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Dilemmas in auditory assessment of developmentally retarded children using behavioural observation audiometry and brain stem evoked response audiometry

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Abstract

The records of 94 consecutive developmentally retarded children with speech retardation and suspected hearing loss who underwent auditory assessment by both conventional behavioural observation audiometry (BOA) and brain stem evoked response audiometry (BERA) were analysed. In 54 children (57.4 per cent) there was good agreement between the results of both techniques leading to a clearcut diagnosis. In 22 children a diagnosis was possible only by the results of BERA as the results of BOA were inconclusive. Of the remaining 18 children, two groups could be identified whose results posed a dilemma. Group 1 (n = 7) consisted of children whose BOA test results differed considerably from their BERA results. Group 2 (n = 11) consisted of children in whom there was no discernible response by BERA while the response by BOA was either inconsistent (n = 5) or not elicitable (n = 6). The specific strategies to be adopted for hearing assessment in these situations are discussed.

Key words: Audiometry; Audiometry, evoked response; Child development disorders

Introduction

The diagnostic evaluation of hearing impairment in developmentally retarded children involves the triad of identification of hearing loss, determination of auditory thresholds and localization of the site of the lesion. Early identification of hearing handicap enables correction with suitable amplification. Conversely, audiological documentation of normal peripheral hearing suggests central causes for speech retardation, which may then be investigated in detail.

Currently available auditory testing methods include both behavioural and non-behavioural or objective tests of hearing. Although widely used and fairly reliable in normal children, behavioural tests have their limitations when administered to a developmentally retarded paediatric population. Appropriate examiner skills in handling children and a good sense of timing are imperative. Despite the best of intentions, efforts at determining responses using these tests are frustrated by factors like lack of cooperation and poor psychomotor skills in the child being tested. Further, behavioural tests, while measuring the lowest stimulus intensity to which response occurs, often do not provide an accurate picture of auditory threshold (McCormick, 1994).

During the past two decades several objective tests, including electrophysiological tests, like brain stem response audiometry (BERA), have evolved and become part of the audiological armamentarium. BERA has proved itself to be particularly useful in paediatric auditory testing (Hecox and Galambos, 1974; Mokotoff *et al.*, 1977). Reliability and accuracy in the determination of auditory thresholds, as well as the fact that the responses are unaffected by sedation, make BERA the audiological test of choice in developmentally retarded children. BERA further enables localization of the site of pathology along the auditory pathway from the VIIIth cranial nerve to the upper brain stem.

Despite the considerable number of audiological tests currently in use, difficulties occasionally arise in providing a clearcut diagnostic evaluation. Ambiguity may be present both with respect to identification and estimation of hearing impairment (Jerger *et al.*, 1980; Finitzo-Heiber, 1982), as well as with respect to the site of the problem i.e. whether peripheral, central or both. These dilemmas commonly occur when only one testing method is employed but occasionally occur even when a second 'cross-check' test is performed. The present study aims to define this small group of children in whom ambiguous results may occur and outline features to be taken into consideration when interpreting results.

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Material and methods

Ninety-four developmentally retarded children presenting to our department with histories of suspected hearing loss and speech retardation were prospectively evaluated in the period from December 1992 to March 1994. These children underwent auditory assessment by both behavioural observation audiometry (BOA) and brain stem evoked response audiometry (BERA). Their ages ranged from five months to 11 years. There were 50 boys and 44 girls. All children underwent an assessment by a clinical psychologist and a paediatrician and the degree of developmental retardation present was documented.

Testing methods

(1) Behavioural observation audiometry (BOA).

Based on the mental age of the patient hearing was assessed either by distraction tests, play audiometry or pure tone audiometry. The stimuli used included the following: calibrated rattles, drums, bells and clacker, warble tones of 0.5, 1, 2 and 4 kHz, narrow band noise and speech.

Testing was performed in the conventional manner. The quietest intensity level at which a response was obtained was noted. Responses from both ears separately could be assessed correctly only when headphones were used. Children whose test results were declared inconclusive were assessed no less than three times before arriving at this conclusion.

(2) Brain stem evoked response audiometry (BERA).

All children were sedated using either chloral hydrate syrup at a dose of 65 mg/kg or vallergan syrup at a dose of 4 mg/kg. In some children, repeated attempts at sedation on different days were necessary to achieve adequate sedation.

BERA was performed using a Nicolet Pathfinder I (Nicolet Electrodiagnostics Inc.). Scalp electrodes were placed at the vertex (C Z1, active) and on the mastoid regions (A1 and A2). One of the mastoid electrodes (A1 or A2) served as the reference electrode while the other served as the ground electrode. The impedance was kept below 5 kOhm. Rarefaction click stimuli and frequency specific tone pips were delivered at a rate of 21–1 per second through TDH-39 earphones. The bandpass filters were set at 150 to 3000 Hz for clicks and 50 to 3000 Hz for tone pips.

