

CENTRAL AUDITORY EVALUATION IN MULTIPLE SCLEROSIS

Case report

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ABSTRACT - Herein, we report a case of multiple sclerosis in which peripheral and central hearing, were evaluated through early (brainstem), middle and late auditory evoked potentials before and after corticosteroid therapy. Auditory evaluation revealed better performance on all post-treatment tests. In this case, central auditory function tests (behavioral and electrophysiological) identified the location of the impairment (brainstem), which was in agreement with the patient complaint. The speech in noise test and brainstem auditory evoked potentials are definitely appropriate in confirming brainstem lesions.

KEY WORDS: multiple sclerosis, electrophysiology, auditory perceptual disorders.

Avaliação auditiva central na esclerose múltipla: relato de caso

RESUMO - Relatamos caso de esclerose múltipla em que foi feita avaliação da audição periférica e central utilizando os potenciais evocados auditivos de curta, média e longa latência antes e depois da terapia com corticosteróides. A avaliação auditiva revelou melhor desempenho em todos os testes após o tratamento. Neste caso, os testes que avaliam a função central da audição (comportamental e eletrofisiológico) foram capazes de identificar o local da lesão (tronco encefálico), o que estava de acordo com as queixas do paciente. Os testes de fala com ruído e os potenciais evocados auditivos de curta latência são apropriados para revelar lesões de tronco encefálico.

PALAVRAS-CHAVE: esclerose múltipla, eletrofisiologia, distúrbios perceptuais auditivos.

The advent of magnetic resonance imaging (MRI) techniques represents a major advance in the diagnosis of multiple sclerosis (MS). Although neurophysiological tests, that of evoked potentials in particular, have great value in the diagnosis of MS, they have not been widely used for the diagnosis of MS, despite the fact that MRI is costly and is available at only a few health care facilities. Technical advances, together with new methods of investigating afferent and efferent nervous pathways, seem to have increased the sensitivity of neural dysfunction detection, but the clinical gains have been modest at best. More promising is the use of neurophysiological tests to quantify the extent of white matter involvement¹. The demyelination or sclerosis (scarring) induces a

slowing of nerve impulse propagation. Impaired conductance is reflected in an increase in latency of evoked potentials. Abnormal evoked responses to different types of stimuli provide clues for the location of plaques or lesions, confirm clinically ambiguous lesions and confirm the organic basis of symptoms. A large proportion of patients with established MS also show lesions of the central auditory pathways, which can be identified by brainstem auditory evoked potentials (BAEPs), middle-latency auditory evoked potentials (MLAEPs) and late auditory evoked potentials (LAEPs), as well as by using neuroimaging procedures²⁻⁴. Unfortunately, many professionals are not cognizant of the auditory deficits that may be associated with this pathology. As a result, these deficits

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often go undetected, especially if assessment is limited to a peripheral test battery. Musiek et al.⁵ found that 18% of their MS subjects had significant hearing losses, although more than 40% of their subjects with normal peripheral hearing presented auditory complaints. They also reported that 80% of their subjects presented an abnormality on at least one auditory test when central as well as peripheral hearing tests were administered. Celebisoy et al.⁶ studied MLAEPs and BAEPs in 30 patients with MS. They found BAEP abnormalities in 18 of the patients and MLAEP abnormalities in 22. In 15 of the 30 patients studied, BAEPs and MLAEPs were both abnormal. In 7 of the 12 patients with normal BAEPs, MLAEPs were found to be abnormal. Of the 18 patients with abnormal BAEPs, only 3 presented normal MLAEPs.

The purpose of this paper is to report behavioral and electrophysiological findings, before and after corticosteroid therapy, in an MS patient presenting hearing complaints.

CASE

The patient gave an informed consented for this case report.

A 27-year-old right-handed man, diagnosed with MS 11 years prior and using interferon beta-1a, presented in December 2002 an acute crisis during which he experienced blurred vision and, for the first time, hearing difficulties in the right ear. The hearing complaints included difficulty hearing in noisy environments and a lack of tolerance for loud sounds. One week after the onset of the symptoms,

the patient visited the Speech and Hearing Department at the University of São Paulo School of Medicine for audiologic evaluation. After the first evaluation, the neurologist prescribed corticosteroid therapy. After seven weeks, the patient returned for new evaluation involving the same procedures.

