Importance of Long Latency Potential in Pediatric Hearing Assessment

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Abstract

Numerous researchers' reports caution clinicians that infants with risk factors have reversible features of auditory dys-synchrony (AD). It is important for an audiologist to be able to differentiate such reversible condition or auditory maturational delay (AMD) from non-reversible AD. It is difficult to identify the exact nature and type of hearing loss in infants and toddlers with risk factors using routine audiological test battery as they present inconclusive tests results (ABR, BOA, OAE & Immittance) in a single assessment. So an appropriate test battery becomes essential in order to differentiate these mimicking conditions. Thus, the study was taken up with the aim to check the importance of LLR in pediatric hearing assessment, especially to differentiate AMD from AD and permanent hearing impairment. 55 infants/ toddlers (30 males & 25 females) were taken for the study with age below 2 years at the time of first evaluation. They were divided into four groups on the basis of ABR and LLR results. The results revealed that LLR is an important tool in differential diagnosis of different conditions that are likely to be encountered when dealing with the hearing assessment of pediatric population of infants/toddler less than 2 years of age. In LLR results large variability was observed across the subjects. So, it is recommended that the interpretation of LLR wave be cautiously approached especially with regard to the absolute latency. It is also recommended that rather than looking at the latency it would be better to look for the presence or absence of LLR for the differential diagnosis of different conditions.

Introduction

Approximately 10% of newborns are at risk for medical problems and developmental disability. Most infants at risk are detected either at birth as reflected in low APGAR scores or during the complete physical examination within few hours of birth.

Different aspects of neurogenesis take place somewhat independently yet simultaneously and interactively in infants. Degenerative and regressive events involving cell death, retraction of axonal process and elimination of synapses occur concurrently throughout the development (Berry, cited in Salamy, Eggermont & Elredge, 1994). Literature on developmental outcomes of infants has shown that at risk infants display increased susceptibility to a variety of physical and developmental deficits.

Hearing is critical for normal speech and language development which in turn is vital for most aspects of normal human development. A significant hearing impairment at birth can

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produce major disruption in language learning (Menyuk, 1977) and produce irreversible deficits in the development of central auditory pathways (Moore, 1985). Even a mild hearing impairment has been implicated in delayed development of auditory skills (Quigley, 1978). Early identification of hearing loss followed by appropriate management minimizes the auditory deprivation which can interfere with speech and language learning and central nervous system maturation. So, early identification will waive off a number of problems regarding person’s educational, social and economic development that might arise in future.

Hearing loss in children is silent and hidden disability. It is hidden because children, especially infants and toddlers, cannot tell us about their inability to hear well. However, there are several ways to identify hearing loss in infants. There are subjective and objective methods to identify hearing impairment. Such methods usually do not involve the subjects’ active involvement. Most commonly used behavioral method is Behavioral Observation Audiometry (BOA) and the objective methods are Auditory Brain-stem Response (ABR), Oto-Acoustic Emissions (OAE) and the Immittance audiometry, among which ABR is the most commonly used tool to identify presence or absence of hearing loss.

Galambos and Galambos (1975) found a drop in latency of the ABR peaks with age in the premature infants which they attributed to maturational changes. Roberts, Davis, Phon, Reichet, Sturtevants and Marshall (1982) concluded that ABR failures in the infants in their study resulted mainly from immaturity. Raj, Gupta and Anand (1991) found that on re-evaluation 8 out of 13 infants with risk factors, who had failed on BAER earlier, had developed normal thresholds and by 6 months follow-up, only 3% had hearing loss. Misra, Katiyar, Kapoor, Shukla, Malik, and Thakur (1996) reported BAER abnormalities and their reversibility in neonates with birth asphyxia.

It is evident that infants with risk factors are likely to have varieties of abnormalities in auditory system which might vary from permanent severe hearing loss to normal hearing; reversible audiological test results in Auditory Maturational Delay (AMD) and Auditory Dys-synchrony (AD) of different degrees. An appropriate test protocol is essential to differentiate one pathological condition from other as it not only aids to the early appropriate rehabilitation but also adds to appropriate counseling to the parents.

Need for the study:

Most authors agree that from 2-15% of infants with hearing loss may exhibit auditory neuropathy; that is, one can expect to identify auditory neuropathy in approximately 1-3 infants per 10,000 births (Rance, et al., 2003; Sininger, 2002). The great majority of infants with auditory neuropathy exhibit one or more high risk factors including blood transfusion, hyperbilirubinemia, anoxia, low birth weight, NICU residence or family history (Berlin, et al., 1998).

