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Case Report

# Auditory Processing Disorder as the Sole Manifestation of a Cerebellopontine and Internal Auditory Canal Lesion

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**Abstract**

**Background:** Clinical importance of auditory processing disorder (APD) testing is often overlooked and regarded with skepticism given the challenging interpretation of results and the current growing debate of its nature and clinical entity.

**Purpose:** Presentation of this case is highly educational as APD is the single clinical manifestation of a large cerebellopontine and internal auditory canal lesion.

**Research Design:** A case report.

**Data Collection and Analysis:** The patient underwent a standard audiological evaluation with normal results. She was referred for APD evaluation. The APD test battery consisted of speech in babble (SinB), dichotic digits (DD), frequency and duration of pattern sequence testing, Random Gap Detection Test, and gaps in noise. These were followed by otoacoustic emissions testing, auditory brainstem responses (ABR) and magnetic resonance imaging (MRI).

**Results:** Her auditory processing results showed deficits in SinB and DD limited to the right ear as well as deficits in temporal processing. Both verbal and nonverbal tests exhibited deficits strictly limited to the right ear, which was in accordance with what she was experiencing as reduced loudness for the incoming sounds on the right ear. This less costly evaluation revealed that there was good reason to assess electrophysiologically the auditory system. ABR showed an abnormal waveform with either missing or severely prolongated wave V (depending on stimulus polarity). Otoacoustic emissions were normal. MRI was then implemented revealing a large cerebellopontine and internal auditory canal lesion.

**Conclusions:** This clinical case stresses the importance of testing for APD with a psychoacoustical test battery despite current debate of lack of a gold standard diagnostic approach to APD. In this case, APD diagnosis led to a cerebellopontine lesion identification with extension to the right internal auditory canal. This rare cause of APD demonstrates the efficiency of the current diagnostic test battery in revealing lesional causes of central APD.

**Key Words:** auditory-evoked potentials, auditory processing disorder, case report, diagnostic techniques, otoacoustic emissions

**Abbreviations:** ABR = auditory brainstem responses; APD = auditory processing disorder; CANS = central auditory nervous system; DD = dichotic digits; DPOAE = distortion product otoacoustic emissions; DPS = duration pattern sequence; GIN = gaps in noise; MRI = magnetic resonance imaging; PPS = pitch pattern sequence; RGDT = Random Gap Detection Test; SinB = speech in babble; SNR = signal-to-noise ratio; TEOAE = transient otoacoustic emissions

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## CASE REPORT REFERRAL

A 48-yr-old woman was referred for auditory processing evaluation due to hearing difficulties focused on her right ear, experienced during the last three years with no previous issue. She specifically noticed a progressive deterioration in speech perception in her right ear compared to the left ear. A standard audiological evaluation at the beginning of her symptoms composed of pure-tone audiometry, tympanometry, and stapedial reflexes has yielded normal results. Three years later, because of the persistence of her problems, she sought a second hearing evaluation that further included recording of otoacoustic emissions. Both distortion product otoacoustic emissions (DPOAE) and transient otoacoustic emissions (TEOAE) were normal bilaterally. She was further referred by her physician for auditory processing assessment. There was no other complaint made by the patient concerning tinnitus, vertigo, or imbalance.

## DIAGNOSTIC ASSESSMENT

Otoscopy was followed by audiometric testing. All audiometric tests were conducted in a sound-treated booth. Pure-tone air conduction thresholds were obtained using a GSI (Madison, WI) 61 audiometer calibrated per standard guidelines (BSA, 2011b). Tympanograms and stapedius (acoustic) reflex thresholds were obtained using a GSI 33 middle ear analyzer. Otoacoustic emissions and auditory brainstem responses (ABR) were measured through the Intelligent Hearing Systems (Miami, FL) SmartEP platform. Two verbal and four nonverbal psychoacoustic central auditory processing tests were administered using a compact disc player and the GSI 61 audiometer as per the recommendations of the American Speech-Language-Hearing Association (ASHA, 2005) and the British Society of Audiology (Auditory Processing Disorder [APD] Steering Committee; BSA, 2011a). All presentation levels for the APD tests were at 60dB HL, as is the standard practice in our clinic for patients with normal pure-tone audiometry. Normal pure-tone thresholds were defined as thresholds better than 20 dB HL for each octave frequency between 250 and 8000 Hz. Tympanograms were considered normal if middle ear pressure was $>-150$ mm $H_2O$ and compliance was $>0.3$ mL.

Word recognition was assessed using lists of 50 phonetically balanced and frequently occurring words in spoken Greek developed by author V.I. (Iliadou et al, 2006). The list of 50 words was divided into two, with 25 words being presented to the right ear and 25 words to the left ear as a clinical method used to optimize time attributed to the test during clinical testing conditions. The word lists were administered at 60 dB HL. On the basis of the norms collected in author V.I.'s laboratory, 95% correct or better was considered normal performance for adults.

