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Vestibular Evoked Potentials in Two Patients with Bilateral Vestibular Loss

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Abstract: The Brain Electrical Activity Mapping (BEAM) of Vestibular Evoked Potentials (VestEP) is a new method in the toolbox of the neurootologist. Two cases of bilateral vestibular loss, diagnosed by classical neurootological methods (highlighted by caloric and optokinetic tests' electronystagmography recording), are reported. The additional information, provided by the Vestibular Evoked Potentials (VestEP) and its high clinical significance for a more accurate neuro-otologic diagnosis are discussed.

INTRODUCTION

The aim of following two case presentations is to demonstrate the method of cortical Vestibular Evoked Potentials (VestEP) displayed by its combination with Brain Electrical Activity Mapping (BEAM).[1] The VestEP-BEAM method provides reliable clinical information about the functional state of the vestibular end-organ, its brain stem pathways and cortical representation. Its clinical application for the past five years improved the accuracy of neurootologic diagnosis for patients suffering with complaints of vertigo, tinnitus and hearing loss.[2]

MATERIAL AND METHODS

The stimuli we use for eliciting VestEP[3] are repetitive short lasting rotations, i.e., step-wise angular accelerations of the subject's body around the vertical axis, with acceleration/deceleration phases lasting 1.0 second each, an interstimulus interval of 14.0 seconds and an acceleration of 15°/s^2. A total of 25 acceleration steps are applied for each clockwise (CW) and counterclockwise (CCW) rotations. Recording electrodes are positioned in accordance to the international 10/20 system, as referent electrode being used a common mastoid bridge electrode. An epoch of 1000 msec. following the onset of the stimulus is used for the analysis of the rotationally evoked brain electrical events.

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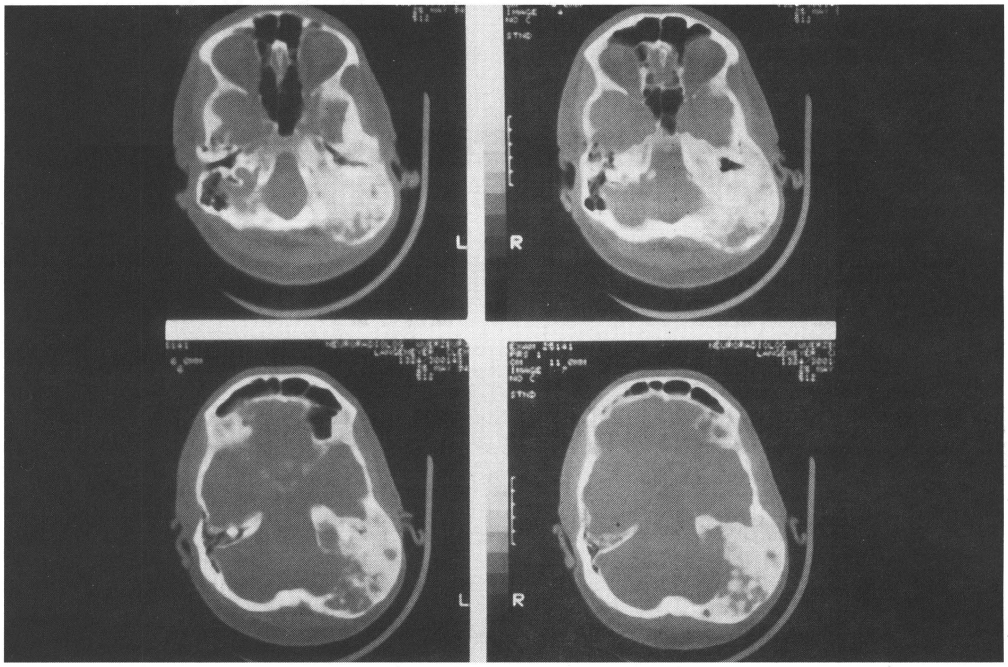
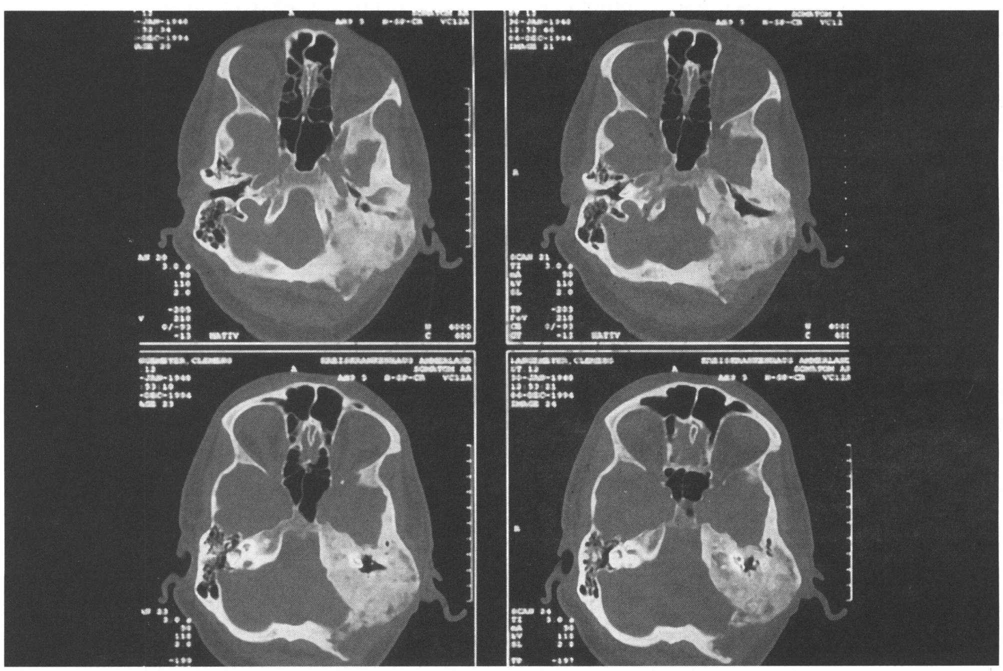
CASE REPORTS

