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Original Research Article

Correlation between brainstem evoked response audiometry with other audiological tests in different types of hearing loss

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ABSTRACT

Background: Brainstem evoked response audiometry (BERA) is most specific and sensitive test for brain stem dysfunction. It is most important objective method for evaluating peripheral auditory system in neonates, infants, sedated and comatose patients and other person who doesn't understand the language. Objective of the study was to evaluate correlation BERA with other audiological tests in different types of hearing loss as well as to study variations of wave forms in different types of hearing loss.
Methods: Patients underwent a complete ENT check up to rule out any actively discharging gears, wax, infection or any middle ear problems. Different audiometric tests: pure tone audiometry (PTA), distortion product otoacoustic emissions, auditory steady-state response (ASSR) and BERA were applied to the patients.
Results: The majority of the patients (32 cases) belonged to the age group of 0-5 years. Maximum cases were of sensorineural hearing loss (60%). ASSR was highly sensitive (85.1%) for estimation of hearing threshold and specificity was 100% ($p<0.001$). BERA was also highly significant for estimation of hearing threshold (sensitivity: 83%; specificity: 92.3%; p value <0.001).
Conclusions: BERA has high degree of accuracy in detecting hearing threshold as an objective test but not as much accurate as ASSR. It is more valuable in terms of identification of site and size of the lesion in auditory pathway and identification for the type of the deafness.

Keywords: Correlation, Brainstem evoked response audiometry, Audiological tests, Hearing loss

INTRODUCTION

Hearing impairment is classified into several types based upon sites of lesions and degree of hearing loss.[1] The most commonly used types are conductive, sensory, neural (peripheral neural and central neural), non-organic, and mixed. Sensory relates to the cochlea and neural to the subsequent sections (peripheral and central) of the auditory pathway.[2] Standard audiometric tests are ineffective in detecting the subtle deficits resulting from pathology in the central auditory nervous system.[3] Thus, a variety of tests have been designed to test the central auditory function. These tests can be divided into behavioural tests and objective tests.[4] Behavioural tests are those, which require patient cooperation in regarding to a given stimulus. Objective tests do not require patient cooperation and provide recordable data in response to an acoustical stimulus.[4] Objective tests include assessment

of stapedial reflex threshold, otoacoustic emission and auditory brainstem response (ABR). Brainstem evoked response audiometry (BERA) is most specific and sensitive test for brain stem dysfunction.[5] It is most important objective method for evaluating peripheral auditory system in neonates, infants, sedated and comatose patients and other person who doesn't understand the language.[5] An objective evaluation of auditory function is also helpful in patient suspected of having non organic hearing loss, malingers and in sedated patients.[5]

The first description of the human ABR is credited to Jewett and Williston in 1971.[6] The ABR measures activity from the auditory nerve up to the level of the brain stem. It measures hearing sensitivity in the range from 2000 Hz to 4000 Hz. In normal person, stimulation of the ear with high intensity clicks (60 dB to 100 dB) may produce 67 waves in first 10 ms.[6] With decreasing intensities, early I to IV waves disappear, but the wave V is most permanent and resistant to change in intensity and persists to a level that relates closely to psychoacoustic threshold. The lowest intensity at which other waves disappear while wave V persists determines the hearing threshold.[7]

Jewett and Willinston (1971) recorded that these auditory evoked waves indicates different anatomical site for their generation: wave I generated from distal eighth nerve; wave II from proximal eighth nerve; wave III from Trapezoid body and olivary complex; wave IV originate from ventricle of lateral laminiscus; wave V generation represents the site of inferior colliculus; and generator sites of wave VI and VII are uncertain.[6] The ABR waves are studied for absolute latency, interweave latencies and amplitude. The latency of each wave decreases with increasing intensity, but the inter wave relationship remains constant.[8] Comparison of waves V latency after stimulation of each ear separately is of value in distinguishing cochlear from retro-cochlear pathology. In cochlear deafness, the interaural difference in wave V latency, which is seen at low intensities, show progressive disappearance with high intensities (due to phenomenon of recruitment). While in retro-cochlear deafness, there is consistent interaural difference in waves V latency.[9]

Thus, BERA (brainstem evoked response audiometry) plays a very important role in assessment of auditory functions.[10] In regards to above facts, a prospective was planned to evaluate correlation BERA with other audiological tests in different types of hearing loss as well as to study variations of wave forms in different types of hearing loss.