Numerous researchers reported (Raj, Gupta & Anand, 1991; Berlin, Morlet and Hood, 2003) and cautioned clinicians that infants with risk factors have reversible features of auditory dys-synchrony. Thus it becomes all the more important for us to be able to differentiate such conditions (AMD) from AD which is most often irreversible. But it is difficult to identify the exact nature and type of hearing loss in infants and toddlers with risk factors using test protocol including ABR, BOA, OAE and Immittance as they present with a conflicting test results in a single assessment. So an appropriate test battery becomes essential in order to differentiate these mimicking conditions.

It may be recalled that during the neural development dendritic arborization and synaptogenesis occur. Due to auditory deprivation the synapses lose their function or die which might lead to absence of LLR in most of the adult cases with AD. However, it is likely that if LLR is administered during synaptogenesis one might observe LLR in most of the infants. Lee, McPherson, Yuen and Wong (2001) reported of two cases (school going children) with AD, one with absent MLR and present LLR while the other had presence of both MLR and LLR (N1/P2). Thus, LLR could be useful tool to identify and differentiate different conditions in children with risk factors, if not all, at least most of the clients.

Aim of the study: Thus the study was taken up with the aim to check the importance of LLR in pediatric hearing assessment especially to differentiate AMD from AD and permanent hearing impairment.

Method

Subjects:

55 infants/toddlers (30 males and 25 females) were taken for the study with age below 2 years at the time of first evaluation. The subjects included both, those with risk factors which are associated with hearing impairment and also those who did not demonstrate any such risk factors. The family history of congenital or acquired hearing loss at an early age was also specifically looked for in these subjects.

Apart from this all the subjects were also distributed into 4 groups based on the results of ABR and LLR irrespective of presence or absence of risk factors for further analysis. Group I was assigned those subjects who had both ABR and LLR present whereas Group II and III consisted of those with ABR absent – LLR present and ABR present at least in second or third evaluation– LLR absent in first evaluation respectively. The final group, Group IV comprised of those subjects who had neither ABR nor LLR present. The number of subjects that formed a part of Groups I, II, III and IV, was 18, 14, 10 and 13 respectively.

Instrumentation:

1. A calibrated two-channel diagnostic audiometer OB922 with impedance matched speakers was used to obtain behavioral responses (BOA)
2. Transient Evoked Oto-Acoustic Emissions (TEOAE) were acquired using ILO292 (software version 5) in full TE menu option in order to examine the status of the outer hair cells to rule out absence or abnormal Auditory Brainstem Responses (ABR) due to cochlear pathology.

3. Intelligent Hearing System (HIS) Smart EP version 3.86USBeZ was used to obtain Auditory Brainstem Responses (ABR) and Long Latency Potentials (LLR) to check the integrity of neural pathway at the levels of brainstem and cortex respectively.
4. An immittance meter, Grason Stadler Inc. (GSI) Tympstar was used to rule out the presence of middle ear pathology causing absence of TEOAE or prolongation of ABR wave latencies.

All the instruments were checked for calibration prior to use on each of the subjects according to manufacturer's recommendations.

Test Procedure:

a. Case History:

Detailed information regarding the history of prenatal, natal and postnatal medical conditions was secured for each of the subjects. Medical records were looked for to obtain information regarding risk factors pertaining to congenital or early onset hearing loss like TORCH infections, neonatal jaundice, birth asphyxia, low APGAR scores, seizures, premature delivery, low birth weight, drinking of Amniotic fluid at the time of delivery, mother getting Chicken Pox in the first trimester of pregnancy and Bronchopneumonia. A detailed report regarding the auditory behavior of the subject at home for various environmental sounds like call bell, dog bark, voices from TV or radio, pressure cooker whistle etc. was obtained from the parents or caretakers. Parents were counseled regarding frequent follow-ups and were asked to look for changes in the auditory behavior and also to report those changes during the next follow-up visit.

b. Test Battery:

1) Behavioral Observation Audiometry (BOA):

Behavioral responses of the subjects were obtained in sound-field condition using warble tones or narrow band noise of 500, 1, 2 and 4 KHz and also speech stimuli. It was carried out in a double-room situation. The subjects were seated on the caretakers lap at a distance of 1 meter from the speakers and at an azimuth of 45° in the observation room. The stimuli were presented sequentially and the starting level was decided based on the parental report about the auditory behavior at home. The lowest levels of presentation of each of the stimuli at which the subject exhibited some sort of auditory behavior was noted down.

