Language outcomes using hearing aids in children with auditory dys-synchrony

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Abstract

There is a considerable debate on the use of hearing aid in children with auditory dys-synchrony (AD). To current date there are no similar Studies which show the effect of acoustic amplification on outcomes on speech and language development in children with AD are found in the literature. Thus, in the present study three children diagnosed to have AD were recruited and subjected to hearing aid trial and fitting based on the late latency response thresholds. Ranging from 7 months to 41 months of intervention period, each child's language age was reassessed and compared with that of their pre-therapy language age. The comparison showed a good improvement in receptive age, though the children had moderate to severe degree of hearing loss. But their expressive age was significantly delayed irrespective of their age of identification and duration of intervention. Thus the delay in expression with a good amount of improvement in reception can be a characteristic of an AD child with amplification.

Introduction

Auditory neuropathy or auditory dys-synchrony (AD) is a clinical syndrome is characterized by the presence of oto-acoustic emissions (OAEs) and/or cochlear microphonics suggesting normal outer hair cell function in conjunction with absent or grossly abnormal auditory brainstem responses (ABRs).[1] The individuals who have been diagnosed to have auditory dys-synchrony exhibit varying clinical characteristics in pure tone average which ranges from normal to profound[2] and their speech recognition abilities show no correlation with the degree of hearing loss.[1] Difficulty in speech recognition extends to greater impairment in presence of noise due to poor temporal processing whereas; some individuals among them demonstrate a reasonably better perceptual ability even in presence of noise.[3,4] The predominant management options for children with AD are amplification devices, cochlear implantation (CI) and/or use of total communication. It is not surprising that there is a considerable debate on the use of hearing aid for children with auditory dys-synchrony, as they have intact outer hair cells and is opposed by two schools of thoughts. To begin with, the amplification by the hearing aid would be louder and distorting and can cause permanent damage to the hair cells.[5,6] There is less evidence to support these thoughts and many studies support benefit from the amplification devices in AD. There is a significant benefit from conventional hearing aids in children with AD and improved speech perception abilities.[7,4] The performance of aided children with AD is comparable with those of CI with AD,[8-10] with these results it implies that children with AD should not automatically be considered for cochlear implant candidates,[4] rather children with AD who don't benefit from amplification can be further considered. There is also a possibility of secondary loss of OAE in children with AD.[2] Hence conventional amplification can benefit pre-lingual AN at least once they have lost OAE. Over the years, due to increased understanding of AD, at least we have identified that the tools such as middle latency response and late latency response (LLR) correlate with their behavioral thresholds.[11,7] However, we still completely lack a reliable tool which can correlate with the amount of speech perception deficit that these clients' experiences. Thus, in the current scenario, it is difficult to choose between the recommended rehabilitations that is hearing aid and cochlear implant. An evidence based systemic review suggests that the effect of amplification in AD children has been systematically reported onlyin 4 out of 202 citations.[12] Out of these four studies, three studies have reported the pre and post treatment improvement in behavioral thresholds (pure tone audiometry) and all four studies examined the effects of amplification on speech perception.[2,9,7] This clearly shows that literature regarding outcomes on speech and language among children with AD is very limited even though it is rising to be a relatively common condition among the hearing impaired. Thus, it is important to explore the effectiveness of strategies that are much less invasive and less expensive than cochlear implants. There should also be sufficient evidence to hold up the possible speech and language outcomes when these children are provided with amplification devices. But to current date, there are no available studies which show the effect of acoustic amplification on outcomes on speech and language development in children with AD. Thus the present study aims to check for the language development in children with auditory dys-synchrony provided with amplification devices.

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Materials and Methods

AD is a highly heterogeneous disorder where the clinical picture can be quite different from patient to patient. The current study aimed at unveiling the effect of hearing aid on language development in this population, for this purpose we recruited three children who vary slightly in their age of identification nevertheless all were below the critical period for language development. Three children with presenting complaint of delayed speech and language and further diagnosed to have AD with the criteria of presence of OAE with absent ABR were chosen for the study. Informed consent was obtained from the parents prior to administration of test procedures. The demographic data for the three children are shown in Table 1.

Procedure

All the children were subjected to a battery of tests, including behavioral observation audiometry (BOA), tympanometry, reflexometry, OAE, ABR and LLR.

Behavioral tests

Behavioral observation audiometry and was done using the Grason-Stadler Inc. (Eden Prairie, MN, USA)-61 (GSI-61) Clinical Audiometer along with sound field system and ascending method was used to find out the lowest level at which the change in baseline behavior occurs in all the cases. Two clinicians (tester and observer) had to agree for correct responses to be considered as a reliable threshold. Warble tone from 250 Hz to 4000 Hz and live speech served as a stimulus.