The monaural low-redundancy Greek Speech-in-Babble (SinB) Test (Iliadou et al, 2006; 2009) uses two of the three equivalent lists of 50 words developed by Iliadou et al (2006), one list administered to each ear at 60 dB HL. For each ear, sets of ten words are presented at five different signal-to-noise ratios (SNR; +7, +5, +3, +1, and −1). The participant's task was to repeat the words heard. Performance is measured using the basic Spearman–Karber formula: 50% correct speech identification = $i + 1/2(d) - [(d) \times$ (no. corrects)]/$w$ (where $i$ = initial presentation level in dB [SNR], $d$ = step size, $w$ = number of items per decrement [per step]). Adult normative data collected in author V.I.'s laboratory ranged from −0.6 to 0.2 dB HL.

The Greek DD Test (Tzavaras et al, 1981; Iliadou et al, 2010) includes two practice digit pairs and 40 digit pairs for testing. A pure tone precedes each pair of digits as a cue to capture and sustain the participant's attention. The test is composed of naturally spoken digits from 1 to 9. A different pair of digits is presented simultaneously to each ear at 60 dB HL, and the listener is instructed to repeat all four digits. The number of digits correctly repeated for each ear is converted into a percent correct score.

The Pitch Pattern Sequence (PPS) Test (available from Auditec, St. Louis, MO) presents a three-tone sequence incorporating a high (1430 Hz) and a low (880 Hz) frequency tone, each of 500 msec duration with a 10 msec rise and fall time and each separated by a 10-sec interval. Each sequence is composed of two tones of the same frequency and one tone of a different frequency. Patient is instructed to label the tones (e.g., high–high–low, low–high–low). A total of 30 patterns are presented monaurally to each ear at 60 dB HL following a brief practice session. The percentage of correct patterns was computed for each ear.

The Duration Pattern Sequence (DPS) Test (available from Auditec, St. Louis, MO) presents three consecutive 1000 Hz tones with two different durations. Patient is asked to indicate the duration pattern of the three-tone sequence (e.g., short–long–short, long–long–short). A total of 30 patterns are presented monaurally to each ear at 60 dB HL following a brief practice session. The percentage of correct patterns is calculated for each ear.

The gaps in noise (GIN) involves the monaural presentation of gaps of varying duration interspersed in white noise at 60 dB HL (Musiek et al, 2005; Shinn et al, 2009). The GIN is composed of four lists with 32–36 trials each: list 1 has 35 trials, list 2 has 32, list 3 has 29, and list 4 contains 36 trials. Each trial consists of 6 sec of white noise with a 5-sec intertrial interval. Each gap duration (2, 3, 4, 5, 6, 8, 10, 12, 15, and 20 msec) occurs six times within each list. Patient is told that she is going to hear noise in which there might be short gaps with no noise, that some gaps would be shorter than others, and that in some cases no gaps would be present. She is instructed to indicate detection of a gap by pressing a button. Ten practice items

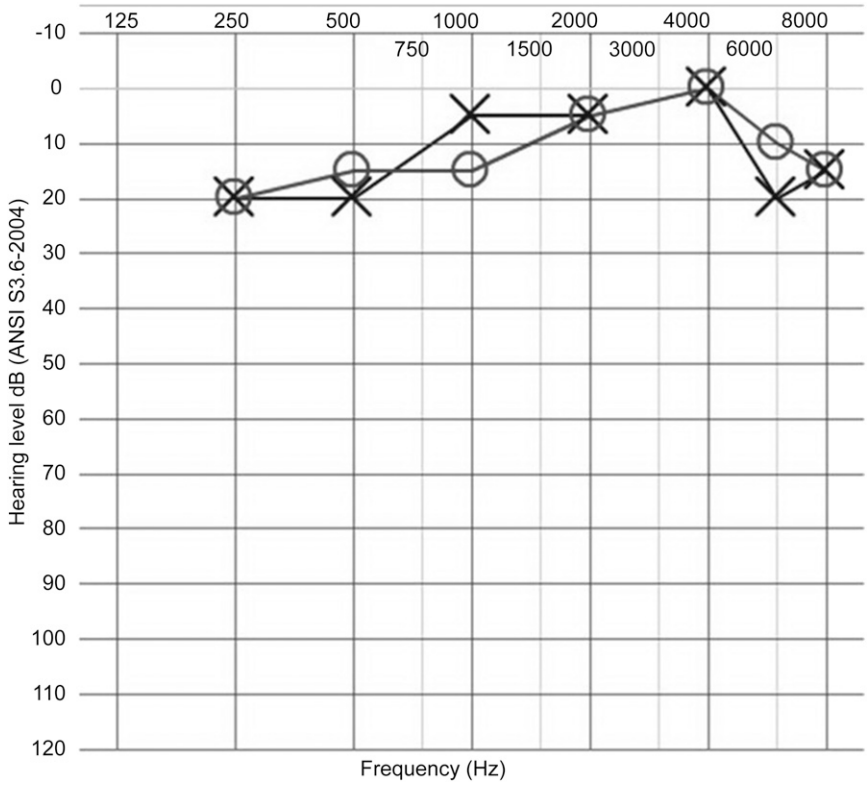


**Figure 1.** Pure-tone audiometry showing normal symmetrical hearing sensitivity. Circles represent right-ear thresholds, and × represents left-ear thresholds.

preceded the test. Gap detection threshold is calculated as the shortest gap duration detected on ≥4 of 6 gaps.