2) ABR and LLR:

Single channel ABR and LLR were recorded in sleep condition using IHS Smart EP instrument. The electrodes were placed at Fz (high forehead) for Non-inverting (positive), A1 (left ear mastoid) for inverting or ground and A2 (right ear mastoid) for ground or inverting. Neo-prep was used for preparing skin at these electrode sites in order to obtain allowable impedance values. Independent at each site and inter-electrode impedances were maintained within 5 KΩ. TDH-39 headphones were placed taking care not to dislodge the electrodes from their positions and the electrode impedances were re-measured to make sure that the impedance stayed within the desired levels at each of the electrodes. The parameters used for ABR and LLR recording have been shown in Table 2 and 3 respectively.

Table 2: Parameters used to acquire ABR

Acquisition Parameter	Stimulus Parameters
Sensitivity- 50 µV Band-pass Filters- Low Pass- 3 KHz High Pass- 30 Hz Notch Filter- Off Artifact Rejection- On Electrode Montage- A1- Fz- A2 Time Window- 15 msec.	Type of stimulus- Click Polarity- Rarefaction Intensity- Variable Number of stimuli- 1500 Repetition rate- 11.1/sec. (to obtain better waveform morphology)

Table 3: Parameters used to acquire LLR

Amplifier Set-up	Stimulus Parameters
Sensitivity- 50 µV Band-pass Filters- Low Pass- 300 KHz High Pass- 1 Hz Notch Filter- Off Artifact Rejection- On Electrode Montage- A1- Fz- A2 Time Window- 750 msec.	Type of stimulus- Click Polarity- Rarefaction Intensity- 70 dB Number of stimuli- 300 Repetition rate- 1.1/sec.

Presence of ABR (wave I or V) at lowest level was taken as threshold and used for interpretation as wave I is likely to be more prominent in infants. In case of LLR, the latencies of the two positive peaks (P1 and P2) and the two negative peaks (N1 and N2) were noted whenever these were present. In case any one or more of these peaks were absent, only the latencies of the peaks that were present were noted. LLR responses were shown to three experienced Audiologists to identify the peaks.

Infants with presence of ABR at 30 dBnHL and also presence of LLR were not followed-up. Only those infants who demonstrated absence of one or both of these potentials in the first evaluation were asked to follow-up to monitor any changes with development and diagnosis.

3) Oto-Acoustic Emissions (OAE):

TEOAEs were obtained using ILO292 instrument with a foam tip positioned in the external auditory canal so as to give a flat frequency spectrum across the frequency range. The stimuli were clicks filtered with a band-pass filter encompassing 500 to 6000 Hz. The duration of the rectangular pulses (clicks) was 80 µsec. The level was maintained at 80 dBpkSPL in the external auditory canal and the inter-stimulus interval was kept constant at 20 msec. A total of 260 averages above the automatic noise rejection level of instrument were stored for analysis. The presentation mode included a series of four stimuli, three at same level and of same polarity and the fourth of three times the level of either of the three and opposite in polarity. This, called the non-linear averaging, is used for artifact reduction during the response acquisition. The responses were considered as emissions based on the reproducibility and the signal-to-noise ratio (SNR). The overall SNR of greater than or equal to +3 dB and the reproducibility of greater than 50% were considered (Dijk & Wit, 1987) for it to be considered as a presence of an echo or emission.

4) Immittance:

Tympanometric measurements were done using 678 Hz probe tone (since infants and toddlers have mass dominant middle ear system) or 226 Hz based on the age of the subject at the time of evaluation. This was done to rule out absence of OAE due to middle ear pathology. Appropriate probe tips were used to obtain hermetic seal and comfortable pressure for the subject. The parameters documented were types of tympanogram to go with ear canal volume, acoustic admittance and the tympanometric peak pressure. The results were also correlated with the ENT findings.

Result and Discussion

The subjects were divided into four groups based on the presence or absence of ABR and LLR irrespective of the presence or absence of risk factors to understand the role of LLR in pediatric hearing assessment. Group I consisted of 18 subjects, Group II of 14, Group III of 10 and Group IV of 13 subjects. The profile includes findings of different tests (BOA, OAE, Immittance, ABR and LLR) that were administered on each of the subjects who participated in the study. Table 4 shows the audiological profile of cases who demonstrated presence of both-ABR and LLR (Group I).