Case 1

A 47-year-old male was referred in 1995 to the Neurootology department with a presumed diagnosis of total bilateral vestibular loss. His neurootological complaints included unsteadiness increased by eye closure and in the dark; episodic severe vertigo; anacousia left; severe hypoacousia right served by a hearing aid.
The history of his disease began in 1963, with the diagnosis of a complete cochleovestibular loss left, due to a *Jaffé-Lichtenstein Syndrome* (fibrose bone dystrophy with secondary cysts formation at left petrous bone, mastoid process, and occipital squama). The highlights of the clinical course of the cochleovestibular complaints also include:
1980 - Right *otitis herpetica* with right facial palsy and the complication of *encephalitis herpetica* with resultant progressive severe hearing and vestibular loss right.
1992 - Diabetes mellitus type II, with secondary poly-neuropathy.
1994 - Retromastoid craniectomy with extended cyst resection left. (Figure 1B)
The most significant examinations and tests done prior to referral in 1995 included:
BERA
Absent potentials for the left ear and pathological morphology of the curves, with prolonged interpeak latency of the waves III-V for the right ear.
CT
Large alterations of the left mastoid, petrous and temporal bones (bone proliferation, fibrosis, chronic inflammation, cicatrization); slight compression upon the left



A. Before surgery (May 1994)



B. After surgery (Dec. 1994)

Figure 1. Case 1 - C.T. images. *Jaffé-Lichtenstein Syndrome* affecting left petrous bone, mastoid process, and occipital squama: **A.** Before surgery; **B.** After surgery.

cerebellum hemisphere; and a non-specific enhancement of the right labyrinth. (Fig. 1A and 1B)

Neurological examination
Left anacousia; severe right hypoacousia; complete bilateral vestibular loss; very slight paresis of the right facial nerve; and no signs of compression of the cerebral structures.

The complete neurootological investigations,[4] done in our department, revealed:
Electronystagmography (ENG)[5]
- Optokinetic testing: normal.
- Calorization: total right areflexia and impossible left calorization, because of the obstruction of the external auditory canal by bone proliferation.
- No response to rotatory testing.

Cranio-Corpo-Graphy (CCG)[6]
Extreme ataxia. The patient was not able to perform more than 30 steps during Unterberger test (normal=100) and to keep standing more than 27 seconds (normal=60 sec.) in Romberg test. (Fig. 2)

VestEP displayed by BEAM
No proper VestEP was recorded at either CW or CCW rotations.[7] (Fig. 3A and 3B) However, very small and hardly detectable responses with latencies of about 330 msec. (normal latency of wave III) can be seen in BEAM during both the rotations, mostly over the frontal areas, but they do not resemble the typical VestEP. (Fig. 4A and 4B)
These could be cognitive potentials due to proprioceptive stimuli, coming from cervical and body receptors.

The neurootologic diagnosis included:
- Complete bilateral vestibular loss.
- Central (cerebellar) ataxia.
- Left anacousia, severe right hypoacousia.

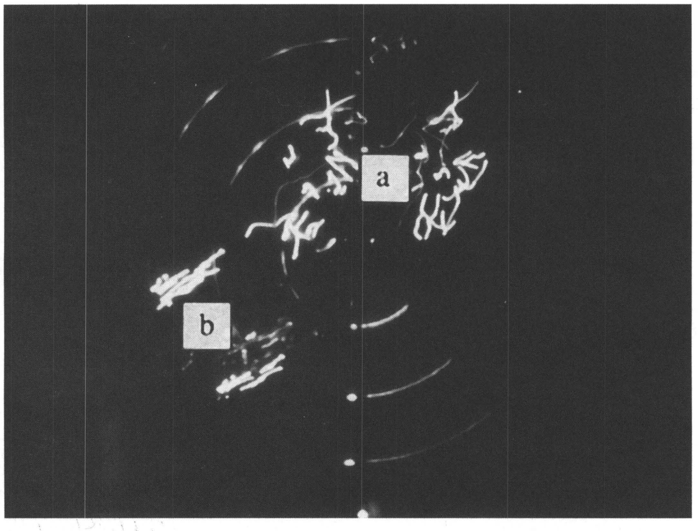
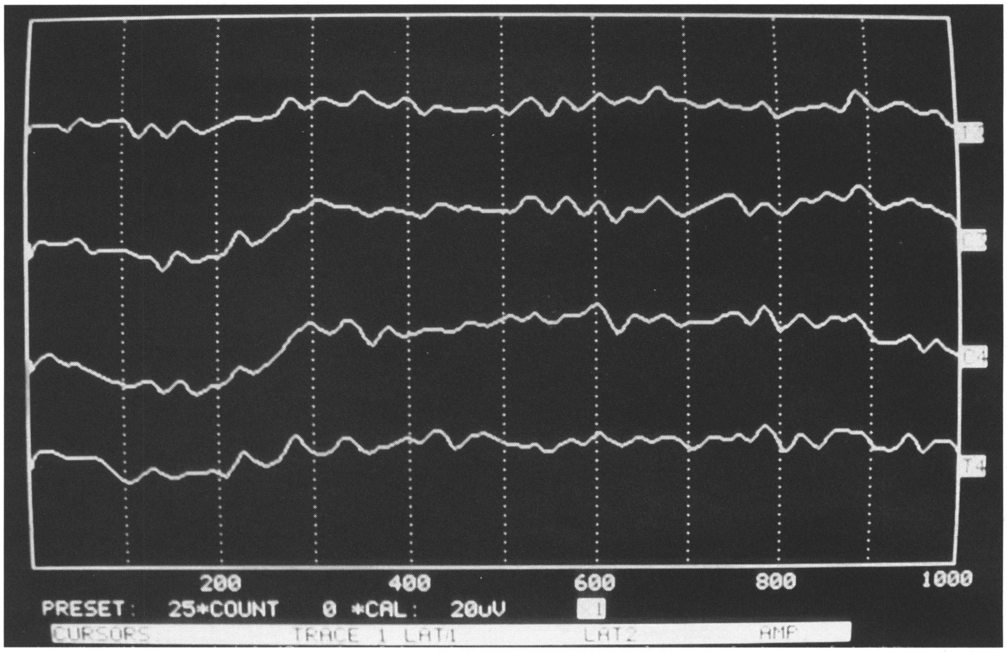
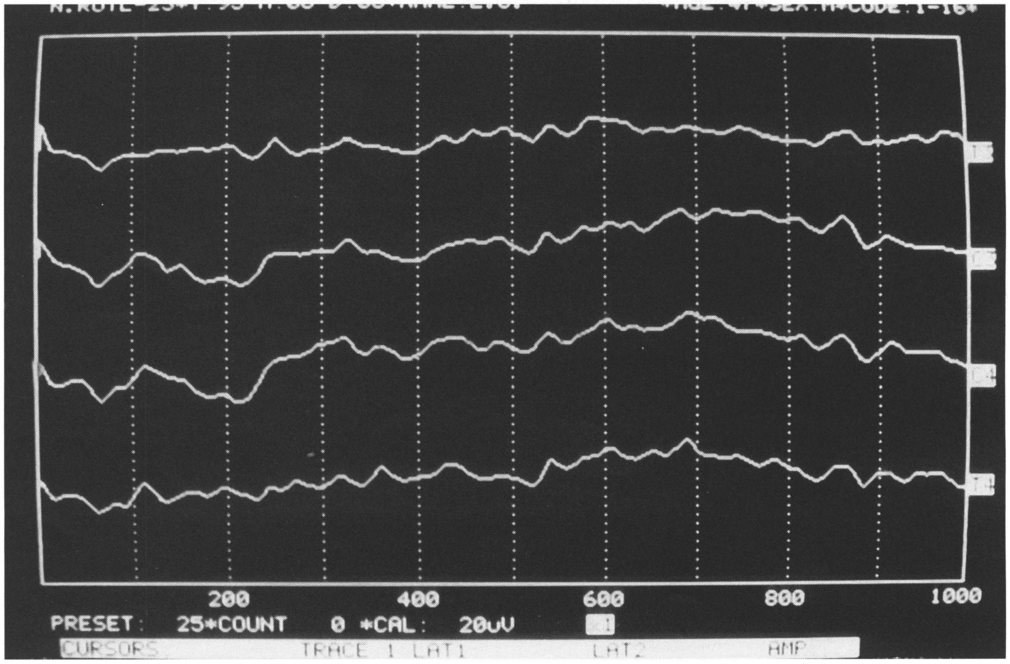