The analysis time was set at 10 or 20 ms and 1024 or 2048 averages were obtained for each recording. Two replicable tracings at each intensity level were used for analysis.

The BERA recordings were analysed to note the following: (i) lowest intensity at which wave V was recordable; (ii) interwave interval; (iii) absence of any or all waves; and (iv) amplitude of waves.

Criteria for assessment

(1) BOA

A child's hearing was declared to be normal if its response to the given stimulus was correct for the appropriate intensity level and developmental age. Established standards were used for comparison (Northern and Downs, 1994). If a response was obtainable only at levels more than two standard deviations above the recommended levels, hearing loss was diagnosed.

(2) BERA

A normal response was defined as the presence of a detectable wave V when a stimulus of 20 dB (or less) intensity was delivered. Absence of wave V with or without the presence of earlier waves was considered to be an absent response.

Results

Most children were found to have moderate to severe developmental retardation. The various associated diagnoses made in these children are listed in Table I.

The results of the auditory assessment by BOA and BERA are shown in Table II. Agreement between the results of both testing methods resulting in a clearcut diagnosis regarding hearing sensitivity was present in 54 children (57.5 per cent). In 15 children the results of BOA were either inconclusive (n = 9) or not elicitable (n = 6) and hence threshold determination was possible only by the results of BERA. Additionally, seven children who had unilateral hearing loss, were diagnosed only on the basis of BERA results.

Of the remaining 18 children, two groups could be identified whose results merited special consideration. Group 1 (n = 7) consisted of children whose BOA results differed from their BERA results. Of these, there were six children in whom a diagnosis of bilateral severe hearing loss was made by BOA using the criteria set forth above. These children were shown to have normal auditory thresholds by BERA. One child in Group 1 was found to have normal hearing by BOA although BERA revealed elevated auditory thresholds. Group 2 (n = 11) consisted of children in whom BERA showed no measurable wave V even at stimulus levels of 105 dB nHL. In four of these children wave I alone was discernible in the recorded response. BOA in these 11 children showed unreliable or inconsistent

TABLE I
ASSOCIATED CONDITIONS IN TESTED CHILDREN (N = 94)

Diagnosis	Number of patients
Cerebral palsy	15
Cleft palate	1
Congenital rubella syndrome	2
Cortical blindness	2
Downs syndrome	4
Hydrocephalus	2
Infantile autism	1
Microcephaly	10
Phenylketonuria	1
Post-encephalitic sequelae	2
Waardenberg's syndrome	2

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TABLE II
DISTRIBUTION OF RESULTS OF BERA AND BOA IN RETARDED CHILDREN (N = 94)

Results of auditory assessment	Number of patients
Children in whom results of BOA and BERA agreed	54
Children in whom diagnosis was made only by BERA:	22
(a) Inconclusive BOA	9
(b) No response by BOA	6
(c) Unilateral hearing loss	7
Children whose test results required special interpretation	18
(Group 1 (results of BOA and BERA different):	
(a) Normal BERA abnormal BOA	6
(b) Normal BOA abnormal BERA	1
Group 2 (no response by BERA):	
(a) Inconclusive BOA	5
(b) No response by BOA	6

responses in five children and no response in the remaining six.

Discussion

The present study serves to address three problem situations which confound paediatric auditory assessment, viz, inconclusive responses using BOA, discrepancies between BOA and BERA results and absent responses. Evidently, both BOA and BERA are essential for the auditory assessment of developmentally retarded children. A little over half of these children could probably have been diagnosed by BOA alone. This figure may be higher in centres solely concerned with the management of retarded children. However, in a multidisciplinary centre such as ours, where time is at a premium because of the heavy workload, a more detailed auditory assessment by BOA is not feasible. A reasonably quick, objective and accurate auditory assessment is what is required. Administration of a combination of both techniques appears to provide the maximum information.

Unreliable or inconclusive responses with BOA

In the present study, the absence of reliable responses occurred in 17 children. McCormick (1994) stated that under ideal conditions children aged six to 18 months respond only 70 per cent of the time to stimuli used in BOA. In a developmentally retarded child, sustaining the child's attention is even more challenging. Response assessment is further hampered by concomitant motor disability like spasticity and involuntary movements. When results using these tests are inconsistent after two or three trials, it may become necessary to abandon further testing in favour of BERA.