Pre- and post-treatment audiologic evaluation included conventional pure tone audiometry (250-800 Hz), performance-intensity functions, immittance measures, the speech in noise test and the staggered spondaic word (SSW) test, as well as measurement of BAEPs, MLAEPs and the P300 component of the LAEPs. A two-channel audiometer (GSI 61; Grason Stadler) was used for pure tone audiometry, speech audiometry and behavioral tests of central auditory processing. A GSI 33 middle ear analyzer (Grason Stadler) was used for immittance measures.

Electrophysiological procedures were carried out using Biologic Traveler Express equipment. For BAEP we used rarefaction clicks at 80 dB HL, presented at 19 clicks/sec. The presence and absolute latencies of waves I, III and V, as well as the interwave latencies I-III, III-V and I-V, were analyzed. For MLAEP 70-dB HL rarefaction clicks were presented at 10 clicks/sec using the following electrode sites: forehead (as ground), left and right ears (A1 and A2) and both temporal lobes (C3 and C4). The MLAEP wave Pa latency and amplitude measures were registered ipsilaterally (C3/A1 and C4/A2) and contralaterally (C3/A2 and C4/A1). For P300 we used clicks at 75 dB HL, presented at 1.1 clicks/sec, to analyze the presence and latency of P300.

Results – After corticosteroid therapy, pure tone thresholds in the right ear improved by 10-15 dB at all frequencies (Table 1). As can be seen in Table 2, the acoustic reflexes in the right ear also improved by 10-15 dB after treat-

Table 1. Pre- and post-treatment pure tone audiometry.

	0.25 kHz		0.50 kHz		1 kHz		2 kHz		3 kHz		4 kHz		6 kHz		8 kHz	
	Pre	Post	Pre	Post	Pre	Post	Pre	Post	Pre	Post	Pre	Post	Pre	Post	Pre	Post
AC-RE	20	10	20	5	15	5	15	0	5	0	10	0	10	10	15	5
BC-RE			20	5	15	5	15	0	5	0	10	0				
AC-LE	20	10	15	10	5	5	0	0	0	0	10	5	15	10	10	5
BC-LE			15	10	5	5	0	0	0	0	10	5				

AC, air conduction; RE, right ear; BC, bone conduction; LE, left ear.

Table 2. Pre- and post-treatment acoustic reflexes.

	0.50 kHz		1 kHz		2 kHz		4 kHz	
	Pre	Post	Pre	Post	Pre	Post	Pre	Post
IPSI RE	100	85	95	85	95	90	95	90
CONTRA RE	115	105	100	95	105	95	95	85
IPSI LE	95	90	85	85	80	85	80	90
CONTRA LE	95	100	90	95	90	95	85	85

IPSI, ipsilateral; RE, right ear; CONTRA, contralateral; LE, left ear.

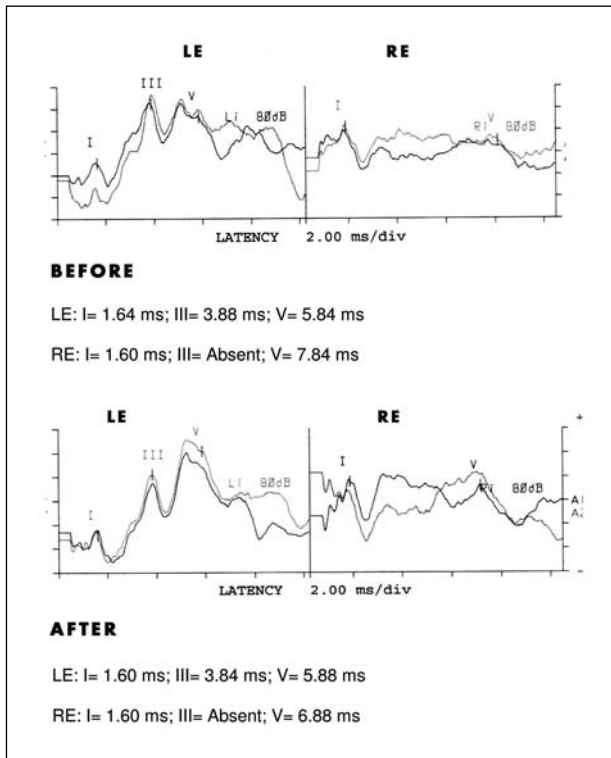


Fig 1. BEAPs before and after treatment. LE, left ear; RE, right ear.