Figure 2. Case 1 - CCG recording: extreme ataxia in both, stepping test (a) and standing test (b).

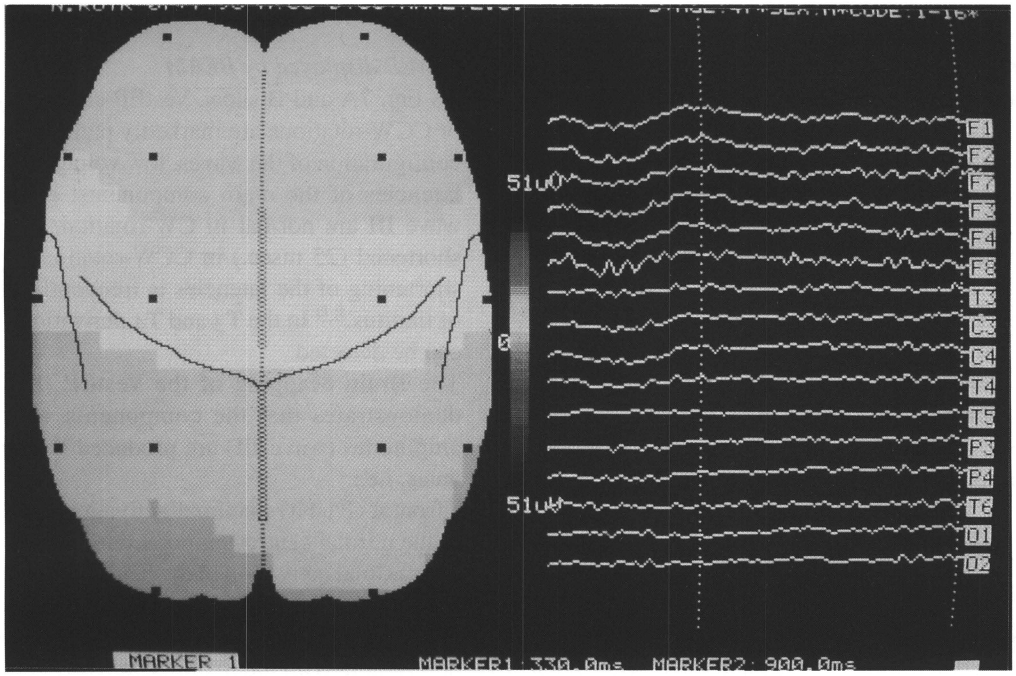


A. Clockwise (CW) rotation

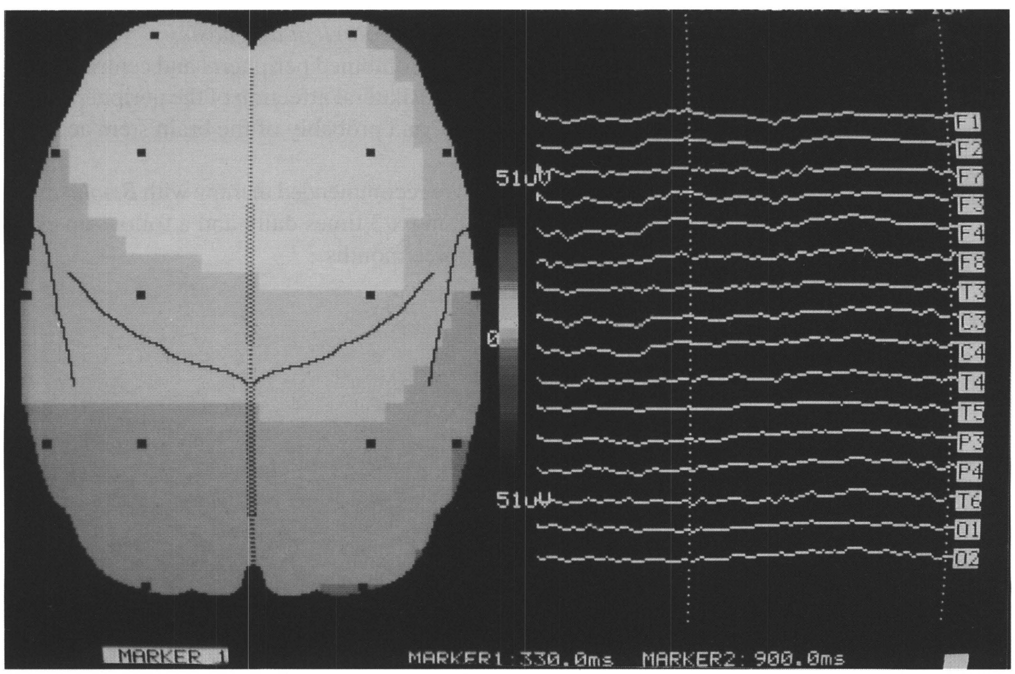


B. Counterclockwise (CCW) rotation

Figure 3. Case 1 - Cortical electrical activity as recorded at the T3, C3, C4, T4 electrodes/derivations, during vestibular stimulation through: **A.** CW-rotation, and **B.** CCW-rotation. No proper VestEP can be recognized.