METHODS

A prospective study correlating BERA with other audiological tests and the study of variations of wave forms in different types of hearing loss was carried out in Department of ENT, Sir Takhtasinhji General Hospital, Bhavnagar from January 2010 to June 2011. Study was approved by Institutional Review Board. Patients were clearly explained regarding the study and written informed consent was obtained from each patient before their inclusion in the study. Patients with normal ears and auditory functions; unilateral or bilateral hearing problems; delayed development of speech and language in paediatrics; high risk new borne babies and malingerer were included in the study. While patients, with active ear discharge, external ear pathologies in whom ear plug cannot be applied, were excluded from the study.

Patients underwent a complete ENT check up to rule out any actively discharging gears, wax, infection or any middle ear problems. Audiometric tests were applied to the patients as per the performa pure tone audiometry (PTA); impedance audiometry esp. middle ear compliance and stapedial reflex; distortion product otoacoustic emissions (DPOAE), auditory steady-state response (ASSR). Evaluation of the hearing by BERA was applied after the patient goes through all these tests. For paediatric patients, they were sedated by administering triclofos after obtaining consent form parents or legal guardians.

A complete and detailed history was taken in every case. Each patient was subjected through general, local and systemic examinations. Routine and special investigations will be carried out as needed to rule out systemic disease. All the findings so obtained were recorded accordingly on the performa especially made for it.

PTA

This was carried out by MAICO MA 52 (diagnostic 6 mbh berlin, salzufer 13/14D-105087 berlin) audiometer calibrated to ISO. Audiometry was done using Hugson and Westlake (5 up and 10 down) procedure by audiologist. It was carried out in a sound treated audiometry room. Before each procedure the audiometer was calibrated with a co-student whose hearing was already tested and noted in the same audiometer. The subject was seated in front of the examiner and explained the procedure. The head phone was put on. Mode switch turned to 'right 'or 'left' depending upon the ear to be tested. Audiometer was switched to 'on'. The frequency dial was adjusted to 1 KHz. And minimum intensity at which the subjects hears was taken as threshold for air conduction (AC) at the frequency. Similarly, all the other frequencies 2 KHz, 4 KHz, 8 KHz, 0.5 KHz and 0.25 KHz were tested for air conduction threshold and noted.

DPOAE, ASSR and BERA

Tests were carried out in air-conditioned room designed especially for the audiological tests. The instrument used was a fully computerized machine (intelligent hearing systems, smart USB lite box, Miami, FL USA) with the

facility of artefact rejection. USB lite box consists of 33 MHz digital signal processor (TMS320C31), 1 channel optiamp receivers, 1 channel OAE amplifiers and USB interface. For measuring DPOAE, 10D OAE probe was used. The skin was cleaned with spirit and skin preparatory paste. The electrodes were placed as follows: RED: right ear (negative when testing right/ground when testing left). BLUE: left ear (negative when testing left/ground when testing right). BLACK: positive.

Procedure

Subject lying supine with a pillow under his head. Room was kept quiet and made specially for the purpose of audiometry (sound proof room). Electrodes electrolyte gel was used and electrodes were fixed. Impedance was kept under 07 kohms and were within 02 kohms of each other. The stimuli used were presented via insert ear phones (Etymotic ER3A). Time windows of 25 m/sec were used to record the click evoked ABRs. Click stimuli were 25 microsec in duration and were presented at a rate of 21.7/sec with rare faction polarity. Total clicks given: 1024. Filter settings used: between 100-3000 Hz. BERA tracing started at around 90 dB. Averaging process started and continued until the required repetition accomplished and intensities decreased by10 dB. Existence of the peak V was considered as sound stimulus heard and perceived by the auditory mechanism. Calculate the peak-interpeak latencies for the BERA waves. Continue taking BERA tracing still V^{th} wave disappears from the tracing and the intensity at which it disappears is considered as the potential threshold of the hearing for that particular ear. Tracings were reproducible. ASSR and DPOAE were recorded by the same instrument. All the findings so obtained were recorded accordingly on the performa especially made for it the relevant data were analysed keeping separately the sex, age, disease, site, etc. and their significance was discussed in the light of available literature.

Hearing was evaluated at 500 Hz, 1000 Hz, 2000 Hz and 4000 Hz only (speech frequencies). Grading of hearing level was done according to following criteria.[11] 0-25 dB: normal hearing, 26-40 dB: mild deafness, 41-55 dB: moderate deafness, 56-70 dB: severe deafness, 71-90 dB: very severe deafness, above 90 dB: profound deafness.