It can be seen in table 4 that subjects 1, 2, 4, 9 and 10 had positive history for risk factors that are associated with hearing loss. Subjects 1, 2, 4 and 10 had history of Neonatal Jaundice (NJ) that did not require blood transfusion. In addition to NJ subject 2 also reported of delayed birth cry by 1 minute, Neonatal Meningitis (NM) and Febrile Seizures at the age of 2 months. Subject 9 had history of delayed birth cry by 1 minute without other associated complications. In spite of presence of risk factor/factors that have been found to be associated with hearing loss in literature these subjects' audiological results fall well within normal limits for BOA, OAE, ABR, Immittance and LLR except for subjects 4 and 9 who showed absence of OAE and slightly elevated BOA values for FM tones and speech stimuli. Occurrence of conductive pathology as indicated by B-type Tympanogram and positive history of ear discharge might have resulted in abnormal OAE and BOA results in these two subjects. This group was considered to have normal hearing based on the test battery result except for subject 4 and 9 as they had conductive component.

Table 4: Audiological Profile of subjects with ABR and LLR present (Group I)

Sub	Risk Factors	BOA Level (dBHL)		Tympanogram Type		OAE		ABR Threshold (dBnHL)		LLR at 70 dBnHL	
		T	**S**	**R**	**L**	**R**	**L**	**R**	**L**	**R**	**L**
1	P	40-55	35	A	A	P	P	30	30	P	P
2	P	45-50	40	A	A	P	P	30	30	P	P
3	Ab	35-50	35	A	A	P	P	30	30	P	P
4	P	50-65	50	B	B	Ab	Ab	50	40	P	P
5	Ab	40-55	40	As	As	Ab	Ab	40	40	P	P
6	Ab	40-55	40	A	A	P	P	30	30	P	P
7	Ab	45-50	45	A	A	P	P	40	40	P	P
8	Ab	35-45	30	As	As	P	P	30	30	P	P
9	P	50-60	45	B	B	Ab	Ab	50	50	P	P
10	P	50-55	40	A	A	P	P	30	30	P	P
11	Ab	35-45	35	A	A	P	P	30	30	P	P
12	Ab	30-45	30	A	A	P	P	40	40	P	P
13	Ab	30-40	35	A	A	P	P	30	30	P	P
14	Ab	45-55	45	A	A	P	P	50	50	P	P
15	Ab	35-40	30	A	A	P	P	30	30	P	P
16	Ab	35-40	30	A	A	P	P	35	40	P	P
17	Ab	35-40	30	A	A	P	P	30	30	P	P
18	Ab	30-40	30	A	A	P	P	30	30	P	P

‘P’ - ‘present’; ‘Ab’ - ‘absent’; ‘T’- ‘tone’; ‘S’ - ‘speech’; ‘R’-‘right’ and ‘L’ - ‘left’ ear

The youngest age of the subject in whom LLR could be recorded was 16 days and all the components of LLR (P1, N1, P2, and N2) were found to be present in this subject. It has also been reported by McPherson, Tures, and Starr (1989) that LLR or at least a component of LLR can be present in normal hearing infants even at birth. Though all the components of LLR could be recorded in the subject 12, it is not always necessary that all the LLR components would be observable at birth. But there is high possibility of presence of at least one of the LLR components at high intensity if the infants follow normal developmental pattern. Thus, it suggested that LLR might be able to substitute ABR provided the normative data is established for this population and also the relationship between the behavioral and LLR threshold is established.