A. Clockwise (CW) rotation



B. Counterclockwise (CCW) rotation

Figure 4. Case 1 - Brain Electrical Activity Mapping during CW- and CCW-rotation. The coloured scales (centers) show the possible intensities of the cortical activity, related to the indifferent mastoid electrode, i.e.: dark red = very high cortical activation; yellow/light green (0) = basical, spontaneous activity; dark blue = very low level of cortical activity. In none of the images any areas with a proper activation can be recognized. The most active areas (dark yellow), corresponding to the suspected electrical events with latencies of about 330 msec., have a frontal distribution.

Case 2

A 57-year-old male was referred to our department in September 1995 for neurootologic investigation, for a presumed diagnoses of complete bilateral vestibular loss and suspicion of right acoustic neurinoma. His neurootological complaints were subjective bilateral tinnitus of *Tinnitus cranii sive cerebri* type, and a sudden aggravation of the bilateral high frequency hearing loss. The severity of these complaints was relatively mild at onset in 1989, but then it increased progressively. Since 1985, the patient also complained of sinoatrial bradycardia and tendency to low blood pressure. In 1987, he had a sport accident with light head and neck trauma, of the concussive type.
Tests done prior to referral included:

Conventional Audiometry (Fig. 5A and B)

BERA
Delayed acoustic brain stem evoked potentials, to the level of the *Colliculus inferior*, bilaterally, with either left and right stimulation.

MRI
Normal image of cerebello-pontine angle, meatus acousticus internus, VIIIth cranial nerve, cochlea, and semicircular canals.

Complete neurootologic investigation done in our department revealed:

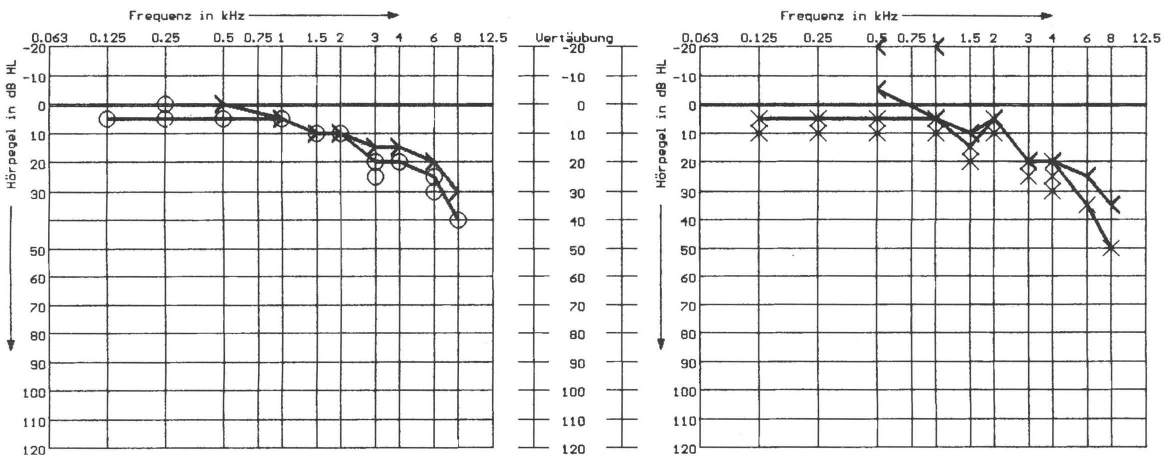
ENG - Optokinetic testing: normal
- Complete bilateral areflexia in both caloric and rotatory testing.

CCG - Pathological right deviation in Unterberger testing. (Fig. 6)

VestEP displayed by BEAM
As Fig. 7A and B show, VestEP elicited by either CW- or CCW-rotations are markedly pathological: abnormal configuration of the waves, low voltage amplitudes. The latencies of the main components, and especially of wave III are normal in CW-rotation, but significantly shortened (25 msec.) in CCW-rotation. A pathological shortening of the latencies is frequently found in cases of tinnitus.[8,9] In the T3 and T4 derivations no potentials can be detected.
The Brain Mapping of the VestEP (Fig. 8A and B) demonstrates that the components with the highest amplitudes (wave III) are produced in atypical cortical areas, i.e.:
- frontal (F3-F4) maximal activations, in contradiction to the normal centro-temporal ones (T3-C3-C4-T4);
- a maximal activation in the hemisphere contralateral to the side of rotation, i.e., the right hemisphere in rotation to the left (CCW) and vice-versa (normally, the hemisphere with the maximal activation is ipsilateral to the side of rotation). No interhemispheric differences were identified.

Neurootological diagnosis:
- Combined peripheral and central vestibular disorder.
- Bilateral affection of the peripheral acoustical organs and probably of the brain stem acoustical pathways.

We recommended therapy with *Betahistine* (Vasomotal®) 8 mg x 3 times daily and a follow-up examination after three months.



A. Right ear **B.** Left ear

Figure 5. Case 2 - Conventional pure tone audiograms.