Identifying the type of deafness: for each and every patient latency intensity graph was plotted for the wave V. Hearing of classified as per below.[8] Normal: there is a shaded area for the normal person. If your points fall in this area then the person is having normal hearing. Conductive deafness: the latency-intensity graph plotted will be above and parallel to the shaded area. Sensorineural deafness: the graph plotted will be irregular and not forming a curve. Cochlear deafness: graph will be shallow.

Statistical evaluation

In the present study the statistical significance of the observations was tested using Pearson chi-square test. The values of the probability (p) for the calculated chi-square values was inferred and sensitivity and specificity were calculated for the given test.

RESULTS

A total number of 60 patients of normal as well as those coming to test for their hearing loss were included in this study. In the present study, the majority of the patients (32 cases) belonged to the age group of 0-5 years. The youngest patient was a child of 1.5 months and the oldest was of 41 years of age. Male patients outnumbered female patients and male to female ratio was 19:1 (Table 1 and Figure 1).

All the patients were classified into normal hearing and unilateral or bilateral hearing loss as shown in (Table 2). The patients were classified for type of deafness according to BERA by the means of latency intensity curve as defined in materials and methods and the results were described in (Table 3). Maximum cases were of sensorineural hearing loss (60%) followed by normal hearing patients (23.33%).

Table 1: Distribution of the patients according to age (n=60).

Age (years)	No. of patients	Percentage (%)
0-5	32	53.33
6-10	9	15.00
11-15	2	3.33
16-20	6	10.00
21-25	3	5.00
26-30	4	6.67
>30	4	6.67
Total	60	100.00

Table 2: Distribution of the patients according to hearing level (n=60).

Hearing level	No. of patients	Percentage (%)
Normal	14	23.33
Unilateral	10	16.67
Bilateral	36	60.00
Total	60	100.00

Table 3: Distribution of the patients according to type of hearing loss (n=60).

Type of hearing loss	No. of patients	Percentage (%)
Normal	14	23.33
Conductive	1	1.67
Sensorineural	36	60.00
Cochlear	6	10.00
Retro-cochlear	3	5.00
Total	60	100.00

Table 4: Distribution of patients according to absolute latencies and interpeak latencies (n=60).

Deafness	Latency	Absolute latencies			Interpeak latencies	
		Wave I	Wave III	Wave V	I-III	I-V
Normal	Absent	0	0	0	0	0
	Normal	13	14	11	14	14
	Prolonged	1	0	3	0	0
	Total	14	14	14	14	14
Conductive	Absent	0	0	0	0	0
	Normal	0	0	1	1	1
	Prolonged	1	1	0	0	0
	Total	1	1	1	1	1
Sensorineural	Absent	22	22	22	22	22
	Normal	9	14	8	14	14
	Prolonged	5	0	6	0	0
	Total	36	36	36	36	36
Cochlear	Absent	3	3	3	3	3
	Normal	0	0	0	3	3
	Prolonged	3	3	3	0	0
	Total	6	6	6	6	6
Retro-cochlear	Absent	1	1	1	1	1
	Normal	2	1	0	2	0
	Prolonged	0	1	2	0	2
	Total	3	3	3	3	3

Table 5: Comparison of PTA and ASS at different frequency level (n=60).

Frequency (Hz)	PTA	ASSR		Total	Significance	
		Hearing loss	Normal hearing			
500 Hz	Hearing loss	40	7	47	χ^2=29.47 df=1 p<0.001	Sensitivity=85.1% Specificity=100%
	Normal hearing	0	13	13		
	Total	40	20	60		
1000 Hz	Hearing loss	40	8	48	χ^2=17.27 df=1 p<0.001	Sensitivity=83.3% Specificity=83.3%
	Normal hearing	2	10	12		
	Total	42	18	60		
2000 Hz	Hearing loss	40	13	53	χ^2=8.06 df=1 p<0.001	Sensitivity=75.5% Specificity=85.7%
	Normal hearing	1	6	7		
	Total	41	19	60		
4000 Hz	Hearing loss	41	11	52	χ^2=7.43 df=1 p<0.001	Sensitivity=78.8% Specificity=75.0%
	Normal hearing	2	6	8		
	Total	43	17	60		
Average	Hearing loss	40	7	47	χ^2=29.47 df=1 p<0.001	Sensitivity=85.1% Specificity=100%
	Normal hearing	0	13	13		
	Total	40	20	60		

Table 6: Comparison of PTA and BERA (n=60).