The Random Gap Detection Test (RGDT) (Keith, 2000) involves the binaural presentation of pairs of pure tones separated by silent intervals. Silent intervals for the practice section begin at 0 msec and gradually increase to 40 msec. In the main section of the test, the silent intervals are presented in random order for each of the following pure tones: 500, 1000, 2000, and 4000 Hz, which are tested in sequence. A 4.5-sec intertrial interval is used to allow the participant time to respond. Nine trials are presented in the practice section and nine trials for each of the frequencies tested are presented in the actual test. Each trial for each pure-tone frequency is presented once with a unique silent interval (i.e., gap). A total of 36 trials are used to calculate the overall gap detection threshold. The patient's task is to report whether one or two sounds was heard. The threshold of gap detection is calculated for each frequency as the shortest interval for which the participant reports perception of two tones. Averaging the gap detection threshold for each of the four frequencies tested provides the average gap detection threshold.

Abnormal performance on the central auditory tests was defined as scores more than two standard deviations below the normative mean for adults (AAA, 2010). Diagnosis of APD required failure (i.e., ≥2 standard deviations below the mean) on at least two central auditory tests, including one verbal and one nonverbal measure (AAA, 2010; BSA, 2011a).

The patient underwent further clinical investigation by other related clinicians. Specifically, neurological and otolaryngological evaluations yielded normal results.

## PROTOCOL FOR ABR

Click-evoked ABR were recorded according to the laboratory's standard neuroaudiology protocol for adult patients and with the implementation of the SmartEP platform by Intelligent Hearing Systems. Recording

**Table 1. Results of Central Auditory Processing Tests Showing Abnormal Results in Almost All Tests Administered with the Exception of DPS and a Marginally Abnormal RGDT**

| | Right Ear | Left Ear | Adult Normative Data |
|---|---|---|---|
| Speech audiometry (%) | **92** | 100 | ≥95 |
| SinB (dB HL) | **2** | −0.6 | −0.6 to 0.2 |
| DD (%) | **75** | 95 | >85 |
| PPS (%) | **80** | 90 | >85 |
| RGDT (msec) | 8.3 | | <8 |
| DPS (%) | 100 | 100 | >67 |
| GIN (msec) | **20** | 6 | <8 |

*Notes:* Speech audiometry = suprathreshold speech audiometry in quiet. Abnormal results are shown in bold.

took place in a sound-treated room. The inverting electrode was placed on the mastoid, the noninverting on the vertex, and the ground electrode on the nasion. Interelectrode impedance was <2 kΩ and auditory stimuli were presented through ER-3A insert earphones (Etymotic Research, Elk Grove Village, IL). Clicks were presented monaurally at an intensity of 80 dB nHL and at a rate of 19.1/sec. Two separate trials were recorded and replicated: one with rarefaction and one with condensation click polarity, which were afterward digitally summed. Electrical recordings were filtered through a range of 100–3000 Hz, and 2,048 sweeps were collected from each ear. The examiner monitored the procedure in real time.

## PROTOCOL FOR TEOAE

TEOAE were recorded with the SmartTEOAE platform (Intelligent Hearing Systems) with standard parameters of nonlinear click stimuli of 80 dB SPL presented at a rate of 49.1/sec. Passing criteria consisted of SNR of ≥6 dB for every frequency band and a correlation percentage of ≥90%.

## PROTOCOL FOR DPOAE

DPOAE were recorded with the SmartDPOAE platform (Intelligent Hearing Systems) with standard intensity parameters of f1 = 65 dB SPL and f2 = 55 dB SPL tone pairs, a frequency ratio f1:f2 = 1.22, and five frequencies per octave.

## RESULTS

As a first step before auditory processing assessment, peripheral hearing was re-evaluated with a repeat pure-tone audiogram, which was found to be normal and symmetrical with hearing thresholds of 20 dB HL or better for both ears tested (Figure 1). Results are presented in Table 1. Tympanometry and stapedial reflexes are presented in the Appendix. Suprathreshold speech audiometry at 60 dB HL revealed normal results for the left ear (100% correct responses) and abnormal for the right ear (92% correct responses, 95% cutoff). SinB testing documented a 2-dB SNR for 50% correct word identification for the right ear, which was abnormal, and a normal −0.6 dB