Physiological and electrophysiological tests

Duly calibrated middle ear analyzer (GSI-Tympstar) was used for assessment of middle ear (Compliance between 0.3 and 1.75 mL and middle ear pressure between +/- 100 dapa). ABR and LLR were recorded in all the subjects following natural sleep in an acoustically and electrically shielded room. Electrode sites were cleaned using NuPrep skin preparation gel and electrodes were placed on sites with Ten20 conductive gel. Absolute and inter electrode impedance were within 2 kOhms, and the electrode montage used was Fz-A2 and the ER-3A insert earphones used to present the stimuli introduced a delay of 0.9 ms between the electrical signal at the transducer and the presentation of the acoustic stimulus at the ear canal. Obtained electroencephalogram was online filtered between 100 Hz and 3000 Hz for ABR and 1-30 Hz for LLR with artefact rejection of 23.8 μV and was epoched between -12 ms and + 12 ms for ABR and overall 600 ms for LLR. For ABR total of 1000 stimuli were presented at a rate of 11.1/s with alternating, rarefaction and condensation polarity and recording was repeated three times at each intensity constituting three averaged waveforms of 1000 stimuli. Wave obtained from rarefaction and condensation was used to identify the cochlear microphonics. Whereas for LLR a total 500 stimuli were presented at a rate of 1.1/s with alternating polarity and recording was repeated three times at each of the intensity in 10 dB decrement from 90 dBnHL till the threshold. All the subjects were stimulated monaurally in both ears. ABR and LLR were recorded using the IHS Smart EP Software, Version: 3.94.

Otoacoustic emissions

Transient evoked oto-acoustic emissions (TEOAE) was recorded in both the ears in all the cases using ILO292 DP Echo port with V6 software. An SNR of 6 dB and above was considered as the presence of OAE.

Hearing aid fitting

Hearing aid trail and fitting was done for all the three children as they exhibited hearing loss in both LLR and behavioral responses. All the participants procured DSP based hearing aids and were first fitted using DSL-I/O in NAOH software with HI-Pro interface. The target gain was set based on the behavioural thresholds. Hearing aid verification was done using functional gain measurement by observing the difference between unaided and aided response for both warble tones and speech stimuli, using the Grason-Stadler Inc.-61 (GSI-61) Clinical Audiometer with the sound field system. The hearing aid gain was further fine tuned so that the aided responses were within speech spectrum. Following the hearing aid fitting all the three children were recommended for speech and language intervention.

Language assessment

Receptive and expressive language skills were assessed by administering the diagnostic tool assessment of language development (ALD) which measures the language development from birth to 7 years 11 months. In order to identify the receptive and expressive age the child was initially observed and the parents were also asked questions regarding the skills that he/she have achieved. The assessment was done during the pre and various post therapy intervals. The average testing time ranged from 20 to 40 min during each of the assessment sessions. All the children are continuing with speech and language intervention with hearing aids and parents are highly motivated.

Results

In all the 3 children TEOAE and cochlear microphonics were present in both ears; with bilateral absence of acoustic reflexes and ABR. In Child I, BOA responses were between 75-85 dBHL for 500 Hz, 1 kHz, and 2 kHz. LLR was recorded up to 70 dBnHL bilaterally, suggestive of severe hearing loss. Prior to the intervention the baseline language age was 0.6-0.11 years of reception and 0.0-0.5 years of expression. Auditory verbal therapy was carried out in the same center for the duration of 2.4 years. The child's present language age is 3.0-3.5 years of reception and 2-2.5 years of expression. For Child II, BOA responses were 60-65 dBHL for 500 Hz, 1 kHz and 2 kHz. LLR was recorded in both ears up to 60 dBnHL. Prior to the intervention the baseline language age was 0.6-0.11 years of reception and expression. Auditory verbal

Table 1. Demographic data of the three children.

	Child I	Child II	Child III
Date of birth	7/3/09	25/8/07	23/11/09
Deprivation period	1 year 2 months	2 years 4 months	2 years 3 months
Age of identification	1 year 10 months	2 years 6 months	2 years 3 months
Present chronological age	3 years 6 months	5 years 1 month	2 years 10 months

therapy was carried out for the duration of 2.4 years. The child's present language age is 5.0-5.5 years of reception and 2.0-2.5 years of expression. For child III, BOA couldn't be done and LLR was recorded up to 50 dBnHL, suggesting bilateral moderate hearing loss. Prior to the intervention the baseline language age was 2-2.5 years of reception and 1-1.5 years of expression. Auditory verbal therapy was carried out for the duration of 7 months. The child's present language age is 2.6-2.11 years of reception and 2.0-2.5 years of expression (Table 2).