PTA average	BERA threshold		Total	Significance	
	Hearing loss	Normal hearing			
Hearing loss	39	7	46	χ^2=22.70 df=1 p<0.001	Sensitivity=83.0% Specificity=92.3%
Normal hearing	13	1	14		
Total	52	8	60		

According to the recordings obtained by BERA, the cases were divided into 5 groups of nature of hearing. From the curves obtained by it, we classified them as absent, within normal limit and prolonged curve latency as per the reference tables given in materials and methods. As we can see most of the cases did not show any waves on the tracings (26 cases). For our study main wave to be taken into the consideration is wave V which was prolonged in total 13 cases out of which 6 cases were of sensorineural deafness, 2 were of retro-cochlear and 1 was of cochlear deafness. Interpeak latencies of wave IV were prolonged in total 2 cases which were of retro-cochlear deafness (Table 4).

As per the (Tables 5 and 6), we also attempted to compare BERA with the standard audiological tests for the threshold estimation. Pure tone audiometry was taken as a standard test for threshold estimation and ASSR and BERA were compared to it for the sensitivity and specificity. As we can see ASSR was highly sensitive (85.1%) for estimation of hearing threshold and specificity was 100%. The p value obtained was highly significant ($p<0.001$). The same results were obtained at 1000 Hz also but at this frequency the specificity of ASSR was 83.3% and specificity was also the same with highly significant p value (0.000032). At 2000 Hz the results were same but the sensitivity was somewhat reduced (75.5%) with still highly suggestive p value (0.0045). At 4000 Hz almost same sensitivity of ASSR as compared to PTA for threshold estimation can be seen (78.8%) with 75% of specificity and with still highly suggestive p value (0.0064). We had compared average PTA threshold at 500Hz, 1000 Hz, 2000 Hz and 4000 Hz with that of average ASSR threshold at same frequencies.

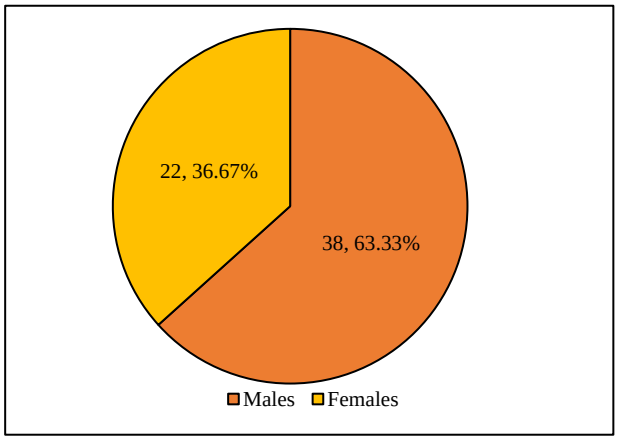

Figure 1: Distribution of the patients according to gender (n=60).

Pure tone threshold was also compared with BERA (click stimulus BERA) thresholds and the results were highly significant for BERA (sensitivity: 83%; specificity: 92.3%; p value <0.001). ASSR is the better tool than BERA (sensitivity 85.1%, specificity 100%). BERA is not as specific as ASSR for as hearing threshold estimation as compared with PTA.

DISCUSSION

60 consecutive patients, who undergo hearing assessment in the ENT Department of ENT, Sir Takhtasinhji General Hospital, Bhavnagar. As ASSR is a new tool to assess hearing sensitivity in patients, PTA was compared with ASSR and BERA to know which test is better for the estimation of hearing thresholds of the patients. The youngest patient in our study was a 1.5 months old male child and oldest was 41 years old male patient. Majority (32%) were in the age group of 0-5 years of age group. This age group is of particular importance and regarded as critical age for acquiring language. The child who does not get auditory stimulation during this period loses neural plasticity.[12] Neural plasticity is essential for learning to listen (i.e., to recognize words). It fades completely between 6-8 years.[12] Experimental evidence in animals suggests changes in both the brain stem auditory nuclei and primary auditory cortex.[13] These structural changes can be reversed early in life. In order to get benefit from the critical period of linguistic development, the identification of hearing impairment, use of appropriate amplification and stimulation of hearing must occur as early as possible.[13]

