

Kraus

ABSENT AUDITORY BRAIN STEM RESPONSE: PERIPHERAL HEARING LOSS OR BRAIN STEM DYSFUNCTION?*

NINA KRAUS, Ph.D.,
ÖZCAN ÖZDAMAR, Ph.D.,
NANCY REED, M.A.,
Chicago, IL.

LASZLO STEIN, Ph.D.,

ABSTRACT.

Interpretation of auditory brain stem response (ABR) findings can be problematic in cases where waves III and V are absent. Such findings can be attributed to profound hearing loss, brain stem neuropathology, or both. Over a 3-year period, 48 patients with no known brain stem damage and on whom audiologic data were available were found to have no response by ABR or absent waves III and V. Severe to profound hearing loss was documented in 38 cases, audiometric data were equivocal in 3 cases, and 7 patients showed pure tone sensitivity ranging from normal hearing to moderate impairment. Thus 15% had better hearing sensitivity than might have been expected from their ABR findings. Each of these patients also exhibited abnormal acoustic reflex findings. We report the electrophysiological (ABR, MLR, acoustic reflex), medical (history, neurological, EEG, CT scan) and behavioral (audiometric, speech and language, learning disabilities, psychological) data which characterize this group of patients.

Auditory brain stem response (ABR) is now widely used as a test of auditory sensitivity and neurological status of the auditory brain stem pathways.¹⁻⁵ When there is no indication of brain stem neuropathology, ABR and audiological findings are generally in agreement.⁶ Yet there have been reports of exceptions. Davis and Hirsh⁷ mention instances of absent ABRs in children who behaviorally responded to moderate or low intensity sounds. More recently, Worthington and Peters⁸ published 4 case reports of patients with absent ABRs and no worse than severe hearing loss. Finally, Lenhardt⁹ reported a case in which only wave I was obtained from a patient with normal pure tone hearing. Although Schulman-Galambos and Galambos¹⁰ and Galambos¹¹ warn that such findings may be due to "technical error," these reports point to the need for re-examining and further delineating cases where ABR and audiological findings appear "paradoxical."

A further issue concerns the possible relationship between such ABR results and other clinical findings, particularly with regard to communication skills. The reports of both Worthington and Peters⁸ and Davis and Hirsh⁷ mention the presence of language disorders in their cases. If such an association exists, ABR might assist in the identification of a subpopulation of patients with communicative disorders.

Over the course of 3 years, we have accumulated data on 48 patients who: 1. had absent ABRs or no detectable waves III or V on repeat testing, 2. had no clinical signs of brain stem neuropathology, and

3. were tested by behavioral audiometry. Of the 48, 7 had audiometric findings ranging from normal hearing to no worse than a moderate hearing loss. We report electrophysiological, medical, and behavioral data pertaining to these 7 patients.

Three illustrative case histories are presented. Repeated ABR measures on separate occasions by different clinicians and manipulation of stimulus and recording variables were done to minimize the possibility of "technical error." Middle latency responses (MLRs) and acoustic impedance measures were employed as additional measures of the physiological status of the auditory pathways. Incidence and associated medical risk factors are presented. Behavioral data from audiological speech and language, learning disabilities, and psychological evaluations were examined in an attempt to define commonalities characterizing these patients.

The present study addresses 1. the issue of absent ABRs or absent waves III and V in the diagnosis of hearing loss and 2. the delineation of the clinical picture which tends to accompany such cases.

METHODS.

Electrophysiology.

Patients were sedated with chloral hydrate (25 mg/kg) and testing was conducted with the patient lying inside an electrical and acoustically shielded room. Rarefaction clicks were generated by rectangular wave pulses (0.1 msec) delivered monaurally to TDH-39 earphones mounted in MX 41/AR cushions at a rate of 20/sec for ABR and 10/sec for MLR recordings. Condensation clicks were also used in cases where wave I only was elicited in order to assess the contribution of the stimulus artifact to the response. Click hearing level (HL) was referred to average thresholds from a group of normal hearing subjects.¹² The peak equivalent sound pressure level re: 0.0002 dynes/cm² of the click stimulus was 32 dB SPL (reference 3 kHz).