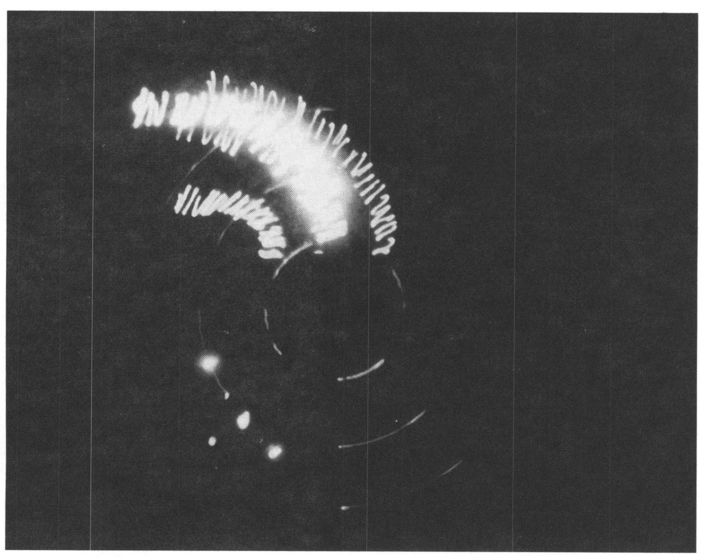
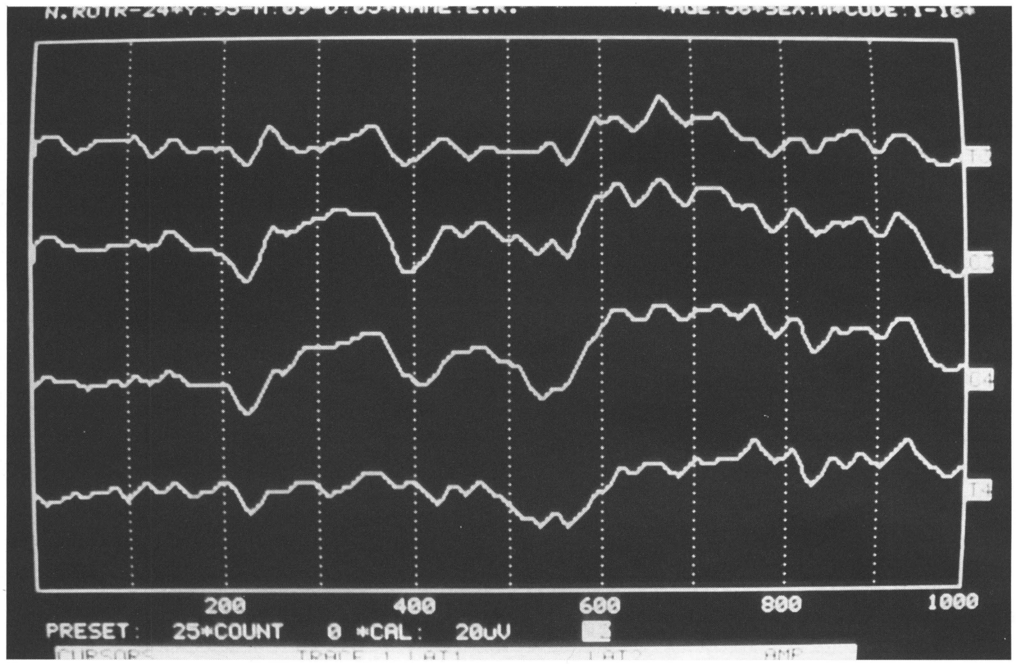
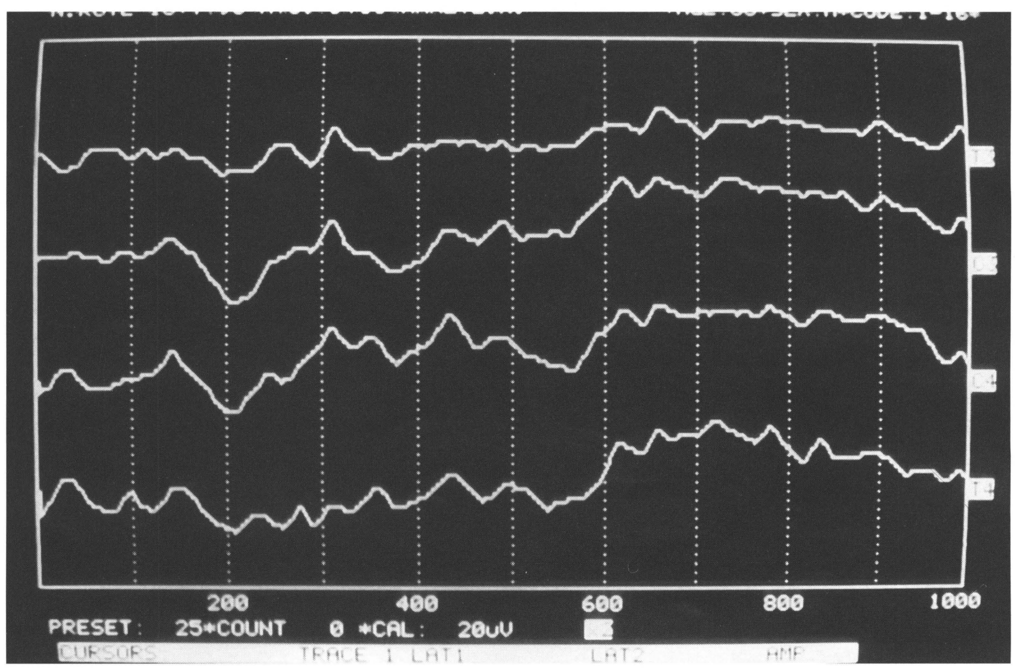


Figure 6. Case 2 - CCG recording: pathological right deviation in the stepping test (mirrored image).