The majority of children who are deaf have some residual hearing; a fact recognized for decades.[3] Profoundly deaf children, if properly aided, can detect most if not all of the speech spectrum.[14] If these children are taught about the active use of amplified residual hearing in early childhood, they may become independent capable of speaking and contributory members of the mainstream society.[15] Students have reported that minimum delay between parental suspicion of hearing loss and audiological assessment is between 7-24 months.[16] This is in agreement with present study as most of the children presented to us for the first time were in age group of 0-5 years. In the present study, the duration of deafness ranged from 1 month to 5 years. 49 patients (81.66%) were below 20 years of age. According to Beyea et al (2016), duration of deafness prior to cochlear implantation significantly affects postoperative speech recognition performance in adult.[17] Seldran et al (2011) reported the similar observation.[18] A correlation between duration of deafness and spiral ganglion cell count has also been proved.[19]

A typical normal auditory brainstem evoked response consisting of five prominent waves with mean absolute latencies and interpeak latencies were recorded and compared to mean normal value as has been proposed by Jacobson (1976).[20] All ages are referenced from conception. 26 cases (43.33%) showed absent response on BERA. In 24 cases (92.30%) the absent waves were found in both ears, while in rest two cases (7.70%) waves were absent in only one ear. In 1 patient wave I, wave III and wave V were delayed, with interpeak latencies of wave I-III and I-V within normal limits. In other words, all the waves have shifted towards right, indicating that these patients were suffering from conductive hearing

loss.[21] The threshold of hearing in this patient was between 30-40 dB. Although approximate prediction of level of hearing loss in this patient can be made by measurement of the rightward shift in waves.[21] The most accurate assessment is derived from the actual ABR threshold.[22] The most prevalent middle ear disorder in young children is otitis media-collection of fluid and thicker glue in middle ear cause increased hearing loss at lower frequencies.[23] In this study cause of conductive deafness was not identified. A conductive hearing loss can also arise as a result of complex external and middle ear malformations, such as atresia of the auricle.[24] In present study, no such abnormalities were detected. In high frequency cochlear impairment (6 cases), wave I and III were absent in 3 cases while wave V was present in 3 cases, wave form morphology was poorer and absolute latency for each wave is increased. However, the interpeak latencies remain same in cochlear hearing loss.[22] In this study, 9 patients (15%) showed delayed and absent wave I with normal I-V interpeak latency in 3 cases, suggesting severe inner ear disorder, affecting the cochlea or the proximal part of the cochlear nerve. Although the most common origin of hearing loss in infant and young children is either in the middle ear or cochlea. It is possible that pathology could be present at higher level of the auditory pathway either the brainstem or cortex.[25]

The ABR will only identify those retro-cochlear hearing losses that occur at a level below the generator sites of wave V (i.e. lateral laminiscus and inferior colliculus).[9] Retro-cochlear part of auditory pathway may get affected by several disorders; the acoustic tumour is the most common. It arises from Schwann cells of vestibular portion of VIII cranial nerve.[26] ABR findings in acoustic neuroma are of complete absence of primary ABR waves (I, III, V).[27] Latency of wave V is significantly greater. Inter aural latency difference of >0.3 m/sec is highly sensitive indicator of acoustic neuroma.[28] In the present study 2 patients (3.33%) showed prolonged I-V interpeak latency while absolute latency of wave I was normal, suggesting that the lesion was situated in the retro-cochlear auditory pathway.[29] In these 2 patients the increased I-V latency was due to increase in III-V latency, which suggested the upper brainstem involvement. About 80% of the group of patients having either upper or lower brainstem involvement were associated with severe degree of mental retardation. This is in agreement with previous study by Lemay et al (2003).[30] This retro-cochlear lesion and associated mental retardation might have been caused by intrauterine or perinatal insult to the pathway leading to the degree of structural involvement.[30] In low frequency hearing defect or severe structural involvement above the midbrain or in malingerers may have complaint of serious hearing problem but may show normal brainstem evoked response.[31] Synonyms for this condition include psychogenic hearing loss, functional hearing loss, malingering and pseudo hypacusis.[31] The commonest reason for this condition is financial and related to claim for damage.[31] The basis of compensation assessment is degree of hearing loss and therefore degree of accuracy required in determining hearing threshold is much greater than that required for diagnostic audiometry.