Discussion

The current study intended to follow up the language development in children who were diagnosed to have AD. Prior to the intervention, assessment was done with ALD and it was identified that for all the 3 children's language development was significantly delayed, following which all the 3 children who exhibited hearing loss based on the LLR degree of hearing loss were fitted with the hearing aid using prescriptive formula DSL-I/O. Ranging from 7 months to 41 months of intervention period their language age was reassessed and compared with that of their pre-therapy language age. The differences in reception age between the baseline and post intervention for the Child I, II, and III were 3 years, 5 years, and 6 months respectively. Differences in expressive language age were 1 year for Child I and Child III; whereas child II had 1 year 7 months. The Child II was the first child we identified and was also followed up for a longer time compared to others. This helped to see another important factor whether is there any slow down or ceiling in the prognosis, however it was not evident and as per the last follow up they continue to progress. The comparison showed (Figure 1) a good improvement in receptive age, though the children had moderate and severe degree of LLR thresholds. But it is also evident that their expressive age was significantly delayed irrespective of their age of identification and duration of intervention.

In AD there is abnormal transmission of impulses by the auditory nerve resulting in deviant neural coding; in spite of this we could observe that there is age adequate improvement in receptive skills in these pre-lingual AD children. This we could comprehend with the understanding of the nature of language development that is; in order to comprehend an acoustic signal child may not require a very precise acoustic cue where even a degraded signal is sufficient enough to understand a message and the repeated exposure to the same stimuli would have formed a deviant auditory image facilitating the child to associate with the respective symbols. However, their expressive ability never matched up to their peers in post therapy assessment (Child I: 1.5 years; Child II: 1. 7 years; Child III: 1.5 years), as expression relies on the receptive abilities which doesn't provide sufficient information for the child to perceive and produce speech as that of the normal developing child. This could also serve as the reason for their imprecise vocal utterances and less intelligibility in the speech of children with AD.

There is a positive correlation observed between the degree of hearing loss and the progress in the language development. As observed in Table 3, Children with moderate hearing loss observed to be performing better in language development than severe hearing loss child. This indicates that amplification provided to these 3 children with poor LLR thresholds do facilitates them to overcome the hearing, but doesn't alter the deviated pattern of the neural coding in them, making the degree of hearing loss to correlate with the language development. Thus delayed expression ability can be considered as the predictor for

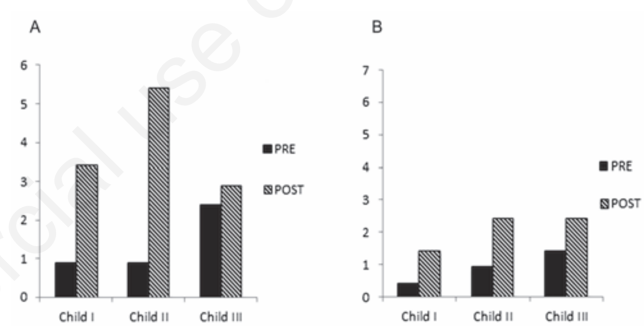

Figure 1. A) Pre and post therapy comparison of receptive language age; B) pre and post therapy comparison of expressive language age.

Table 2. Findings of the audiological evaluation.

	Child I	Child II	Child III
BOA	75-85 dBHL at 500 Hz, 1 kHz and 2 kHz	60-65 dBHL at 500 Hz, 1 kHz and 2 kHz	Could not be done
Cochlear microphonics	Present	Present	Present
Acoustic reflex	Absent	Absent	Absent
ABR	Absent	Absent	Absent
LLR	Up to 70 dBnHL	Up to 60 dBnHL	Up to 50 dBnHL
Impression	Bilateral severe hearing loss	Bilateral moderatehearing loss	Bilateral moderate haring loss

BOA, behavioral observation audiometry; ABR, auditory brainstem response; LLR, late latency response.

Table 3. Pre and post therapy language age.

	Pre therapy		Intervention duration	Post therapy		Progress	
	RLA	ELA		RLA	ELA	RLA	ELA
Child I	0.6-0.11 years	0.0-0.05 months	2 year 4 months	3.0-3.5 years	1.0-1.5 years	3 years	1 year
Child II	0.6-0.11 years	0.6-0.11 months	3 years 5 months	5.0-5.5 years	2.0-2.5 years	5 years	1 year 7 months
Child III	2.0-2.5 years	1.0-1.5 years	7 months	2.6-2.11 years	2.0-2.5 years	6 months	1 year

RLA, receptive language age; ELA, expressive language age.

the degree of the AD and with the above evidence we could conclude that children with AD can be provided with amplification with the expectation for age adequate progress in the receptive language age but not in the expressive language age.

Conclusions

Delay in expression with a good amount of improvement in reception can be a characteristic of an AD child with amplification. The above findings can be implemented for counselling parents of children with AD regarding the expected outcomes in language if their children are fitted with hearing aids.

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