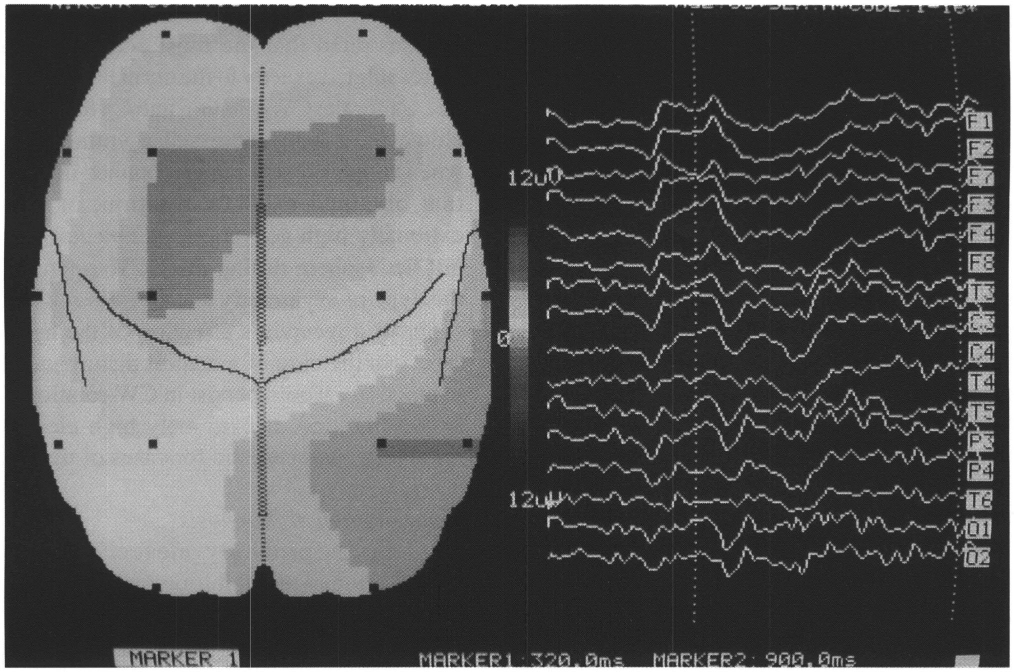


A. Clockwise (CW) rotation

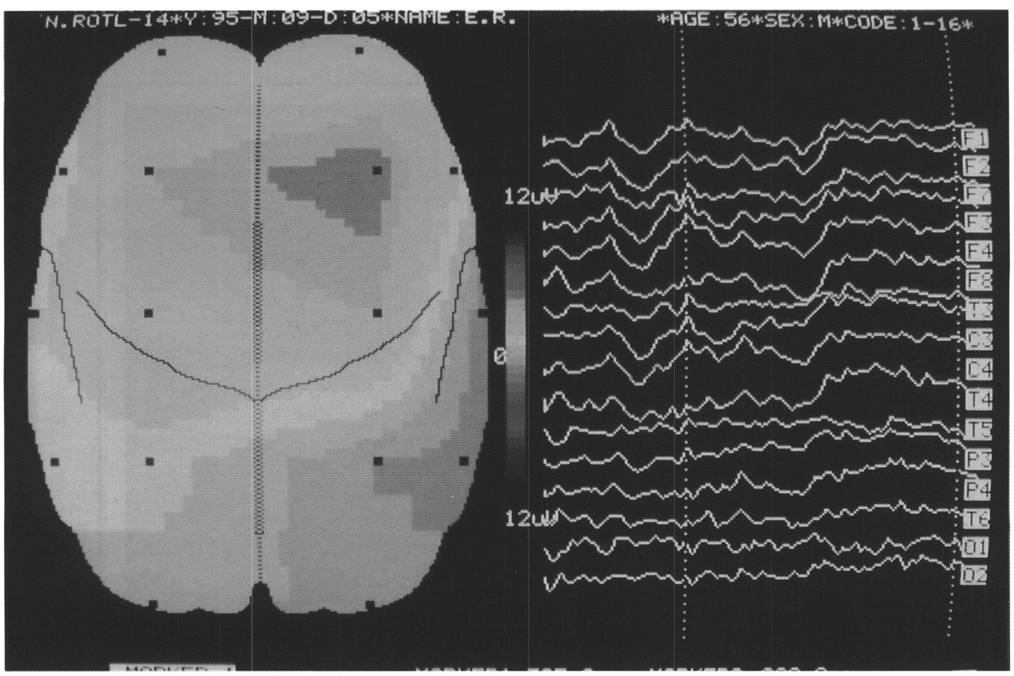


B. Counterclockwise (CCW) rotation

Figure 7. Case 2 - Markedly pathological VestEP elicited by either **A.** CW or **B.** CCW rotations. No potentials are recorded at the T3 and T4 electrodes/derivations.



A. Clockwise (CW) rotation



B. Counterclockwise (CCW) rotation

Figure 8. Case 2 - Brain Electrical Activity Mapping of the VestEP in **A.** CW-rotation; **B.** CCW-rotation.

The patient returned after three months of therapy with unchanged bilateral tinnitus and reported a slight amelioration of the hearing, which was objectified with audiometric testing (10dB at the right ear, 5 dB at the left ear).

BERA
showed the tendency to normalization of the latencies of brain stem acoustically evoked potentials.

The results of our neurootological tests included:
ENG - Optokinetic testing: normal
- Dysrhythmia, convergent dissociation
- Slight, right beating spontaneous nystagmus
- Calorization: symmetric nystagmus reaction, with very low amplitude and frequency
- Rotation: no nystagmus reaction
- Vestibular decruitment

CCG - Minor changes according to the age. (Fig. 9)

VestEP displayed by BEAM
A moderate but clear improvement of the VestEP-parameters could be noticed. The potentials were better detectable in all the examined derivations; the waves had higher amplitudes and significantly better morphologies. (Fig. 10 A and B) There was also a tendency to normalization of the latencies, especially of the wave III, in the CCW-rotation (15 msec. closer to the normal value). The Brain Mapping of the VestEP (Fig. 11 A and B) demonstrated that the most active cortical areas now corresponded exactly to the normally expected electrodes, i.e., C_4 for the CW-rotation and C_3 for the CCW-rotation. However, a well expressed asymmetry could be seen when comparing the image obtained in CW-rotation with that obtained in CCW-rotation, in the sense of an extremely high activation, i.e., electronegativity, in the left hemisphere during the CCW-rotation. We describe this type of asymmetry as labyrinth related, as it usually suggests a receptor's affection of the hyper-excitability type.[7] In the case of a central disturbance, this focus of overactivity would persist in CW-rotation.[8] On the other hand, this kind of extremely high electronegativity is relatively characteristic for cases of tinnitus.[9]

Neurootological diagnosis
- Central (probably mesencephalic) vestibular affection, with inhibition of the nystagmus generator centers.
- Bilateral affection of the acoustical receptors and probably of the brain stem acoustical pathways.

The patient was recommended to continue therapy with Betahistine, and to make supplementary cardiovascular and blood tests (coagulation study, serum cholesterol and triglyceride etc).