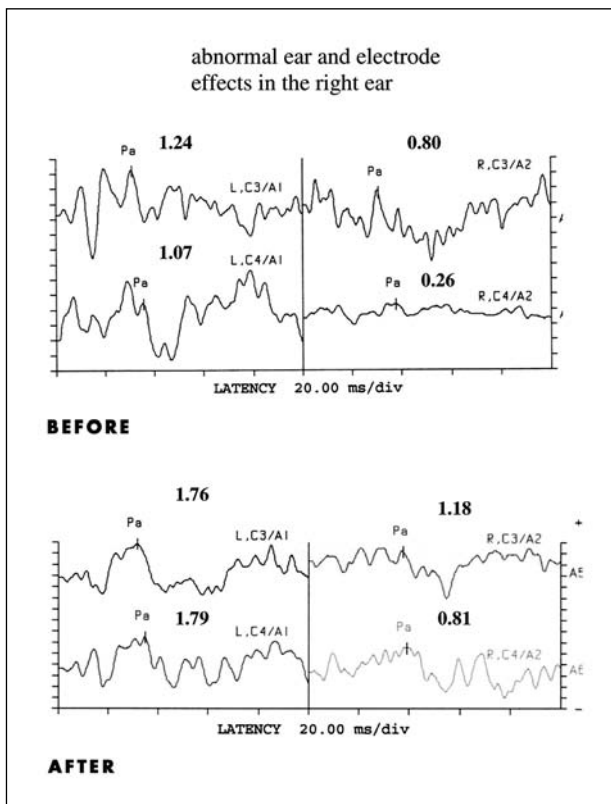


Fig 2. MLAEPs before and after treatment.

Table 3. Pre- and post-treatment behavioral evaluation of the auditory pathway.

	Speech in noise test		SSW test	
	Pre	Post	Pre	Post
RE	64%	84%	90%	90%
LE	80%	84%	90%	90%

SSW, staggered spondaic word; RE, right ear; LE, left ear.

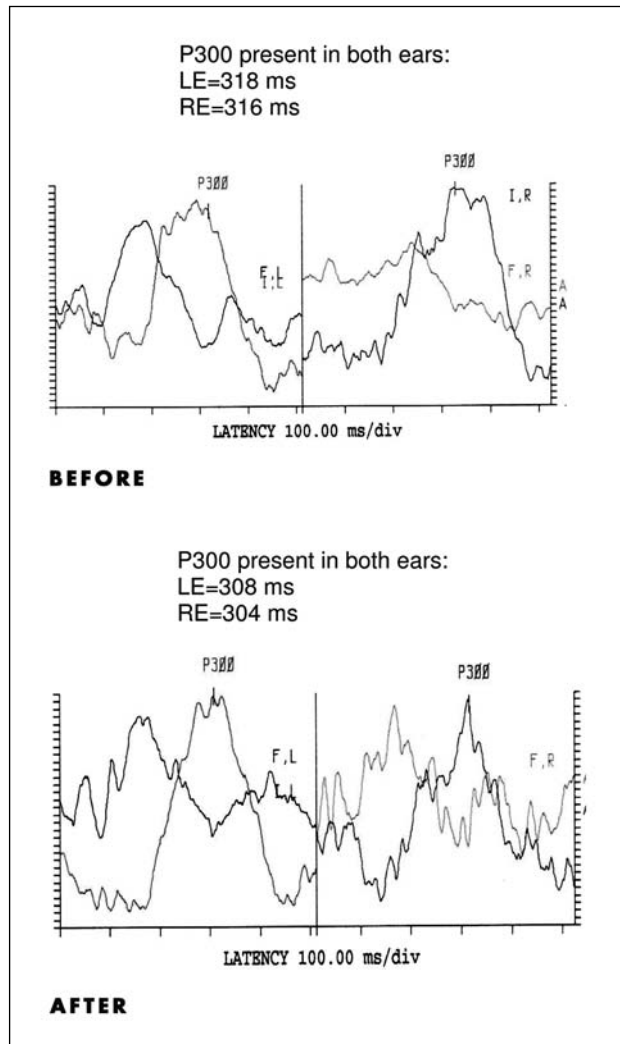


Fig 3. P300 before and after treatment. LE, left ear; RE, right ear.

ment. In addition, the speech in noise performance of the right ear improved after treatment (Table 3). Figure 1 shows all BAEP waves for both ears before and after treatment. The Pa amplitudes for all of the electrode sites (C3A1, C3A2, C4A1 and C4A2) increased after treatment (Fig 2). In Figure 3, it can be seen that there was no significant difference between pre- and post-treatment values for P300 latency in either ear.

