Clinical note

AUDITORY BRAIN STEM AND MIDDLE LATENCY RESPONSES IN A PATIENT WITH CORTICAL DEAFNESS

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Auditory stimuli with abrupt onset elicit a series of scalp-recorded evoked potentials. According to their latency these potentials are classified as early (0—10 msec), middle (10—50 msec) and late (50—500 msec) components (Picton et al. 1974; Davis 1976). Early responses are generated by the activation of the auditory brain stem pathways (Jewett et al. 1970; Starr and Hamilton 1976) and are known as auditory brain stem responses (ABRs). There is little agreement as to the origin of the middle and late responses. Although cerebral cortex and thalamus have been suggested as possible generator sources for the middle latency responses (MLRs), the issue is still open to question (Geisler et al. 1958; Picton et al. 1974; Goff et al. 1977; Knight et al. 1980; Mendel 1980).

The recent success of ABR in clinical electrophysiology and audiology has spurred renewed interest in middle latency responses. Currently MLRs have been shown to be stable in normal resting subjects and free of muscle activity (Mendel and Hosick 1975; Kraus et al. 1980). The extensive normative data gathered in our laboratory (Özdamar and Kraus 1981) have confirmed that MLR component Pa is stable in every adult individual tested. The use of MLRs as a clinical tool is appealing as no active participation from the subject is required and the responses are unaffected by light sedation. Thus MLRs may provide a convenient measure for clinical testing of cerebral and possibly thalamic function.

Research on the origin of the middle latency responses has centered on normal subjects for the purpose of evaluating hearing sensitivity (e.g., Mendel 1980). Little or no investigation on the sources of these responses has been done on patients with cerebral disorders. A very recent report studied a patient with bilateral temporal lobe lesions and reported normal MLRs (Parving et al. 1980). Patients with such lesions offer a rare opportunity to study the origin of these responses since bilateral representation in the auditory central nervous system is a general rule.

The present work reports auditory brain stem and middle latency responses in a patient with bilateral temporal lobe lesions and a unique case of cortical deafness. The rare patients with bilateral lesions have been reported to exhibit disorders ranging from cortical deafness to auditory agnosia and pure word deafness (Jerger et al. 1969, 1973; Kanshepolsky et al. 1973; Adams et al. 1977; Goldstein et al. 1977; Leicester 1980; Parving et al. 1980). In the present work a detailed behavioral evaluation of the patient is reported so that comparison with such studies is possible. Roentgenological findings are provided to document the lesion sites.

Case history and behavioral evaluations

Case history

The patient is a right-handed, 36-year-old white female admitted to the hospital with the 'worst headache' of her life. She was previously in good health and denied any history of hypertension, loss of consciousness, or seizures. However, she had suffered episodic headaches for much of her adult life. On admission the patient was alert and oriented to time and place. Short-term memory and fund of knowledge were good. Her speech was fluent and there was no evidence of paraphasia or oral motor apraxia. She did complain of a 'plugged' sensation in her ears and of slight difficulty in hearing. Arteriograms showed an aneurysm involving the left middle cerebral artery. A diagnosis of subarachnoid hemorrhage due to a ruptured left middle cerebral artery aneurysm was made. A clipping of the aneurysm was done 10 days later. Postoperative computerized tomography (CT) findings showed infarction of the left temporal and parietal lobes.

The patient improved rapidly until 2 weeks later when she had another subarachnoid hemorrhage. CT scan showed a hematoma about the right sylvian fissure and parietal lobe. Forty-eight hours later she developed adult respiratory distress syndrome and

was intubated. Her subsequent course was complicated by hypoxia secondary to plugging of the endotracheal tube. The patient was comatose for about 1 month. In the middle of this period, a repeat CT scan showed dilatation of the ventricles and a ventriculo-peritoneal shunt was placed, resulting in improvement of her level of consciousness. A right carotid arteriogram showed an aneurysm of the right middle cerebral artery that had not been present on the initial arteriogram. Surgery was not done at this time.

When the patient recovered consciousness, she did not respond to speech, non-speech sounds or music. Comprehension of gestures was fairly good although impaired. She also had severe dyslexia and dysgraphia. However, she spoke in grammatically correct sentences. Vocalization was impaired due to incomplete adduction of the vocal folds and she had some trouble with drooling and swallowing. The patient was discharged from the hospital after a total stay of about 3 months. At discharge she was alert and could walk. She complained of total deafness, right-sided weakness, altered smell and temperature sensation.