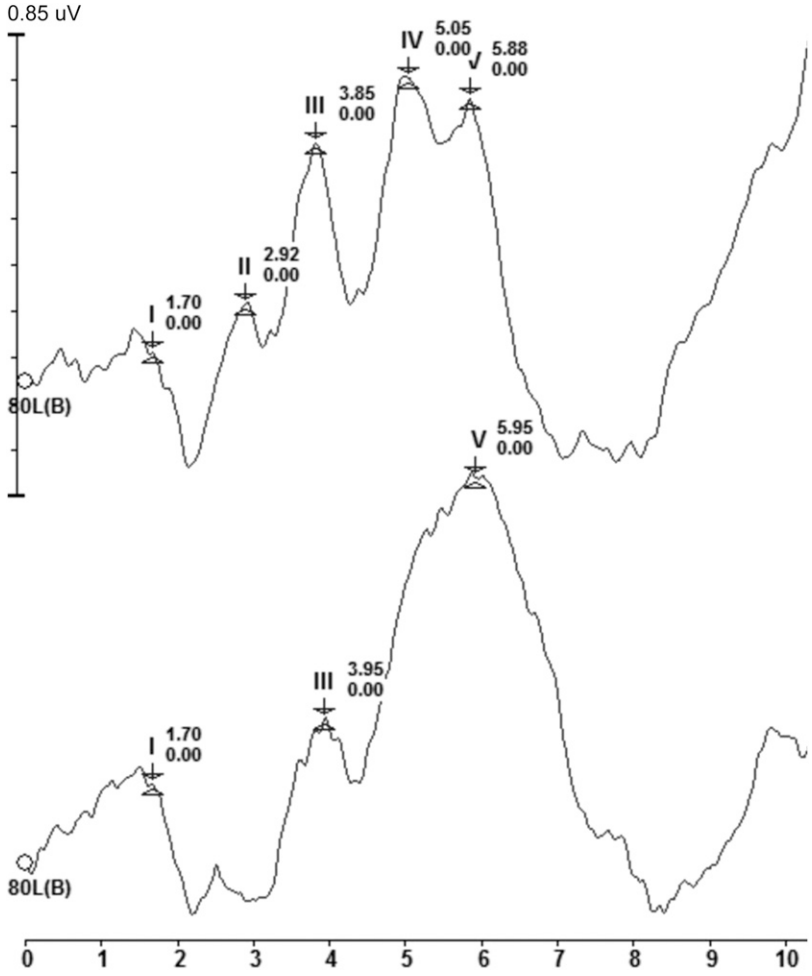


**Figure 2.** ABR left ear result at 80 dB nHL clicks. Stimulus presentation polarity is rarefaction and condensation starting from the top. Clearly defined peaks within the normal absolute latency and interwave latency intervals of our laboratory are visible.

for the left ear (adult normative results: −0.6 to 0.2 dB HL). Dichotic digits (DD) percent correct results were 75% for the right ear and 95% for the left ear (adult normative data are >85% for both ears). PPS test gave 80% correct results for the right ear compared to 90% for the left one (adult normative data are >85% correct results). RGDT revealed an 8.3-msec threshold (adult normative data are <8 msec). DPS Test gave a score of 100% for both ears tested (adult normative data are >67%). GIN Test revealed a threshold of 20 msec for the right ear and 6 msec for the left one (adult normative data are <8 msec).

## ABR Results

Recordings from the left ear (Figures 2 and 3) rendered a clear waveform, with clearly defined peaks and within the normal absolute latency and interwave latency intervals of author N.E.'s laboratory (I = 1.70 msec, III = 3.92 msec, V = 5.90 msec, I–III = 2.22 msec, III–V = 1.98 msec, and I–V = 4.20 msec). However, in the right ear (Figure 4), there was a prolongation of wave III latency (5.33 msec) and therefore of the I–III interwave latency (3.65 msec), while the absolute latency of wave I remained within normal limits (1.68 msec) Most notable was the prolongation of wave V with an absolute latency of 8.50 msec, and with a prolonged I–V interwave latency of 6.82 msec, III–V interwave interval was also prolonged with a value of 3.17 msec. These findings were also noticed at the singular recordings with rarefaction and condensation polarity clicks (Figure 5). Furthermore, there is a significant interaural difference of waves V at 2.6 msec.

The notable prolongations of waves III and V in the right ear recordings and of all interwave intervals as well as the abnormal interaural difference of waves V suggested the possibility of a retrocochlear pathology, and the patient was referred for magnetic resonance imaging (MRI).

## TEOAE Results

Recordings presented as normal (Figure 6) in both ears satisfying all passing criteria (for passing criteria see section Protocol for TEOAE).