DISCUSSION

The patient presented noticeable improvement in auditory symptoms after corticosteroid therapy. Although post-treatment improvement was observed in the pure tone audiometry results, as well as in those of the acoustic reflex test, it is of note that peripheral hearing was found to be within normal limits. Therefore, had a more thorough hearing examination not been performed in order to evaluate central auditory processing, it would not have been possible to identify any alterations.

The post-treatment results of the speech in noise test were normal for the right ear, and the SSW test results were essentially normal before and after treatment (Table 3). These results confirm the fact that the patient presented abnormalities at the level of the brainstem but not at the more central level.

Impairment of binaural processing can occur in MS due to the demyelination of many pons structures, such as the superior olives and a region between the two inferior colliculi, that are responsible for the function involved in processing speech in noise.

In the electrophysiological evaluation of the right ear, the first BAEP evaluation demonstrated no wave III and a 2-ms delay (7.84 ms) in absolute latency of wave V in comparison to the left ear, evidencing brainstem impairment on the right side. In the second evaluation, we noticed a decrease in absolute latency of wave V (6.84 ms), 1 ms sooner than in the first evaluation. Matathias et al.² also reported BAEP abnormalities in 50% of the MS patients studied. Walsh, Kane and Butler⁷ reported that BAEPs are more likely to be abnormal when demyelination affects the brainstem clinically, but they can also detect "silent" lesions in approximately 40% of patients who do not have symptoms or signs of brainstem involvement. Delay is probably indicative of the MS-related demyelination of the auditory pathways. It is important to remember that vascular, inflammatory, neurodegenerative, metabolic and infectious conditions may mimic MS lesions on MRIs. Therefore, a diagnosis of MS may not be made based on the presence of MRI lesions alone and requires corroboration through appropriate clinical or other tests. When MS presents as a clinically isolated syndrome, the criteria for dissemination over time must be met in order for a definitive clinical diagnosis of MS to be made. Dissemination in space may often be demonstrated by evoked potential testing.

Bergamaschi et al.³ found that BAEP abnormalities decreased progressively to normalization that

coincided with clinical recovery. Although, for our patient, we had no access to BAEP test results obtained prior to symptom onset, recovery was coincident with the decreased wave V latency. Therefore, we strongly recommend that all patients diagnosed with MS (even those presenting no auditory symptoms) be submitted to BAEP testing as a baseline evaluation for later comparison.

The first MLAEP test revealed ear and electrode effects for the right ear. The ear effect was not found in the second evaluation. Myelin not only protects nerve fibers but enables their function. When myelin or nerve fiber is damaged or destroyed, the ability of the nerves to conduct electrical impulses to and from the brain is disrupted, and this produces the various symptoms of MS.

Celebisoy et al.⁶ found that 60% of the patients with confirmed brainstem involvement presented BAEP abnormalities. In addition, the MLAEP results were abnormal in 73.3% of the patients. They concluded that the use of BAEP measures in combination with MLAEP measures is a more comprehensive means of evaluating brain function in such patients. Another study⁸ showed that up to 79% of MS patients with clinical evidence of brain stem involvement presented abnormal BAEPs.

The P300 and SSW test results were within normal limits before and after treatment, demonstrating that the disease had not affected the higher levels (cortical areas) of the auditory pathway. However some researchers have found abnormalities in the LAEPs of patients with MS^{4,9,10}. In such cases, one might infer that the disease had not affected the auditory areas related to the generation of such potential. Therefore, the behavioral and electrophysiological results were in agreement, contributing to the determination of the site of lesion^{2,3,4,6,10}.

All tests of central auditory function (behavioral and electrophysiology) were good predictors of lesion. We feel that the results of this case study are significant since impairment of at least two distinct, unrelated functions is necessary in order to establish a diagnosis of MS. Auditory complaints are quite common and almost always unspecific. Therefore, physicians are typically more attentive to the more frequently reported visual alterations, to the detriment of the auditory complaints, which, as a consequence, are rarely investigated. There is no doubt that MRI brain scans represent an advance in the diagnosis of MS, which is a dynamic disease, lesions always develops, and many of those lesions are clinically silent.

Therefore, we must prioritize the allocation of existing resources. It is feasible to use those resources with precision, thereby achieving two main objectives: the follow-up treatment of pre-established profiles; and the diagnosis of new lesions that might develop.

Using central auditory tests (behavioral and electrophysiological), we were able to identify the exact location of the impairment (brainstem), which was in agreement with the patient complaint. We found the speech in noise test and the BAEP test both to be definitely appropriate in confirming brainstem lesions.

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