Auditory brain stem and middle latency responses were recorded as previously described.^{13,14} A differentially recorded EEG signal (G1: vertex; G2: ipsilateral mastoid; Ground: forehead) was averaged based on 2048 stimulus presentations for

*From the Siegel Institute, Electrophysiology Laboratory, Michael Reese Hospital and Medical Center and Department of Surgery (Otolaryngology), University of Chicago.

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Send Reprint Requests to Nina Kraus, Ph.D., Electrophysiology Laboratory, Siegel Institute, Michael Reese Hospital and Medical Center, 3033 South Cottage Grove Ave., Chicago, IL 60616.

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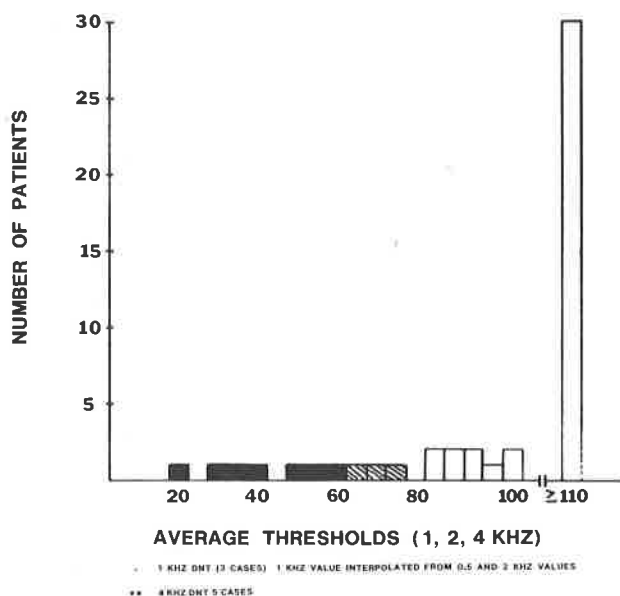


Fig. 1. Average of 1, 2 and 4 kHz behavioral threshold values for all patients (n=48). Shaded areas represent patients with the distinctive ABR/audiometric findings which are the subject of this paper. Cross-hatched areas represent "equivocal" cases.

ABR and 1024 for MLR. Low pass filters were at 100 Hz for ABR and at 3 and 10 Hz for MLR with the high pass filter at 2 kHz. The computer was triggered by the leading edge of the electrical click signal and 20 μ sec sampling bins were used. Twenty and 80 msec of post stimulus time were recorded for ABR and MLR respectively. Sound intensity was incremented from threshold (the lowest intensity at which sound evoked bioelectric activity was obtained), in 10 dB steps to a maximum intensity of 90 dB HL. At least two recordings were obtained for each stimulus condition.

Acoustic impedance measures were obtained with an Amplaid 702 Impedance Meter. Acoustic reflexes were elicited with ipsilateral and contralateral reflex signal presented to each ear at 0.5, 1, 2 and 4 kHz. These measures were documented by the graphic output of an X-Y plotter. Reflexes were considered abnormal if reflex thresholds were absent or elicited at or above 100 dB (HL) in the presence of normal tympanograms.

Audiologic and Other Clinical Evaluations.

Audiologic follow-up was available in 48 patients with no ABR response or absent waves III and V. Pure-tone audiograms were obtained under earphones in 17 cases. Visual response audiometry (VRA) in sound field was used with 31 patients. Therefore, information in the better ear only could be obtained in these cases.

Testing of the 7 patients who did not exhibit severe to profound hearing loss was more extensive. Pure tone audiograms were obtained on at least two separate occasions for 6 patients. VRA in sound field was necessary in one instance. Speech discrimination testing was done in 5 cases.