Some time LLR might be a better tool for assessing hearing sensitivity when there is mild degree of dys-synchrony which might lead to noisy ABR morphology but might show better LLR responses. This might help to identify hearing sensitivity in such cases. 5 subjects had undergone three evaluations each at a interval of 3 months or more between two follow-ups and also 2 subjects who underwent two evaluations apiece at interval of three months or more. Rest of the 7 subjects underwent only 1 evaluation each. This group showed interesting finding of absence of ABR peaks even at 90 dBnHL in spite of BOA showing normal hearing in most of the subjects to severe hearing loss in subjects 1 and 3. However, all the subjects showed A-type tympanogram in presence of recordable TEOAE and presence of LLR at 70 dBnHL. Considering that the maturation of auditory system occurs from peripheral to central (auditory nerve to auditory cortex) (Romand, 1983; Montandon, Cao, Engel & Grajew, 1979; Stockard & Stockard, 1981, 1983), ABR also should have been present if LLR were to be present. But this was not to be in these subjects which gives an indication towards some permanent abnormality at the level of Auditory Nerve and Brain-stem which contain the generators for different ABR peaks. ABR is actually a test of neural synchrony and is dependent upon the ability of neurons to maintain precise timing and respond synchronously to external stimuli (Jewett & Williston, 1971). So the absence of ABR peaks could be because of dys-synchronous firing of the ANFs or in other words, Auditory Dys-synchrony (AD). LLR requires much lesser degree of synchrony of firing of the ANFs (Kraus, et al., 2000) which is why probably it was found to be present in these subjects. The physiology behind this aspect has been reported by Kraus et al. (2000). The cortical potentials reflect neural synchrony differently than ABR. The ABR peaks reflect synchronous spike discharge generated in the nerve tracts whereas the peaks in cortical responses reflect the summation of excitatory post-synaptic potentials. In other words the ABR reflects action currents in axons while the cortical potentials reflect slow dendritic events. Because unit contributions to the ABR are biphasic and of short duration ABR peaks tend to cancel when discharges are separated by fractions of a millisecond. In contrast, for cortical potentials, the waves are so slow that contributions separated by several milliseconds contribute to these later waves. While the ABR reflects highly synchronous discharge with millisecond precision the synchrony required for cortical potentials is on the order of several milliseconds.

This highlights that after 3 evaluations in 5 subjects it was possible to label them as having AD. In the first evaluation itself ABR was found to be absent and LLR present which prompted to diagnose the subjects as having AD. The follow-up testing was done only to confirm this status. All other subjects in the group were suspected to have AD based on similar findings in the first or first two evaluations but they could not be followed up owing to time constraint or their inability to come for follow-up. Thus we may be able to arrive at the diagnosis of infants or toddlers having AD after the first evaluation itself if ABR is absent but LLR present. This also receives support from Starr, Picton, Sininger, Hood and Berlin (1996) who reported presence of LLR in the subjects with Auditory Neuropathy which is more recently being called AD.

Subjects 3, 5, 6, 8 and 9 had absence of TEOAE which could be accounted by As-B, As-B, B-B, As-As, and As-As type of tympanogram respectively suggesting presence of conductive pathology in these subjects. The conductive pathology hampers the reverse transmission of OAE through the middle ear system and thus prevents the OAE from being recorded at the level of ear canal. If LLR was not done it would not have been possible to categorize these subjects into AD as most of the available literature on AD highlights the presence of OAE being an integral part of the test battery for its identification. Thus, it goes to show how important can LLR be in identifying AD in infants and toddlers at an early age even if OAEs are absent owing to some middle ear problem. This can implicate in early and appropriate use of intervention strategies or measure like cochlear implantation can be taken very early in life as the literature supports the usefulness of cochlear implantation in cases with AD (Peterson et al., 2003 cited in Kirk, Firszt, Hood, and Holt, 2006).

Also in subject 10 there is presence of ABR at 90 dBnHL only which suggests a milder degree of dys-synchrony in the ANFs' firing. LLR was found to be present at 70 dBnHL in this subject. Hyde (1997) found 10 dB discrepancy between the LLR threshold in asleep condition and the behavioral threshold (LLR threshold> Behavioral threshold) and suggested about 10-15 dB discrepancy between ABR threshold and behavioral threshold (ABR threshold> Behavioral threshold). This implies that LLR and ABR thresholds should roughly coincide which is not what was found in subject 10. This subject showed a discrepancy of grater than or in worst case equal to 20 dB between ABR and LLR. Also, BOA responses were observed at 45-55 dB and 45 dB for FM tones and speech respectively which suggests that the subject was hearing sounds at much lower intensities than suggested by ABR finding. All this in conjunction led to this subject being put in the category of having AD (i.e., Group II). The inclusion of this case in this group also receives support from Sininger (2002) who said that the neural response (ABR) will be poor or completely absent but will occasionally show a small wave V response (at high stimulus intensities). However, the subject did not come for follow up to confirm the diagnosis. Since LLR requires lesser synchrony than ABR, even small amount of demyelinization might have led to its presence whereas a small amount of demyelinization of lower structures (auditory nerve and brainstem) could have produced synchronous firing only at much higher level (90 dBnHL). So, this could even be classified in to AMD. But due to lack of information about further follow-up it can only be a matter of debate whether to call it AD or AMD. So if such cases are encountered follow-up evaluations are advisable to confirm the actual existing condition.