Lack of correlation between BOA and BERA

BERA thresholds have been shown to correlate well with mean BOA thresholds in the 2 to 4 kHz frequency range (Coats and Martin, 1977). In the present study, six children appeared to have bilateral hearing loss by BOA while BERA revealed normal auditory thresholds. What could be the reason for this discrepancy? Kaga and Tanaka (1980) have shown that behavioural thresholds closely approximate BERA thresholds only above the age of one year. Below that age differences of up to 60 dB have been observed. In the present series, three children with a developmental age of around one year, had behavioural thresholds exceeding the BERA thresholds by 50 to 60 dB HL. This might well represent the process of normal maturation of neural structures in the central nervous system (CNS). Such children probably may be managed best by a repeat testing three to six months later in order to note improvement in behavioural thresholds with time. Taylor (1964) described a group of children with cerebral palsy whose initial testing (using behavioural techniques alone), suggested profound deafness. Subsequent testing several months later, however, revealed normal responses to sounds. He suggested that this phenomenon could be due to delayed maturation of the integrating mechanisms of the brain in these children. The improvement in responsiveness to sound may, however, be incomplete.

Although neural maturation may to some extent account for the discrepancies in hearing level estimation, it is also possible that BOA may have overestimated the degree of hearing loss in some. Accurate assessment of responses in BOA is dependent upon several variables like the child's level of attentiveness, ambient noise, type of stimulus used and the skill and experience of the tester. Despite the use of optimal conditions, false-positive rates of BOA are still quite high. Clearly, where BERA thresholds are obtained at normal levels, the diagnosis of a normal peripheral hearing system may be made, even if BOA results are different.

The reverse situation in which one child had normal hearing on BOA but elevated thresholds on BERA also poses a dilemma. In this child, aged four years, BERA thresholds were established at 80 dB for clicks and tone pips. Waveform analysis revealed prolonged interwave latencies suggesting brain stem dysfunction. One possible explanation for this

phenomenon is that in the presence of neurological dysfunction the amplitude of wave V at high intensities is markedly reduced. Desynchronization of the auditory pathways may prevent a recordable response from being obtained at lower intensities (Picton *et al.*, 1992).

Discrepancies between threshold estimation by BOA and BERA may also be because the pathway followed by the auditory brain stem response is different from the pathway subserving hearing as tested by behavioural methods. The two pathways may be affected independently of each other (Picton *et al.*, 1992). Such patients evidently do not require amplification. The existence of such a phenomenon highlights the need for both BOA and objective methods such as BERA in assessing the hearing of brain damaged children.

In the present study, none of the seven children with unilateral hearing loss could be identified by BOA. While three children gave inconsistent responses, four children were labelled as having either normal hearing or bilateral hearing loss. The importance of BERA in identifying this small, but well defined group, needs to be underscored.

Lack of response by BERA

Approximately 11 per cent of the children in our study demonstrated no response by BERA. Jerger *et al.* (1980) found that six per cent (nine children) out of a total of 151 children, with and without other evidence of CNS pathology, showed no responses by BERA. While he included these children in the group with profound hearing loss, he cautioned that the absence of response by BERA was not a definite indication of hearing loss. He suggested that involvement of central pathways may preclude any sort of response on BERA. Similarly, other authors (Worthington and Peters, 1980; Kraus *et al.*, 1984) have described obliteration of the auditory brain stem response despite the presence of a normal peripheral auditory system. Finitzo-Heiber (1982), however, maintains that such children indeed have a severe hearing impairment and assessment of CNS involvement may not be possible with BERA because of the absence of waves.

The absent response is, perhaps, the most ambiguous of results in children with obvious evidence of CNS involvement. Analysis of the auditory brain stem response cannot help to distinguish peripheral from central pathology in these situations. Electrocochleography may be necessary in order to localize the site of the disorder. Identification of wave I, without the presence of subsequent components, points to underlying neurological pathology. However, the presence of concomitant peripheral and central pathology cannot be discounted.

Sohmer and Student (1978) studying the auditory brain stem response in children with psychomotor retardation and minimal brain dysfunction, inferred that the prolonged brain stem conduction time found in most of these patients was due to diffuse brain stem pathology. In patients in whom at least waves I and III are present, it is possible to note this abnormality. However, in children in whom no waves are detectable in the auditory brain stem response, as was the case in seven children in this study, such localization is not possible. The best strategy in these children would be to have regular follow-up with assessment by BOA and BERA at three to six monthly intervals. Observation of response at a subsequent follow-up visit would indicate progression of neuromaturation.

It may be pertinent to mention here that evoked otoacoustic emission (EOAE) testing (Kemp *et al.*, 1990), which is a new and promising method of detecting cochlear function, could be of help in determining whether the hearing mechanism up to the cochlear level is intact. The results of EOAE testing in children with absent responses by BERA should indeed prove very useful in localization. However, limitations in EOAE testing such as the necessity for standardization of response evaluation and instrumentation (Rupa and Musiek, 1991), as well as the lack of sufficient information regarding its specificity (Cope and Lutman, 1994), need to be addressed first.

It is important to realize that electrophysiological and behavioural testing have complementary roles in hearing assessment. Their reliability and accuracy are mainly dependent upon the technical expertise as well as facilities available in any particular audiological set-up.

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