BERA being an objective test is invaluable in diagnosis and assessment of degree of hearing loss in such cases.[4] The incidence of non-organic hearing loss has been reported to be (20-25)%, but varies depending on the patient population.[32] As observed in our study 26 cases (43.33%) were not showing any waves, but on examining the rest of the cases (34 cases) we saw a pattern in each wave latency. As we decreased the intensity of sound stimulus starting from 90 dB on gradual decreasing order of 10 dB, we observed that the absolute latencies of all the waves (I, III, V) were gradually increasing in each and every case establishing an inverse relationship with that of intensity and latency. We also observed that at highest intensity, all the curves (I, III, V) were present but as intensity decreases, with the gradual prolongation of the absolute latency curves start disappearing and at lower intensities each wave were absent except wave V. Wave V was last to disappear which was the threshold point for the particular patient's hearing level.

In our study we had diagnosed a case of Waardenburg syndrome, in which diagnosis was made with the combined efforts of ENT Department and Ophthalmology Department of our hospital.[33] We had found bilateral stable hearing loss which was of sensorineural type, no response on free field audiometry, no response on right ear ASSR other than at 1000 Hz at which she had 80 dB of hearing threshold. BERA waves were absent in right ear but in left ear she had prolong wave I and V with normal I-III and I-V interpeak latencies and disappearance of wave V in between 50-60 dB. DPOAE suggested refer in both ears. She had white for lock over head with white iris of both eyes which were typical of Waardenburg syndrome.

Similarly, we had diagnosed a case of Apert syndrome with the help of pediatrics department of our hospital.[34] We had observed bilateral hearing loss of sensorineural type with poor response on free field audiometry and severe hearing loss on ASSR in both ears. In this case wave I and V were prolong with normal interpeak latencies of wave I-III and wave I-V, DPOAE suggested refer in both ears. This patient had syndactyly of upper and lower limbs with exophthalmos and typical facial features of Apert syndrome.[34] Patient was responsive to very high intensity sounds.

In our study, we had also compared pure tone audiometry with more sophisticated and objective test like ASSR for the purpose of threshold estimation at 500 Hz, 1000 Hz, 2000 Hz and 4000 Hz as well as average pure tone threshold and average ASSR threshold. The results were significant for the ASSR in terms of sensitivity and specificity of threshold estimation at 500 Hz (sensitivity: 85.1%; specificity: 100%), 1000 Hz (sensitivity: 83.3%;

specificity: 83.3%), 2000 Hz (sensitivity: 75.5%; specificity: 85.7%) and 4000 Hz (sensitivity: 78.8%; specificity: 75%). In all these frequencies ASSR was capable of estimating hearing thresholds more accurately than pure tone audiometry. Similar results were obtained on comparing average pure tone threshold and average ASSR threshold (sensitivity: 85.1%; specificity: 100%). In all these comparison p value obtained was highly significant for ASSR ($p < 0.001$). This suggests that for the purpose of threshold estimation ASSR is much better tool than pure tone audiometry. The sensitivity however decreases on higher frequencies.

Pure tone threshold was also compared with BERA (click stimulus BERA) thresholds and the results were highly significant for BERA (sensitivity: 83%; specificity: 92.3%; p value <0.001). This suggested that BERA is more sensitive and specific for the purpose of threshold estimation as compared to pure tone audiometry. As per above results on comparing BERA (click stimulus BERA) with that of ASSR average threshold, we can indirectly interpret that for the purpose of threshold estimation ASSR is the better tool than BERA (sensitivity: 85.1%; specificity: 100%). BERA is not as specific as ASSR for hearing threshold estimation as compared with pure tone audiometry. These are the similar results as obtained by Werff et al (2002) which suggest that BERA is more reliable tool for identifying size and site of lesion in auditory pathway and ASSR is highly sensitive and specific for hearing threshold estimation as compared to pure tone audiometry and BERA.[35]

CONCLUSION

BERA is a valuable and reliable tool in the diagnosis and management of the patients with sensorineural hearing loss. Its main application is as an objective test for estimation of auditory sensitivity in all cases in which the cooperation of the patient is unobtainable or doubtful and who are unfit for behavioural tests. BERA has high degree of accuracy in detecting hearing threshold as an objective test but not as much accurate as ASSR. It is more valuable in terms of identification of site and size of the lesion in auditory pathway and identification for the type of the deafness. Thus, BERA is one of the essential and valuable audiological investigations. It should be made available in all institutes where ENT services prevail.

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