Apart from this Group II comprised of 8 subjects out of a total of 14, who had history of risk factor/factors. These were subjects 4, 6, 7, 8, 10, 12, 13 and 14. Subjects 4, 6, 10, 12 and 13 had history of Neonatal Jaundice but only 4 and 13 required blood transfusion and phototherapy respectively. Subject 8 demonstrated multiple risk factors in terms of history of mother having Chicken Pox in the first trimester of pregnancy, consumption of amniotic fluid by the subject at the time of delivery, low blood sugar level at the time of birth and febrile seizures at 1 month of age. Subject 10 also had the history of birth asphyxia to go with neonatal jaundice and subject 14 had birth asphyxia followed by seizures few days later. So in all, 55.55% of the subjects clubbed under the AD group (Group II) had history of severe degree of risk factor/factors pertaining to hearing loss. Thus, it suggests that severe degree of risk factors showed-up in high chances of auditory abnormality and hence such infants and toddlers must be considered for detailed audiological evaluation and LLR must be the part of the test battery to identify AD.

Group III consisted of 10 subjects 4 of whom had history of one or more risk factors that have been suggested to cause hearing loss. Subject 1 and 3 had history of neonatal jaundice to go with seizures in early infancy whereas subject 9 had history of premature delivery. Subject 8 was confronted with multiple risk factors that included birth asphyxia, low APGAR scores, bronchopneumonia, and seizures.

Six (1, 3, 6, 8, 9, and 10) out of the total of 10 subjects of this group showed presence of OAE in absence of any recognizable ABR peak even at 90 dBnHL in first recording. Based on the reports in literature that talk of absence of ABR in presence of OAE being the feature of AD (Starr et al., 1996), these subjects could have been diagnosed as having AD. But follow-up test results in these subjects (second follow-up for subjects 3, 6 and 10 and second and third follow-ups for subjects 1, 8 and 9) suggested other wise. These follow up recordings demonstrated presence of ABR and gradual progression towards normalcy in terms of peak latencies in those with two or three evaluations. These recordings also showed absence of LLR in presence of ABR in first recording itself which is a big indicator that the maturation has not fully occurred at the central level (at the level of cortex). The routine audiological testing (which includes BOA, ABR, OAE and immittance) in such cases, give the findings that are similar to the findings in cases with AD (i.e., absent ABR, near normal or slightly elevated BOA, A-type tympanogram and present TEOAE) at least in first evaluation. This can lead to a case being misdiagnosed as AD though it could be a case of delayed maturation (Auditory Maturational Delay or AMD). Comparison of the profiles of subjects in Table 2 and 3 can give a valuable introspection in this regard. The presence of LLR in absence of ABR gives an indication towards normal cortical functioning in lieu of sub-normal or abnormal peripheral (auditory nerve and brain-stem) functioning. Based on the earlier discussion with regard to table 2, the label of AD (based on peripheral-to-central course of maturation) can safely be put forth for such cases. Absence of LLR, when ABR is present (normal or abnormal) or when it is absent, shows a trend towards either AMD or severe hearing loss. But it would be better if cases with such findings are monitored with regard to the changes in auditory behavior at home and also through regular follow up (preferably at 3 months intervals) till a clearer picture of the condition evolves (preferably upto 2 to 3 years of age), more so if OAEs are absent.

In subjects 4 and 6 again, the absence of OAE can be accounted by B type tympanogram in both ears of both the subjects. These have been considered in this group based on the BOA findings which suggests near normal responses to FM tones and speech in subject 4 and shows a maturational course, indicated by improvement in response levels in second evaluation, in subject 6. So, if ABR is present at any level and LLR is absent in the first evaluation itself the case can be diagnosed as AMD if OAEs are found to be present. This receives support from the findings of 4 out of 6 subjects in table 3. These subjects (1 and 8 in third evaluation and 9 and 10 in second evaluation itself) showed the presence of LLR and also improvement in the ABR peak latencies with increase in age, thus, supporting the diagnosis of AMD which was established based on absence of LLR in the first recording session itself.