Sites of lesion

During her hospitalization the patient had several CT scans and arteriograms. They were consistent with the clinical picture of bilateral lesions of the temporal lobes. CT scans showed hematomas and infarcts in the right and left temporal lobes subsequent to ruptured aneurysms. A reconstruction of the infarcts taken from the CT scans 2.5 months after the second subarachnoid hemorrhage is shown in Fig. 1. Diagrammatic sections were prepared utilizing the lesion localization study of Gado et al. (1979). Interpretation and reconstruction of the lesions were made by a neurologist who did not know the patient or outcome of this study. Analysis of the reconstruction using the templates of Gado et al. (1979) suggests complete bilateral destruction of auditory areas 41 and 42 and possible or incomplete lesions in association area 22. The lesion on the left side extended lower in the brain (in the infraventricular region) than the lesion on the right.

Convalescent course

The patient was evaluated on 3 separate occasions over a 7 month period. On each of these occasions, a behavioral assessment of the patient's hearing, communication, and other higher cortical functions was performed, and auditory brain stem and middle latency responses were obtained.

The patient resumed many of her previous homemaking activities. She initially reported auditory sensations of buzzing or roaring but was unable to hear the speech of others or environmental sounds,

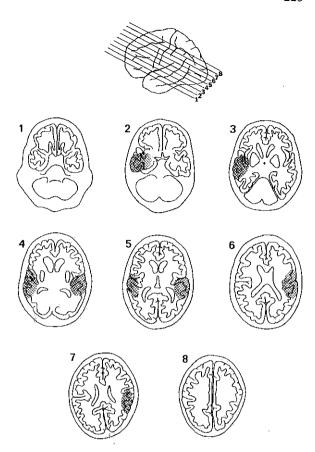


Fig. 1. Reconstruction of the lesion based on CT scan of the patient taken 2.5 months after the accident. Cross-hatched areas in the left and the right temporal lobes represent areas of infarct.

although she felt she could hear herself speak. Family members reported she had occasional auditory hallucinations such as hearing a radio. During the seventh month after discharge she showed awareness and recognition of some sounds such as the telephone ringing, although reporting these as faint and distorted. Senses of taste and smell were diminished, but not absent. She denied being able to feel light touch on any part of her body. She was unable to distinguish hot and cold and denied any sensation of pain throughout her body. Eight months after discharge she began to have some awareness of pain as when pinched. Her emotional status was one of depression, lability, and low frustration tolerance which improved somewhat during the convalescence.

The patient was seen by an otolaryngologist on several occasions. Otologic findings were negative, with the exception of lack of response to auditory 226 Ö. ÖZDAMAR ET AL.

stimuli. A unilateral vocal cord paralysis was diagnosed, thought to be secondary to prolonged tracheal intubation.

Behavioral assessment

Hearing. All audiometric tests were carried out in a double walled test booth. At the time of the first testing (2 months after discharge) there was no response to acoustic stimuli. No startle response could be elicited. She did not respond to pure tone stimuli and reported no vibration sensation of low frequency tones with the bone vibrator placed anywhere on her head. She did, however, report vibratory sensation with the vibrator on her hand. Impedance audiometry revealed normal tympanic membrane mobility bilaterally. Both ipsilateral and contralateral acoustic reflexes were elicited at normal reflex thresholds and there was no reflex decay.

Audiometric testing at 5 months post-discharge showed no change. Prior to the third testing she had begun to show some response to high intensity sounds such as door slams and telephone ringing. The audiometric test results at 7 months post-discharge are presented in the lower right corner of Fig. 3. At this time she responded to all frequencies tested, indicating a severe hearing loss bilaterally. Similar responses were obtained to bone conduction signals.

Oral language and reading comprehension. The patient showed no capacity to understand spoken language unless she received visual cues. Initially she made little use of lip reading, but by the seventh month post-discharge she showed skill and reliance on lip reading.