All of the 7 patients had neurological examinations, 5 had EEG testing and 3 had CT scans. The older patients were evaluated by Speech Pathology, Learning Disabilities, Psychological and Psychiatric services within the Institute. The speech examination evaluated language and articulation skills and structure and function of the oral mechanism. Testing done by Learning Disabilities focused on the assessment of school task performance and specific testing of the auditory and visual modalities. IQ testing was performed during the psychological evaluation as was assessment of the patients' emotional status.

RESULTS.

Over a 3 year period, 543 patients with no clinical indication of brain stem damage were tested by ABR for the assessment of hearing. Of these, 48 showed no ABR response or absent waves III and V. Audiometric information was available in all 48 cases. Severe to profound hearing loss was documented in 38 patients with no response by ABR. Three patients appeared to have hearing losses of moderate to severe degree, but these cases were regarded as "equivocal" due to the extent of the losses and because reliable audiometric thresholds could not be established with certainty. Of primary interest are the 7 patients with audiometric data clearly ranging from normal hearing to moderate impairment.

It is generally believed that ABR click threshold correlates best with high frequency hearing sensitivity.^{15,16} Figure 1 shows the average behavioral threshold values obtained at 1, 2 and 4 kHz for all 48 cases. Because ABR and audiometric findings were bilaterally symmetric and because audiologic information was frequently obtained in sound field, we plotted the threshold of the better ear. The shaded areas represent the thresholds of the 7 patients of primary concern to this study and the cross-hatched areas designate the "equivocal" cases.

Total absence of sound evoked, bioelectric activity was found in 3 of the 7 patients. Another 3 showed evidence of ABR wave I or waves I and II only. One patient showed no response in one ear and early waves only in the other. Together, these patients comprise 15% of patients without ABR waves III and V in whom absence of response was not attributable to severe to profound hearing loss.

In each patient tested (6 of the 7), acoustic reflexes were either absent entirely or were elicited at elevated thresholds. These findings could not be explained by pure tone thresholds. Middle latency responses were present in only 1 of the 10 ears tested.

The electrophysiological, medical, and behavioral findings characterizing the individual cases are summarized in Table I. Although the range of deficits varied considerably among these cases, there were many common features which are summarized in Table II. Eighty-six percent of the patients were male. Five patients (71%) had suffered a well defined clinical event such as perinatal asphyxia, hyperbilirubinemia or head injury, predisposing them to sensorineural hearing loss as well as to central nervous system damage. Neurologic examination was negative in 86% of the patients and in no instance revealed brain stem damage.

Patients old enough for formal testing performed below age level on the auditory subtests of the Illinois Test of Psycholinguistic Abilities (ITPA) and the Wepman Perceptual Test Battery (WPTB). These tests, which have parallel subtests for the assess-

TABLE I.
Summary of Individual Electrophysiological, Medical and Audiological Findings.

Identifying Case No.	Information Sex	Age at ABR	Electrophysiology			Birth History	Medical				Audiological	
			ABR	Acoustic Reflex	MLR		Medical History	Neuro Exam	EEG	CT	Pure Tone Sensitivity	Audiogram Configuration
1	M	6 yr, 5 mo; 7 yr	NR	NR except R contra elevated	NR	RDS	Concussion febrile convulsions	WNL	WNL	WNL	WNL bilat	
2	M	6 yr	NR	NR	DNT	28w GA 1250 gm RDS hyperbilirubinemia RLD	WNL	WNL	WNL	DNT	Mild-to-mod SN loss bilat	Flat
3	M	5 yr	R NR L early waves	NR	R NR L present	WNL	URI febrile convulsions seizure disorder	WNL	WNL	DNT	Mild-to-mod SN loss bilat	Flat
4	F	11 yr; 14 yr	Early waves	NR	NR	37w GA 2045 gm	Concussion	WNL	WNL	DNT	R mild-to-sev L mod SN loss	Sloping flat
5	M	5 yr, 10 mo; 6 yr; 2 mo	Early waves	NR	NR	WNL	Concussion pneumonia	Macrocephaly	WNL	4th ventricle enlarged	Mild-to-mod SN loss bilat	Flat
6	M	29 yr	NR	NR	NR	WNL	WNL	WNL	DNT	DNT	Mild SN loss bilat.	Flat
7	M	3 mo; 9 mo; 1 yr	Early waves	DNT	DNT	Hyperbilirubinemia	WNL	WNL	WNL	DNT	Mild SN loss bilat	Flat