Rest of the subjects (2, 4, 5, and 7) had undergone only one evaluation. They had presence of ABR and OAE, except subject 4 (absent OAE), but LLR was absent. Absence of OAE could be accounted based on the tympanometry result which showed B-type tympanogram in subject 4. Presence of ABR and absence of LLR helped in the diagnosis of AMD in these subjects. However, follow-up is required to confirm this diagnosis. It has been seen that absence of ABR in the first evaluation could be misleading as there can be case of delay in maturation that lead to this phenomenon. This paradox can easily be solved by the inclusion of LLR in the test battery for the hearing assessment in infants and toddlers. Thus, it is strongly recommended to include LLR in the protocol for assessing hearing in infants and toddlers.

In Group IV (ABR absent- LLR absent) 3 out of a total of 13 subjects (23.08%) had history of one or more risk factors pertaining to hearing loss. Subject 1 had history of neonatal jaundice, subject 13 had birth asphyxia and subject 8 had a cluster of risk factors that included prenatal high blood pressure (at 7th month), premature delivery and low birth weight. All the subjects had BOA responses at much higher levels than normal hearing infants which correlated well with the findings that included absence of OAE, A-type tympanogram (except subjects 1, 2, and 9 in first recording and subjects 1 and 7 in second recording in both and left ear respectively) and no repeatable peaks in ABR and LLR recordings. Absence of LLR along with absence of OAE and ABR gives a fair indication towards subjects having permanent hearing loss. A conductive component in subjects 1, 2, 7 and 9 cannot account for absence of ABR at 90 dBnHL and that of LLR at 70 dBnHL. Thus, this group of subjects can be diagnosed as having permanent hearing loss.

The differentiation between the two- AMD Vs severe hearing loss can be easily accomplished based on TEOAE findings. If the TEOAEs are present it shows that the course of maturation may be slightly prolonged or delayed causing abnormality in ABR and LLR findings whereas absence of TEOAE would indicate abnormality at the level of cochlea too and hence severe hearing loss could be a better recommendation.

It can be concluded based on the results that LLR is an important tool in differential diagnosis of different conditions that are likely to be encountered when dealing with the hearing assessment of pediatric population of less than 2 years of age. The same can be clearly understood from the table 5.

Table 5: Different test results and diagnosis based on them

	BOA	**Immittance**	**OAE**	**ABR**	**LLR**	**Diagnosis**
1.	Normal	A-type	Present	Present	Present	Normal hearing
2.	Normal	A-type	Present	Absent	Present	AD
3.	Normal	A-type	Present	Absent/ Abnormal/ Present	Absent	AMD
4.	Abnormal	A-type	Absent	Absent	Absent	Severe hearing loss

The test results in case of AMD and AD are identical if LLR results are taken away. The Audiologist will, hence, find it very difficult to diagnose the condition or, more realistically, would have to wait until the maturation has fully occurred. This difficulty can be overcome when LLR results are included. The absence of LLR can be considered for the diagnosis of AMD whereas its presence can be termed as AD (based on the pattern of maturation which suggests peripheral to central course for it) based on presence of ABR at any level and absence of ABR even at high levels respectively. Thus LLR can prove to be of immense importance in differential diagnosis of AD and AMD. Also, though other routine audiological tests indicate hearing sensitivity within normal limits, still there may be a case of delayed maturation at higher centers (auditory cortex). This can be ruled out by presence of LLR. Not only that, LLR can also be used as a substitute for ABR to obtain threshold especially if ABR morphology is poor or if ABR is completely absent as in cases with AD. LLR can also be used as supporting tool for the diagnosis of severe hearing loss. Absence of LLR would indicate minimum or no signal reaching the auditory cortex that can evoke a cortical response.

The normative data was also established in the present study for ABR but due to lack in number of subjects a more careful usage of these findings is recommended. In case of LLR norms, large variability was observed across the subjects. So it is recommended that the interpretation of LLR wave be cautiously approached especially with regard to the absolute latency. It is also recommended that rather than looking at the latency it would be better to look for the presence or absence of LLR for the differential diagnosis of different conditions.

Implications of the study:

First and foremost, the study highlights the importance of LLR in differential diagnosis of AMD from AD and permanent hearing loss. So this brings out a solution to the paradoxical nature of hearing assessment in infants and toddlers. The study also suggests the use of LLR in threshold estimation especially if there is case of AD in which ABR is absent and thus implicates in early decision for cochlear implantation and avoid unnecessary psychological trauma to the parents if it is AMD. The study also tried to establish norms for ABR and LLR which could be used for arriving at conclusion if an infant or toddler is developing normally or not, though a careful use of the findings of the present study is recommended.

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