Reading constituted the patient's major means of understanding others. Word-to-picture matching was good for common objects, actions and body parts. She showed good reading comprehension of sentences, but comprehension of adult level paragraphs was at the 40–50% level. Reading comprehension decreased as the material increased in length, or became more abstract.

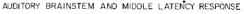
Oral expressive and written language. The patient was able to verbally express feelings, comment, ask questions, and respond to written questions. Her spoken language was grammatically and syntactically intact. She had no difficulty naming common objects, body parts, actions, numbers, or letters. There were occasional word retrieval problems, as well as literal and verbal paraphasic responses, although these were not predominant characteristics of her spoken language.

Copying of printed sentences was good and writing of overlearned material was unimpaired. She would not write elaborated descriptive paragraphs. There were more paraphrasic responses in her written than in her spoken language.

Speech production and oral motor skills. The patient's voice was weak, breathy, and hoarse. There was fading at the ends of phrases and sentences and some speech was produced during inhalation. Prosody was characterized by narrowed intonation and stress contours, with occasional exaggerations particularly when upset. Loudness was poorly modulated. Resonance balance was mildly hypernasal. Fluency of speech was good, but rate of production was reduced. Vowels were prolonged and consonants were imprecise.

There was no buco-facial apraxia. The patient was able to protrude, elevate and lateralize the tongue. There was slight deviation of the tongue to the right side at rest. The velum contracted bilaterally and the uvula was midline. Diadochokinetic rates were reduced. Her cough was weak and breathy. She had no chewing or swallowing problem and no nasal regurgitation of liquids.

Miscellaneous. The patient was able to pantomime serial actions. She demonstrated good design copying skills. Visual perception and abstract reasoning was grossly intact (Raven 1938, 38th percentile). She was able to recognize common objects in either hand with eyes closed. Left-right discrimination was good in relation to her own body and externally. Clock setting and time telling were intact, as was finger recognition.



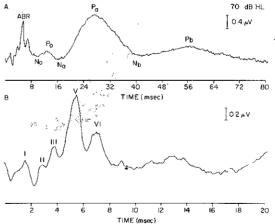


Fig. 2. Auditory brain stem and middle latency responses obtained from a normal female subject at 70 dB HL click intensity. A: recording of 80 msec poststimulus onset showing ABR and MLR components. B: initial 20 msec expanded from trace A, showing ABR components in detail.

Electrophysiological testing

Methods

Electrophysiological testing was carried out with the patient sleeping or resting quietly on a bed in an electrically and acoustically shielded room. She was sedated with chloral hydrate (500 mg, p.o.). Stimuli were rarefaction clicks presented monaurally at a rate of 10/sec. Click hearing level (HL) was referenced to average thresholds obtained from a group of nor-

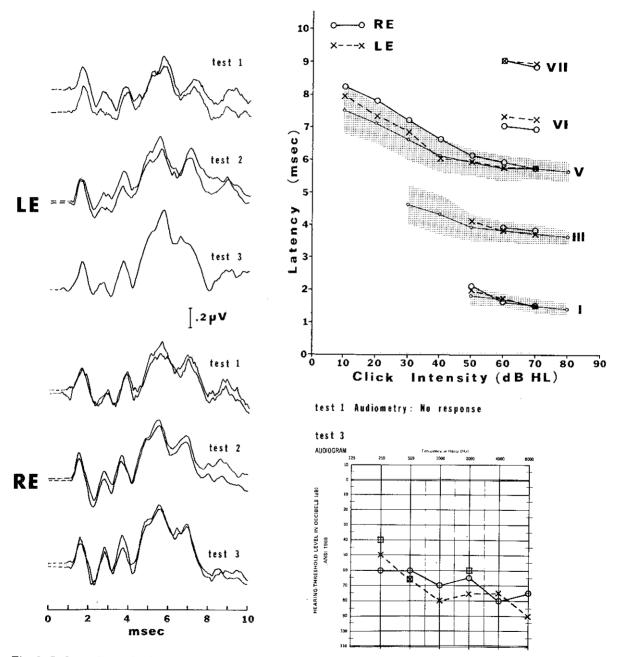


Fig. 3. Left: auditory brain stem responses obtained from each ear on 3 different test dates. Upper right: latency-intensity plots of major ABR components obtained on test date 1. Dotted areas denote two standard deviations above and below means derived from normal subjects. Lower right: audiogram obtained at the time of test 3.

