characteristic WBA spectra for particular ear diseases are still not available, not even from the manufacturer. Specific features of WBA measurements for various conditions are needed, both individually and for group means. Building such a database is a priority for future research.

## References:

- Berrettini S, Forli F, Bogazzi F, Neri E, Salvatori L, Casani AP et al. Large vestibular aqueduct syndrome: Audiological, radiological, clinical, and genetic features. *Am J Otolaryngol*, 2005; 26(6): 363–71.
- Callison DM, Horn KL. Large vestibular aqueduct syndrome: An overlooked etiology for progressive childhood hearing loss. *J Am Acad Audiol*, 1998; 9: 285–91.
- Valvassori GE, Clemis JD. The large vestibular aqueduct syndrome. *Laryngoscope*, 1978; 88: 723–28.
- Gopen Q, Zhou G, Whittemore K, Kenna M. Enlarged vestibular aqueduct: Review of controversial aspects. *Laryngoscope*, 2011; 121: 1971–78.
- Govaerts PJ, Casselman J, Daemers K, De Ceulaer G, Somers T, Offeciers FE. Audiological findings in large vestibular aqueduct syndrome. *Int J Pediatr Otorhinolaryngol*, 1999; 51(3): 157–64.
- Stenfelt S. Model predictions for bone conduction perception in the human. *Hear Res*, 2016; 340: 135–43.
- Aithal S, Kei J, Driscoll C, Khan A, Swanston A. Wideband absorbance outcomes in newborns: A comparison with high-frequency tympanometry, automated brainstem response, and transient evoked and distortion product otoacoustic emissions. *Ear Hear*, 2015; 36(5): e237–50.
- Nakajima HH, Rosowski JJ, Shahnaz N, Voss SE. Assessment of ear disorders using power reflectance. *Ear Hear*, 2013; 34(Suppl. 1): 48S–53S.
- Shahnaz N, Feeney NM, Schairer KS. Wideband acoustic immittance normative data: Ethnicity, gender, aging, and instrumentation. *Ear Hear*, 2013; 34: 27S–35S.