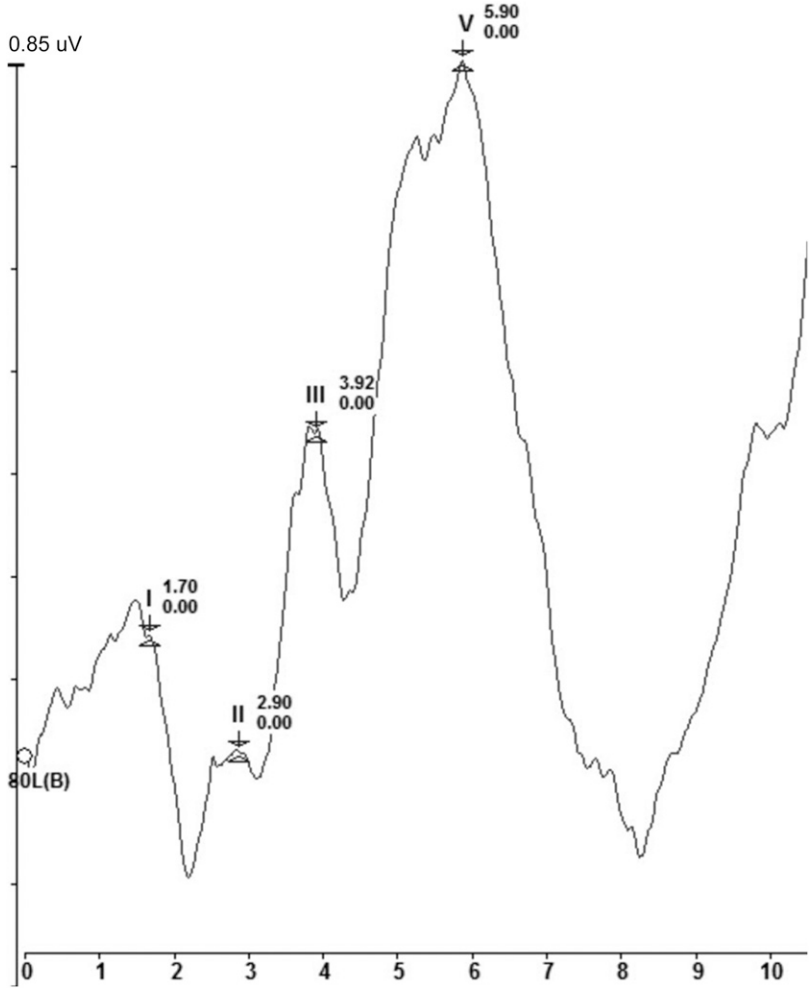


**Figure 3.** ABR left ear result. Digital composite of both click polarities is shown. Clearly defined peaks within the normal absolute latency and interwave latency intervals of our laboratory are visible.

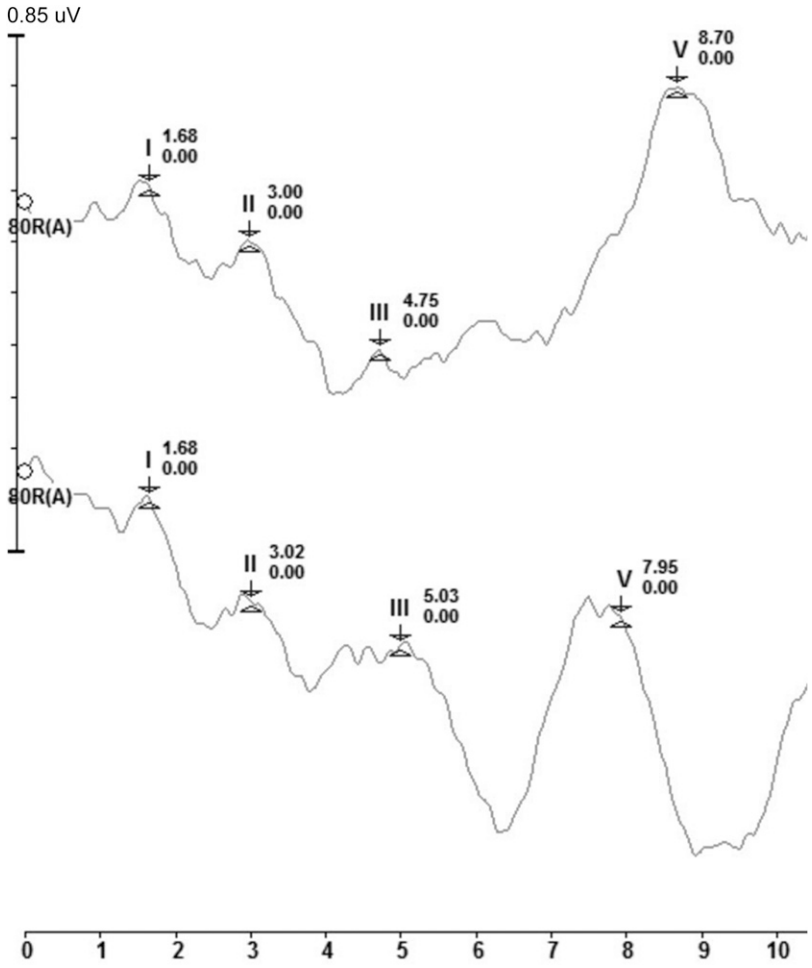


**Figure 4.** ABR right ear result at 80 dB nHL clicks. Stimulus presentation polarity is rarefaction and condensation starting from the top. Prolongation of waves III and V and interwave latencies I–III, I–V, and III–V is evident.

### DPOAE Results

Recordings were compared to laboratory normative data and found to be within normal range (Figure 6). In both ears, absence or abnormality of distortion products was noted in higher frequencies, that is, >6 kHz, and most significant in the right ear, which was expected for a 48-yr-old patient.

### *MRI*

MRI revealed large extra-axial space-occupying lesion of the posterior cranial fossa, located at the right cerebellopontine cistern and the internal auditory canal (Figures 7 and 8).

The mass measured up to $3.5 \times 3.5 \times 2.5$ cm while it puts mild pressure on the pons and the right cerebellar hemisphere.

After intravenous contrast administration, MRI shows prominent and homogenous enhancement (Figure 7).