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mal hearing subjects (Özdamar and Stein 1981). Power spectrum characteristics of the click were the same as described in that study.

Auditory brain stem and middle latency responses were recorded simultaneously as described in other reports (Kraus et al. 1980; Özdamar and Kraus 1981). Briefly, they were obtained by averaging a differentially recorded EEG signal (G1: vertex; G2: ipsilateral mastoid; Ground: forehead). The low- and high-pass filters were set at 3 and 2000 Hz respectively. The computer was triggered by the leading edge of the electrical click signal and 80 msec of poststimulus time was examined using 20 µsec sampling bins. At least 2 tracings, consisting of 1024 click presentations each, were obtained for each stimulus condition.

In Fig. 2A, a typical ABR and MLR recording from a normal subject is presented for comparison purposes. The subject is a 27-year-old, right-handed female with no prior history of otologic or neurologic disorder. The first 20 msec of the response is expanded in Fig. 2B for easy identification of the ABR waves.

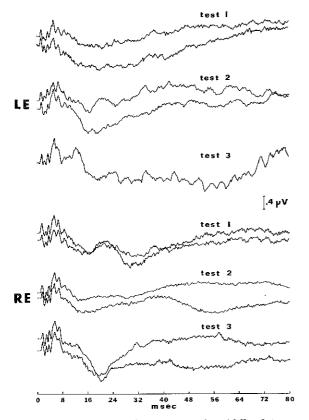


Fig. 4. Auditory brain stem and middle latency recordings obtained on 3 different test dates.

Results

Normal ABRs were elicited bilaterally at all click intensities on all 3 test dates. Responses obtained at 70 dB HL in both ears are presented in Fig. 3A. All ABR components (waves I through VII) were recorded with normal amplitude and latency, indicating integrity of the cochlear mechanism and auditory brain stem pathways. Latency-intensity plots for all waves were within normal range as shown in Fig. 3B. Wave V was measured down to 10 dB HL bilaterally, indicating normal hearing sensitivity by ABR

MRL component P_a , on the other hand, was consistently absent to both right and left ear stimulations as illustrated in Fig. 4. Beyond the brain stem potentials, there was no evoked potential activity to left ear stimulation. Right ear stimulation elicited a small positive response at 20 msec on test date 1. This deflection disappeared on test date 2 and on test date 3 a negativity appeared at about the same time. The absence of the P_a component is obvious when these recordings are compared with the MLRs obtained from a normal subject shown in Fig. 2.

Discussion

Although MLRs were recorded many years ago (Geisler et al. 1958), presently little is known about them, particularly in comparison to early and late auditory evoked potentials. The early controversy regarding the myogenic vs. neurogenic origin (Bickford et al. 1964; Cody et al. 1964) is largely resolved today. The component occurring approximately 15 msec poststimulus onset has been shown to be myogenic, arising from the posterior auricular muscles (Picton et al. 1974; Goff et al. 1977; Streletz et al. 1977). The positive component P_a occurring at about 30 msec is neurogenic, as it persists under sedation and total muscular paralysis (Mendel and Hosick 1975; Harker et al. 1977).

The controversy regarding the origin of P_a continues today. The work of Geisler et al. (1958) and Piction et al. (1974) points to primary auditory cortex and/or thalamus as neural sources for MLRs while this contention is questioned by Goff et al. (1977). Animal research has implicated primary auditory cortex as the generator site for the P_a analogue measured in the cat (Kaga et al. 1980) although this has also been contested (Buchwald et al. 1981).

Recently this issue has been addressed by recording MLRs in a patient with bilateral temporal lobe lesions (Parving et al. 1980). This study, which is most relevant to our findings, reported intact MLRs. Whereas anatomically the patient reported by Parving

et al. (1980) and our patient appear to have similar lesions, they are behaviorally quite different. Parving et al.'s case had essentially normal hearing sensitivity bilaterally whereas our patient initially showed no behavioral response to sound despite persistent efforts. Although some auditory sensitivity was regained by the third test date, pure tone thresholds were indicative of a severe hearing loss bilaterally. The term cortical deafness best describes the patient's marked impairment in hearing sensitivity in spite of the apparent intactness of the auditory periphery and auditory brain stem pathways. The patient of Parving et al. displayed an auditory agnosia in that although hearing sensitivity was intact, he was unable to interpret the meaning of acoustic stimuli, including his own speech. It is curious that our patient reported auditory perception of her own speech. This suggests that the auditory sensation of one's speech may be based a great deal on kinesthetic feedback. The apparent behavioral differences between the two patients suggest that the lesions are not the same.