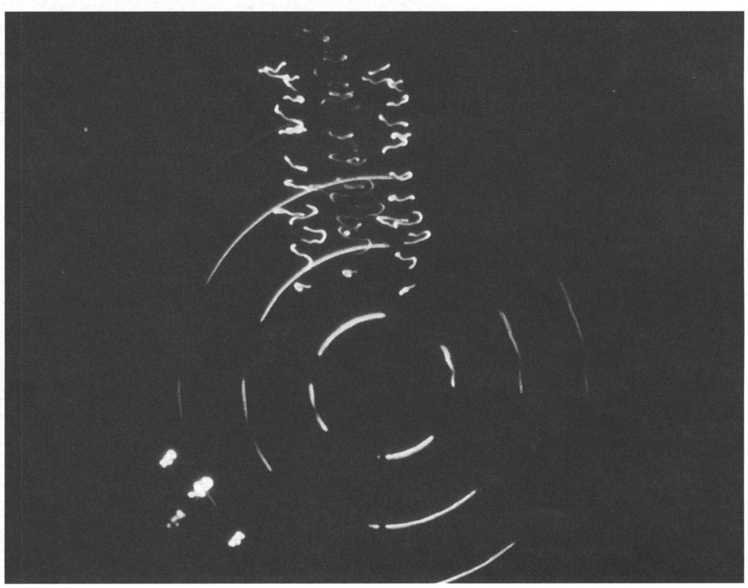
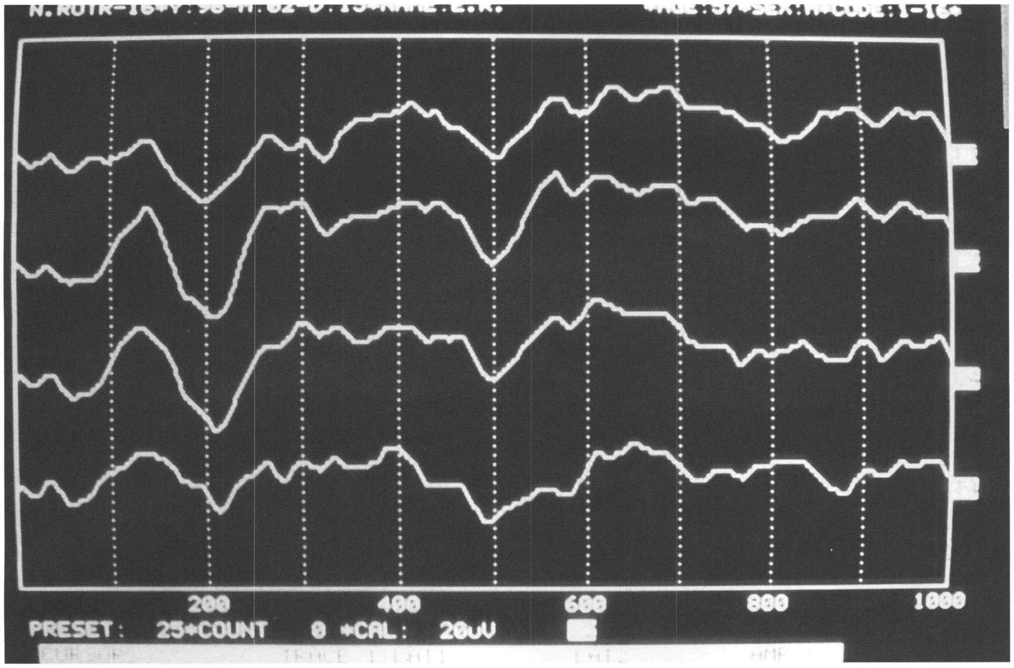
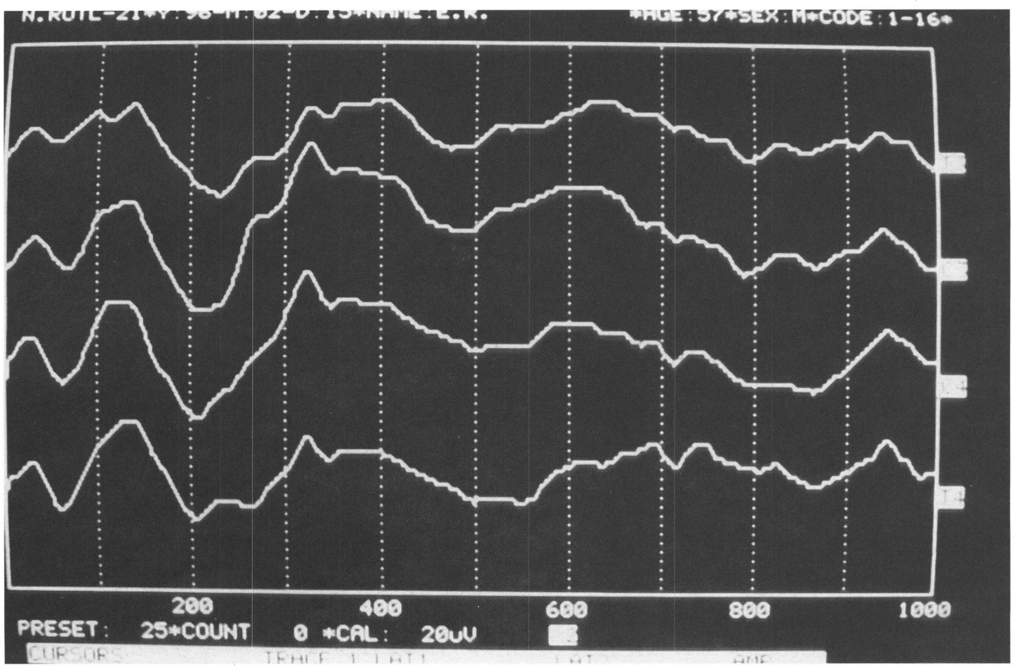


Figure 9. Case 2, follow up examination - CCG recording: normal aspect, based on age of patient.