## DISCUSSION

Clinical importance of APD testing is often overlooked and regarded with skepticism given the challenging interpretation of results and the current growing debate of its nature and clinical entity. Presentation of this case is highly educational as diagnosis of this large cerebellopontine and internal auditory canal lesion was a consequence of addressing the only symptom with which the patient came forward leading to evaluation of her auditory processing abilities. Her auditory processing results showed deficits in SinB perception and DD limited to the right ear as well as deficits in temporal processing and pitch pattern discrimination. Both verbal and nonverbal tests exhibited deficits strictly limited to the right ear, which was in accordance with what she was experiencing as progressive deterioration in speech perception in her right ear compared to the left ear. This less costly evaluation revealed that there was good reason to assess electrophysiologically the auditory system. ABR showed an

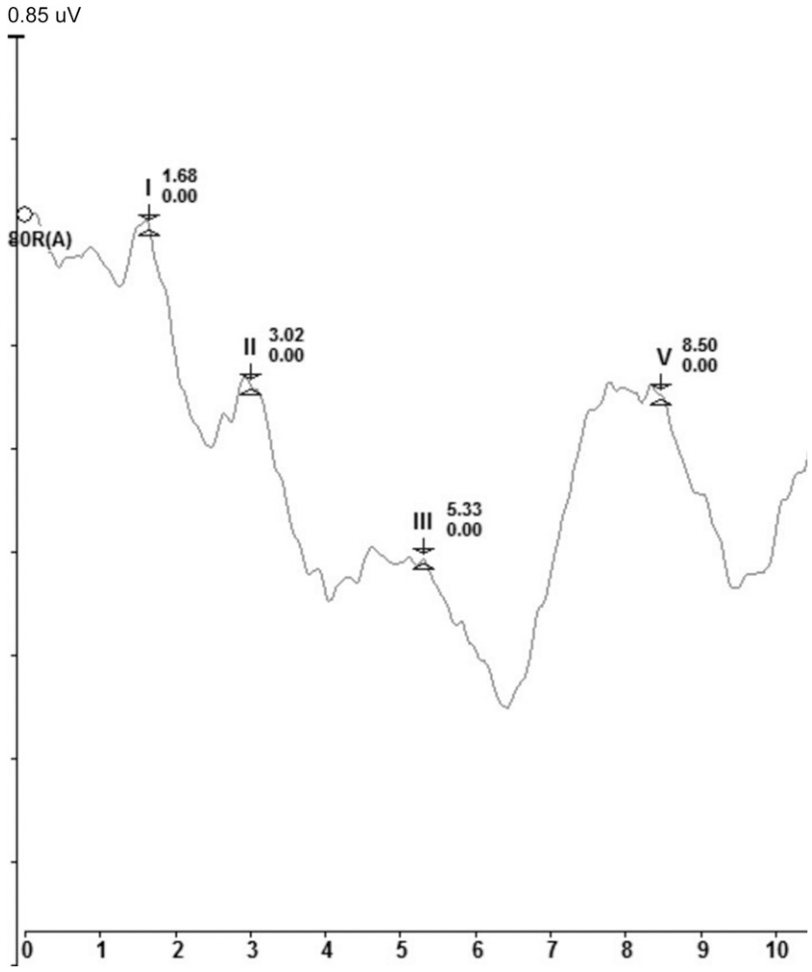


**Figure 5.** ABR right ear result. Digital composite of both click polarities is shown. Prolongation of waves III and V and interwave latencies I–III, I–V, and III–V is evident.

abnormal waveform with either missing or severely prolongated wave V (depending on stimulus polarity). MRI was then implemented revealing a large cerebellopontine and internal auditory canal lesion. Neuroimaging supports either a neuroma or a meningioma as they are both homogenously enhancing solid masses and less an epidermoid inclusion or arachnoid cyst (nonenhancing masses) or an aneurysm (heterogenously enhancing lesion with internal flow voids).

Justification of presenting this clinical case goes beyond current debate on sensory, cognitive, or combined basis of APD. The case shows the clinical need to use currently established diagnostic techniques for APD, as they assist in revealing auditory perceptual abilities and/or deficits going beyond classical audiological evaluation. The latter mostly focuses on pure-tone audiometry, which is considered the answer of "how well someone hears" but in fact is measuring hearing sensitivity, as in hearing pure tones while not accounting for more complex everyday sounds (i.e., speech) and situations (i.e., competing speech and/or other sounds, noisy environments). While research on better understanding the nature of APD and optimizing the currently used diagnostic test battery is extremely important, it should be made clear that current diagnosis of APD is essential in addressing real complaints, which may reveal as shown in the current case, a brain lesion. This may otherwise remain undetected for a long period and be revealed when much more important neurological and/or life-threatening symptoms occur. Early diagnosis may be facilitated by the noninvasive psychoacoustic auditory processing evaluation.

The unilateral nature of the auditory processing deficits in this clinical case indicates the need—as any asymmetry in audiological results—for excluding an eighth nerve or root entry zone lesion. Neuroimaging is rendered necessary to exclude the possibility of existence of a central auditory nervous system (CANS) lesion. However, it may also be the case that a unilateral APD presentation is due to a nonstructural CANS deficit. In the case presented, central auditory processing test battery correlates well with CANS functionality, showing that symptoms of listening difficulties may be measured by these central tests. The unilateral