NR - no response; Early waves - I or I and II; WNL - within normal limits; DNT - did not test; GA - gestational age; RDS - respiratory distress syndrome; SN - sensorineural; RLD - retrolental dysplasia; bilat. - bilaterally.

ment of auditory and visual skills revealed significantly worse performance in the auditory as compared to the visual tasks. Auditory memory performance was impaired in each case. Speech discrimination testing revealed worse scores than would have been predicted from the patients' hearing thresholds in 4 of 5 cases. Speech and language delays were present without exception although the older patients eventually spoke in grammatically intact sentences. In patients too young for formal testing, response to sound was characterized by highly variable and inconsistent responses on repeated audiological testing and were further substantiated by reports from other professionals (such as educators of the hearing impaired and occupational therapists). Three of 4 patients fitted with hearing aids reportedly derived little benefit from amplification although this was not formally tested. Formal intelligence testing was done in 5 cases. Four normal and 1 borderline-normal scores were obtained.

CASE HISTORIES.

Case 1.

This patient was born to an Rh- mother and sustained perinatal respiratory distress. Subsequent medical problems included hospitalizations were at 1½ and 2 years for upper respiratory infections, febrile convulsions, and a concussion at 3 years. He was evaluated at this center at 6 years of age for delayed speech development and equivocal responses to sound. The neurological exam, EEG and CT scan findings were negative. Psychological testing revealed a performance IQ of 111 and no overt emotional disturbance. Significant delays were evident in speech and language skills with performance at the 3 year, 9 month level. He spoke utilizing 3 and 4 word sentences. His age level scores ranged from 3 to 5 years on the auditory subportions of the ITPA while his visual task performance was above age

level (7 years). The discrepancy between auditory and visual modality function was further evidenced by his performance on the WPTB. Depressed processing of both verbal and non-verbal auditory stimuli was also documented by an auditory processing battery (Denver Auditory Phonemic Sequencing Test, Environmental Sounds Recognition Test, Competing Environmental Sounds Test and Test of Nonverbal Auditory Discrimination). Auditory memory and auditory sequencing skills were at the 4 year level (Auditory Memory Span for Digits; Spencer and McGrady Memory for Sentences).

Audiologically, the patient showed normal pure tone sensitivity bilaterally. Speech discrimination ability was tested using the Word Intelligibility by Picture Identification (WIPI) test and yielded fair scores of 72% and 76% in the right and left ears respectively at suprathreshold levels. Ipsilateral acoustic reflexes

TABLE II.
Summary of Common Electrophysiological, Medical, and Behavioral Findings.

	Percent	Number of Ears Tested
Electrophysiology		
ABR results:		
No response	50	14
Waves I, II only	50	14
MLR present	0.1	10
Acoustic reflex abnormalities	100	12
Medical		
Negative neurological exam	86	7
At risk for sensorineural hearing loss	71	7
At risk for central auditory deficits	71	7
Behavior		
Speech and language delays	100	5
Learning disabilities	100	4
Normal IQ	80	5
Sensorineural hearing loss	86	7
Auditory processing deficits:		
Formal testing	100	5