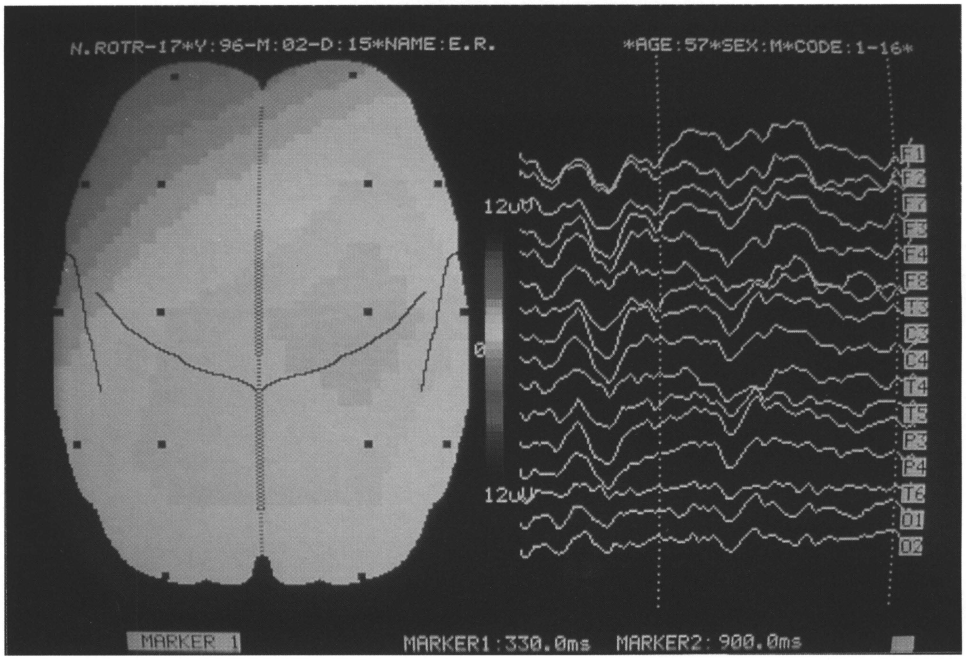


A. Clockwise (CW) rotation

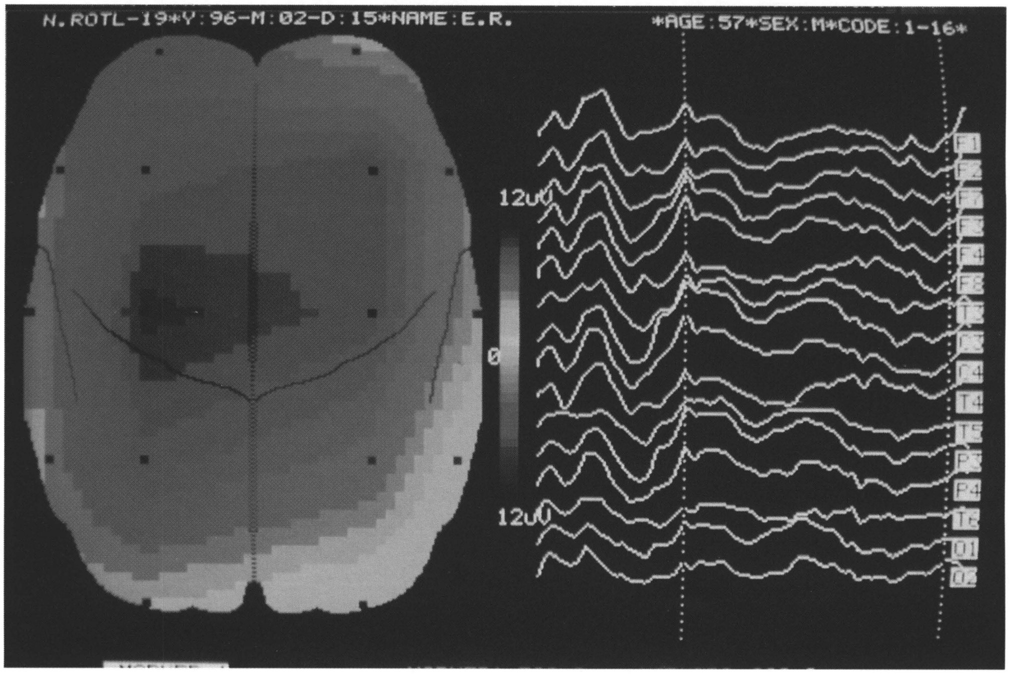


B. Counterclockwise (CCW) rotation

Figure 10. Case 2, follow up examination - VestEP elicited by **A.** CW-rotation and **B.** CCW-rotation.



A. Clockwise (CW) rotation



B. Counterclockwise (CCW) rotation

Figure 11. Case 2, follow up examination - Brain Electrical Activity Mapping of the VestEP in **A.** CW-rotation; **B.** CCW-rotation.

DISCUSSION

A cortical VestEP represents a specific answer/response of certain cortical areas to the vestibular stimulation. The *sine qua non* condition for the generation of normal Vestibular Evoked Potentials is the morpho-functional integrity of the vestibular system, including receptors, brain stem pathways and cortical centers. Partial lesions result in pathologic alterations of VestEP parameters. The more severe and extended the lesions are, the more disturbed will be the recorded VestEP.

The two presented cases were investigated in our department for similar presumptive diagnoses (Complete bilateral vestibular loss), based mainly on the results of the ENG, i.e., no nystagmus reaction to either calorization or rotation.

In the first case, where the symptomatology is dominated by the severe disequilibrium complaints, this diagnosis is confirmed by the VestEP-BEAM investigation. Namely, no cortical response/potential to the vestibular stimulation could be recorded, because of the complete bilateral destruction of the labyrinth receptors. We found extremely interesting the manner in which the complete absence of the vestibular function, as well as the very severe affection of the auditory function, were compensated by the visual and proprioceptive functions, the patient being able to lead a relatively normal life, with a satisfactory degree of social independency.

In the second case, any symptoms suggesting an affection of the vestibular system are absent. The only complaints were bilateral high frequency hearing loss and subjective tinnitus. This case is more complex and thanks to the information supplied by the VestEP-BEAM method, we could make some suppositions about the location and the mechanisms of the neurootological disorders and recommend a correct therapy. Thus, the results of the complete neurootological examination demonstrate that both the acoustic and vestibular systems are affected by the pathological process. Moreover, we can affirm that the disturbances are located not only in brain stem (ENG, BERA), but especially at the receptors (CCG, VestEP/BEAM). Therefore we can speak about a combined acoustic and vestibular, peripheral and central neuro-otological syndrome. It most likely reflects a vascular insufficiency within the vertebro-basilar system due to possible micro-tromboembolization, rheological changes, dynamic circulatory insufficiency, more pronounced on the left side. Sensory cochlear hair cells and vestibular receptors are especially affected by hypoxia, due to the terminal type of arterial circulation of the labyrinth. However, the brain stem structures, i.e., the mesencephalic nystagmus generator center, as well as the acoustical pathways, are affected too. These suppositions are also stressed by the positive effect of the treatment with betahistine.

CONCLUSIONS

The method of VestEP displayed by BEAM is considered to be a modern and very complex investigative tool in neurootology, which permits:

- Functional, topographic and differential diagnosis for balance disorders and also auditory disorders including tinnitus, due to the intimate anatomico-physiological relationship between the vestibular and acoustical end-organs.
- Follow-up and monitoring of the clinical progression of the cochleo-vestibular complaints.
- Objective evaluation of the efficacy of the therapy.

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