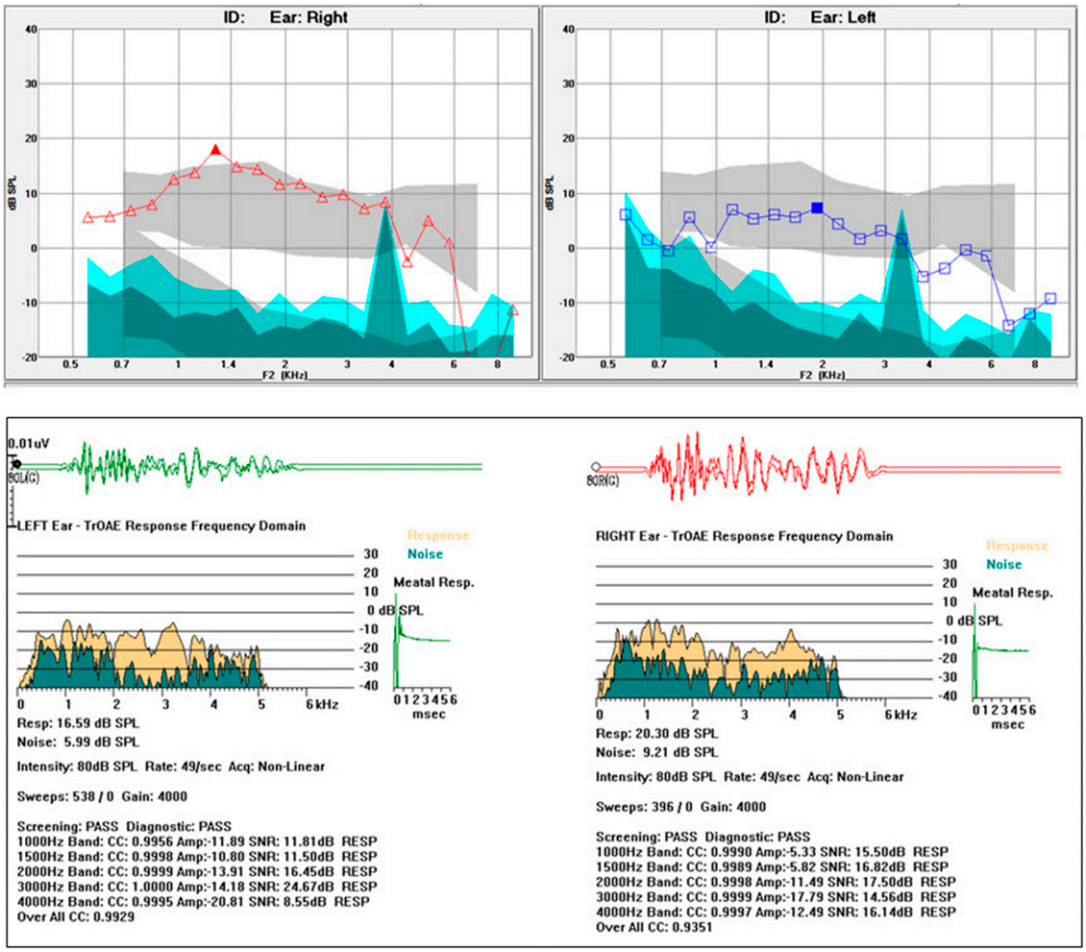


**Figure 6.** TEOAE were recorded within normal range meeting passing criteria, which included SNR of ≥6 dB for every frequency band and a correlation percentage of ≥90%. DPOAE recordings were compared to normative data and found to be within normal range with the exception of higher frequencies (>4000 Hz). (This figure appears in color in the online version of this article.)

nature of APD renders other possibly confounding factors (i.e., cognition, attention, and fatigue) to be thought as less important in the outcome auditory processing evaluation, as these should be equally influencing results of the other ear, which is exhibiting normal auditory processing abilities.

The case presented illustrates the impact of a brainstem lesion in the processing of auditory information. The first case reports exhibiting this impact were audiologically assessed with pure-tone audiometry, speech recognition, tympanometry, contralateral acoustic reflexes ABR, and DD (Musiek et al, 1994). This way an indication of APD may be present but is not documented by an auditory processing test battery approach as is the current recommendations (AAA, 2010). The case presented in this article is the first diagnosed APD adult case with a cerebellopontine angle lesion to the best of the authors' knowledge. The mass identified puts mild pressure on the pons and the right cerebellar hemisphere, influencing the transduction of the auditory information through the right side of the auditory brainstem pathway. This is illustrated in the results of the auditory-evoked potentials testing showing normal processing of information possibly up to the level of the distal auditory nerve (Schwann cell part) on the right side while the left side is unaffected. The I–III latency prolongation implies a neural conduction slowing either at the level of the proximal auditory nerve or the lower brainstem/root entry zone before the generation of wave III. Published data focus on lesions in higher located sites of the CANS (i.e., the lateral lemniscus in Cho et al, 2005; the insula in Bamiou et al, 2006; the auditory cortex in Musiek et al, 2011; the thalamus in Ponzetto et al, 2013). Although it is expected that in cases with large cerebellopontine angle lesions, the contralateral ABR recordings will also be affected, this was not noticed in the presented case report.