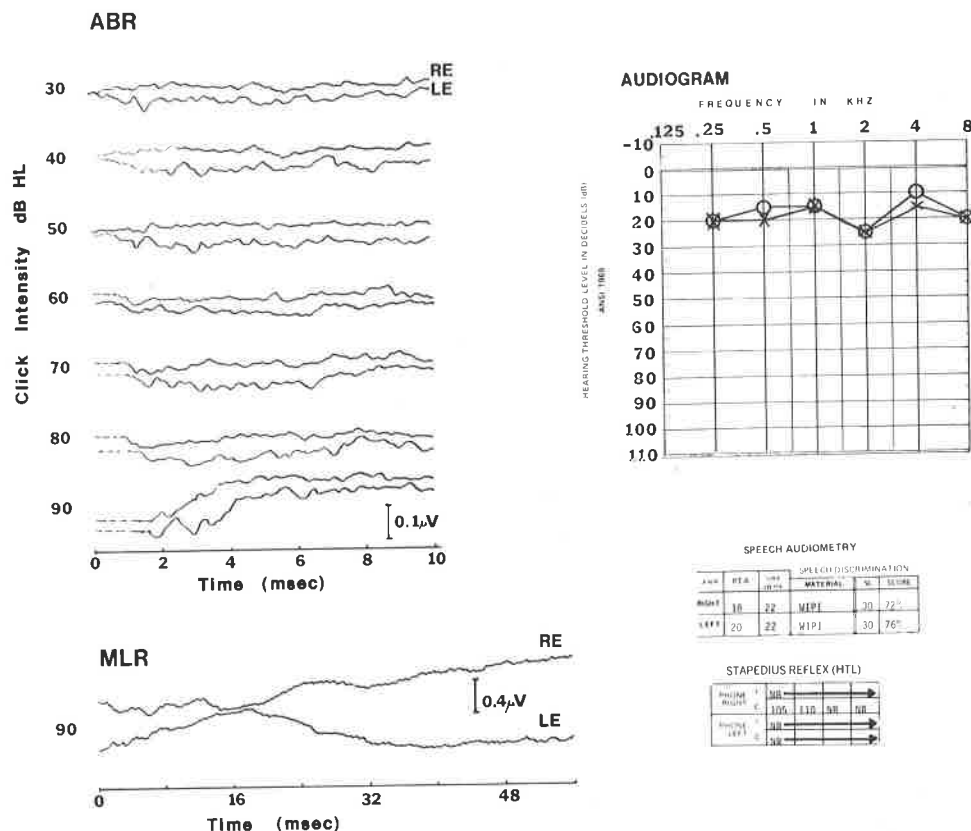


Fig. 2. ABR and audiometric results obtained from Patient 1.

were absent bilaterally as were left contralateral responses; only right contralateral responses were obtained at elevated SPL at 500 and 1,000 Hz. On two separate occasions, no ABR or MLR activity was obtained to stimuli as loud as 90 dB HL in both ears. These findings are shown in Figure 2.

Case 2.

This child was born prematurely with a birth weight of 2045 gm. She sustained a concussion at 2 years. Her evaluation at 8 years of age revealed normal neurological and EEG exams. She demonstrated a non-verbal IQ of 120 and no overt emotional disturbance. Significant receptive and expressive language delays were evident. She spoke in complete sentences, and made considerable use of lipreading. Severe auditory processing deficits and specific learning disabilities were evident. Her scores fell within the 2 to 5 year range on the auditory subtests of the ITPA, with performance at 5 to 6 year levels on the visual subtests. Poorer auditory modality performance was also evident on the WPTB. On a test requiring sentence repetition (Carrow Elicited Language Inventory), she scored at the 3½ year level, with errors characterized by omissions and substitutions of words.

In the right ear she exhibited a mild sensorineural hearing loss for low and middle frequencies with a moderate to severe loss in the high frequencies. The left ear showed a moderate sensorineural loss with a flat configuration. Speech discrimination scores were significantly depressed with scores of 20% and 16% in the right and left ear respectively. Incidentally, she showed no errors in articulation which might have been expected from her hearing loss. She made questionable use of her hearing aid and was perceived as functioning "neither as a deaf nor as a hearing child." Acoustic reflexes were absent bilaterally. ABR testing revealed the presence of atypical early waveforms up to 3.5 msec post stimulus time at high stimulus intensities only. Waves I and II were evident. Wave V was absent as were MLRs. These findings are shown in Figure 3.