Previous studies of patients with bilateral temporal lobe lesions reported that the patients retained surprisingly good hearing. In some cases deafness was temporary, improving in a couple of weeks. When hearing loss persisted, it was a mild to moderate loss affecting the middle and high frequencies (Jerger et al. 1969, 1973; Kanshepolsky et al. 1973; Leicester 1980; Parving et al. 1980). This is an unusual case of cortical deafness following bilateral temporal lesions in which the patient behaved as though functionally deaf or severely hearing impaired. On the other hand, her auditory brain stem responses attest to the integrity of the cochlear mechanism and auditory brain stem pathways. As cases of bilateral temporal lobe lesions are rare, it is likely that no two cases had the same site of lesion. The behavioral discrepancy between our case and previous cases may be due to the lesion sites specific to the patients.

In addition to being a novelty in the human literature, the patient's deafness is also at odds with animal lesion studies. These have shown that animals of numerous species can detect the presence of acoustic stimuli following complete bilateral ablation of the cerebral cortex (Neff et al. 1975). Behavioral intensity threshold and frequency discrimination are unimpaired by such lesions. This may then reflect a species difference although one cannot rule out the possibility that the patient is psychologically denying acoustic stimuli.

Our findings support the notion that MLRs are generated by auditory cortex (primary and/or secondary) and possibly the radiations from thalamus. The delineation of MLR generator sites cannot be based solely on CT scan data as discrete anatomical localization is limited and is further hampered by the anatomical variations of the cerebral cortex among

individuals of the same species. It appears that these auditory areas may be responsible for conscious perception of sound and that total bilateral destruction of these areas results in complete deafness. MLRs may reflect the bioelectric substrate underlying auditory stimulus detection. The combination of behavioral, anatomical and electrophysiological data promises to be an effective strategy for reaching a better understanding of neural generator sites for MLRs and auditory processing in the brain.

Summary

Auditory brain stem (ABR) and middle latency responses (MLR) were recorded in a patient with bilateral temporal lobe lesions. Audiological and higher cortical functions were assessed using conventional behavioral methods. Roentgenological findings were presented for localizing the lesions.

Initially the patient showed no behavioral response to sound. Subsequently the patient reported inconsistent awareness of environmental sounds and pure tone sensitivity was impaired to a severe degree. Higher cortical function was essentially intact and the patient was not aphasic. ABR and acoustic reflex findings were consistent with normal functioning of the auditory periphery and brain stem pathways. MLR component P_a was absent bilaterally.

These findings suggest that MLR component P_a is bilaterally generated in the temporal lobes. Auditory cortex appears to play a role in auditory sensitivity in humans.

Résumé

Réponse auditive du tronc cérébral et de latence moyenne chez un patient avec surdité corticale

Les réponses auditives du tronc cérébral et les réponses de latence moyenne ont été enregistrées chez un malade avec lésions bilatérales du lobe temporal. Les fonctions audiologiques et corticales supérieures on été mesurées à l'aide des méthodes comportementales conventionnelles. Les données radiologiques permettent la localisation des lésions.

Initialement, le malade ne montre aucune réponse comportementale au bruit. Ultérieurement, le malade fait mention d'une conscience inconstante des bruits environnants et la sensibilité aux sons purs est fortement atteinte. Les fonctions corticales supérieures sont dans l'ensemble intactes et le malade n'est pas aphasique. Les données acoustique réflexes et les réponses auditives du tronc cérébral confirment le fonctionnement normal de l'audition périphérique et des voies du tronc cérébral. La composante Pa de

la réponse de latence moyenne est absente des deux côtés.

Ces données suggèrent que la composante P_a de la réponse de latence moyenne prend son origine de façon bilatérale dans les lobes temporaux. Le cortex auditif joue un rôle dans la sensibilité auditive chez l'homme.

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