It is known to clinicians involved in central auditory processing evaluation that the presence of a neurological lesion as a cause for APD is extremely rare (Griffiths, 2002). However, there is a growing tendency to validate psychoacoustical tests used for central auditory

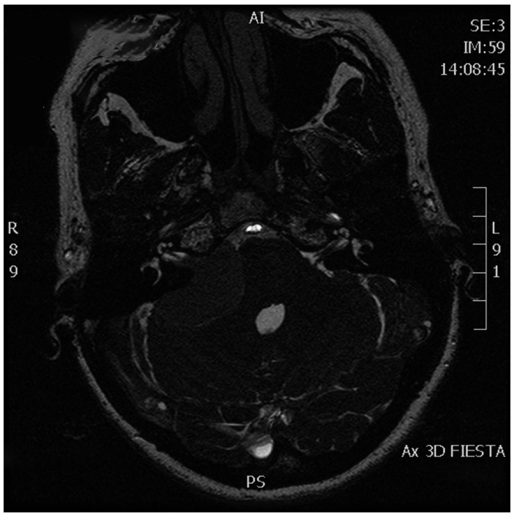


**Figure 7.** Large extra-axial space-occupying lesion of the posterior cranial fossa, located at the right cerebellopontine cistern and the internal auditory canal. The mass measures up to 3.5 × 3.5 × 2.5 cm while it puts mild pressure on the pons and the right cerebellar hemisphere.

processing evaluation in groups of patients with auditory cortex lesions (Chermak and Musiek, 2011; Bamiou et al, 2012). The rationale being that specificity and sensitivity of each test as well as of combination of tests in a central auditory processing battery may be better demonstrated so that clinicians and researchers know what is being tested in each case. The case presented here shows that almost all tests administered were efficient in diagnosing APD. Specifically the SinB Test and the DD together with the nonverbal PPS test and the GIN, all provided a right-ear APD diagnosis. The RGDT was abnormal but since it is binaurally tested there was no indication of specific ear deficit. The only administered test with a negative result for APD (meaning showing normal auditory processing) was the DPS. This could not be attributed to an intact temporal processing as shown by the right-ear abnormal threshold of the GIN. It could be the case that more challenging stimuli might reveal the difficulty in the temporal aspect of auditory processing that is evident in more brief stimuli with rapid transitions (i.e., GIN).

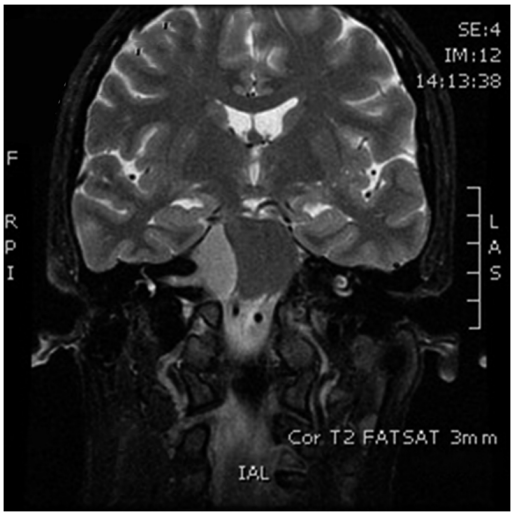


**Figure 8.** After intravenous contrast administration, MRI shows prominent and homogenous enhancement.

In conclusion, this clinical case presentation stresses the importance of testing for APD with a psychoacoustical test battery despite current debate of lack of a gold standard diagnostic approach to APD. In this case, APD diagnosis led to a cerebellopontine lesion identification with extension to the right internal auditory canal. This rare cause of APD demonstrates the efficiency of the current diagnostic test battery for APD in revealing lesional causes in the CANS.

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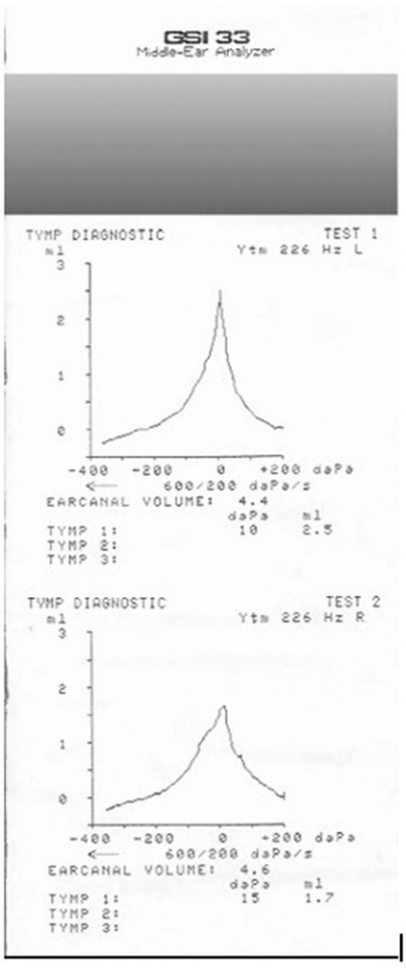
## Appendix




| Ipsilateral stapedial reflexes | 500 Hz | 1000 Hz | 2000 Hz |
|---|---|---|---|
| Right ear | 90 | 90 | 90 |
| Left ear | 90 | 90 | 90 |

Tympanograms were within normal limits for both right and left ears. Ipsilateral stapedial reflexes (500, 1000, and 2000 Hz) were obtained for both ears at 90 dB HL. Technical issues did not permit for testing of contralateral stapedial reflexes and reflex decay.