Case 3.

This child had a normal birth history. He suffered a concussion at 3 years of age and was brought to this Institute at 5 years because of "difficulty understanding speech." Macrocephaly was noted on examination and CT scan revealed a prominent fourth ventricle with no dilatation of lateral or third ventricles. These findings were not thought to have clinical significance. The EEG exam was normal. Psychological testing revealed a normal IQ and "organic brain syndrome." He showed significant delays in receptive and expressive language although he spoke in 4 and 5 word sentences. Mild deficits were noted across stimulus modalities and his behavior was inconsistent on all diagnostic procedures. On the auditory subtests of the ITPA his scores fell within 2 to 4 year age levels and performance was poor on the auditory portions of the WPTB. He scored above age level on the visual subtests of the ITPA (6 years), demonstrating strong non-verbal thinking skills by his ability to analyze pictures. Auditory memory was at the 4 year level (Spencer Sentence Repetition Test) and he tended to remember the rhythmic and syllabic pattern of the sentences with omissions and substitutions of the verbal content.

His audiograms were inconsistent, with response discrepancies of 10-15 dB. He exhibited a moderate sensorineural loss in the right ear and mild-to-moderate loss in the left ear with flat configurations and made questionable use of his hearing aid. ABR testing revealed waves I and II with no subsequent waveforms as shown in Figure 4. MLRs and acoustic reflexes were absent.

DISCUSSION.

The incidence of absent/abnormal ABR in patients with relatively good hearing has been reported only once. According to Davis and Hirsh⁷ the incidence is 1 in 200 (0.5%). Specifics on these pa-

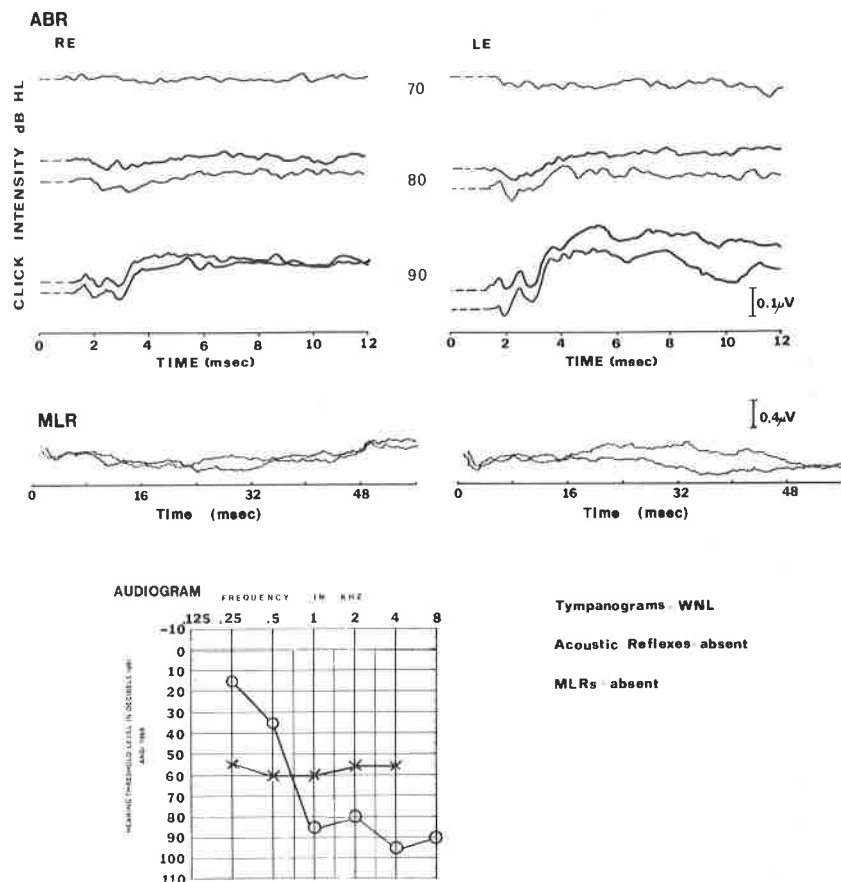


Fig. 3. ABR and audiometric results obtained from Patient 4.

tients were not further reported. Worthington and Peters⁸ have reported 4 cases, and Lenhardt⁹ reported 1 case in which only wave I was obtained. We found that 15% of patients without ABR waves III and V (and no clinical evidence of brain stem dysfunction) fell into this category. This figure represents 1.3% of the total population of 543 patients tested by ABR for the assessment of hearing. The incidence becomes 1.8% if the 3 "equivocal" cases are included.

No response by ABR is problematic because it is impossible to determine, from the ABR alone, whether the results are due to profound hearing loss or to brain stem damage. As audiometric findings were not consistent with profound degree of hearing loss in the 7 cases considered here, we suspect that absence of ABR response reflected neuropathology of the auditory brain stem. In the instances where wave I or waves I and II were obtained in the absence of later waveforms, the ABR itself was indicative of brain stem damage. These cases are complicated nevertheless because one cannot rule out concomitant hearing loss. Therefore, in instances where either no response or early waves only are obtained, ABR is insufficient as the sole evaluator of hearing sensitivity.

There are several physiologic factors which may

contribute to the difference between ABR and audiologic results observed in these cases. The risk factors for cochlear hearing loss (*e.g.*, perinatal asphyxia, hyperbilirubinemia and head trauma) are also those known to cause dysfunction of the central auditory pathways.

Perinatal asphyxia, which occurred in 50% of the patients in the present study, is a well documented cause of sensorineural hearing loss.¹⁷⁻¹⁹ Leech and Arnold²⁰ reported that loss of inferior colliculus neurons with gliosis or ischemic cell changes occurred in 85% of perinatal asphyxia cases. Stockard and Stockard²² have reported the loss of later ABR components (beyond wave II) following perinatal anoxia. Cochlear and auditory brain stem pathology have been linked to anoxia in humans.¹⁹ That brain stem structures are vulnerable to anoxia has been shown in primates²¹ and guinea pigs.²²

A history of hyperbilirubinemia was present in 40% of the patients presently reported. In addition to causing peripheral hearing loss, hyperbilirubinemia has been shown to lesion the auditory brain stem, specifically, the cochlear nucleus.²³⁻²⁷ Disorders of central auditory processing have also been reported in the literature.^{22, 28, 29}

Head trauma has been shown to cause both peripheral auditory and brain stem dysfunction.^{30, 31}

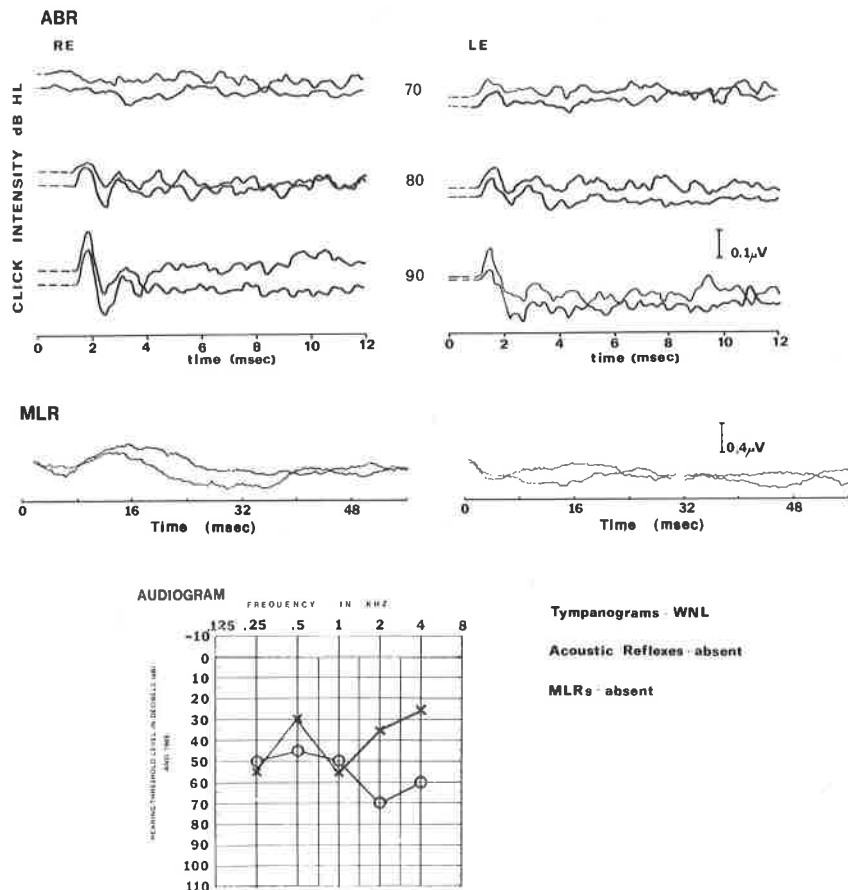


Fig. 4. ABR and audiometric data obtained from Patient 5.

One third of the patients in the present investigation had histories of head injury. Elevated evoked response thresholds have been measured from the inferior colliculus concurrent with normal cochlear potentials (CM) in guinea pigs following head trauma.³² Evidence of unilateral brain stem pathology subsequent to head injury has been documented by ABR in humans.¹¹

With the assumption that both cochlear and brain stem function may be damaged in these patients, the difference between ABR and audiometric test results can be (hypothetically) explained. First, the inferior colliculus has been implicated as a generator site for wave V,³³ which generally correlates best with behavioral hearing thresholds. If the auditory midbrain is particularly sensitive to factors such as asphyxia or head trauma, then it is not surprising that ABR wave V is absent in these cases. Second, waves I and II cannot be observed at low stimulus intensities even in normal subjects. If a concomitant hearing loss exists, one would expect to observe these early waves at intensities higher than behavioral thresholds, if at all.

Absent ABRs or the selective loss of the later waveforms may result from disruption of the neural synchrony necessary for ABR generation. The aforementioned insults to the nervous system may

have resulted from myelination deficits or destruction of cell bodies. Although essential for ABR generation, neural synchrony may not be necessary for stimulus perception. It is also possible that pathways responsible for ABR may be damaged while hearing is preserved through other pathways. The fact that MLRs were observed in one subject is an indication that neural signals are indeed reaching auditory pathways central to the brain stem. Therefore, some form of auditory function exists as is evidenced by audiometric thresholds and understanding for speech.

The present study should only help reinforce the concomitant use of ABR and behavioral audiometry. Diagnosis of hearing loss using ABR must be made with some restraint and with close audiologic follow-up in order to guard against inappropriate interpretation in cases such as these. Electrocochleography may be useful for the assessment of auditory sensitivity in these patients.

The combination of ABR and audiology in children who display this apparent incompatibility in test results may become useful for identifying a subgroup of patients with communicative and/or learning disorders which may result from brain stem dysfunction. That the patients in the present work evidenced deficits in auditory processing raises the

possibility that these problems may be directly associated with dysfunction of the auditory brain stem as measured by ABR. ABR and acoustic reflex findings can demonstrate an organic basis for auditory processing or communicative problems which otherwise could not be explained by peripheral hearing loss alone. Children with these distinctive test findings, then, may require a specific course of management to compensate for dual cochlear and auditory brain stem dysfunction.

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