

A Study of Various Factors Affecting Habilitation Outcome in Children with Severe to Profound Hearing Loss

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Citation

D G Shrestha, S Mahajan. *A Study of Various Factors Affecting Habilitation Outcome in Children with Severe to Profound Hearing Loss*. The Internet Journal of Otorhinolaryngology. 2014 Volume 16 Number 1.

Abstract

Aims: A study was conducted to study the various factors affecting the habilitation outcome in children with severe to profound hearing loss.

Materials and methods: The study was conducted at Command Hospital (Eastern Command), Kolkata over a period of two years.

It was a pilot study including 25 pre-lingually deaf children, aged between 0-14 years, randomly selected, of both sexes, with severe to profound bilateral hearing loss. All children with associated co-morbidities like neurological disorders or multiple medical problems were excluded from the study.

We formulated a novel scoring system. The initial evaluation was done on a total score of 14 followed by detailed evaluation of the children, initiation of amplification and speech and language therapy. Follow up was done at 3, 6, 9 and 12 months respectively. During each follow up, they were evaluated on a total score of 17.

The results were analyzed statistically. P-values of less than 0.05 were considered significant.

Results: All the factors appeared to play a cumulative role in the final habilitation outcome, with the results being statistically significant. The most important factor appeared to be the type and degree of amplification used.

Our unique scoring system also showed promise to evolve into a tool to assess prognosis of children following intervention based on their initial evaluation scores

Conclusion: Our results clearly point out the importance of universal neonatal hearing screening, early diagnosis and earliest possible adequate intervention fortified by parental care & training by skilled audiologists.

INTRODUCTION

It is difficult to imagine a deaf life, as the use of the ear is more instinctive than that of the eye.

In normally developing children, the language of the home is acquired through the channel of hearing. For profoundly deaf children, this normal acquisition is disrupted, leading to the likelihood of communication, speech and language delay, which may result in underachieving educationally and later in employment [1].

The presence of a child with hearing loss is often linked with psychosocial stress in parents and other family members [2].

Children also suffer from social isolation as a result of hearing impairment. There is some evidence that access to

services for hearing loss are dependent on social class and income with those in developing countries and in the poorer sectors of the society having difficulty in accessing the services they need.

Amplification in the form of hearing aids or cochlear implants is the mainstay for the treatment of permanent childhood hearing impairment (PCHI) in most cases. Early audiological assessment and provision of hearing aids at the youngest possible age (as young as four weeks in some cases) makes for the best possible outcome. Supplying, fitting and after-care of young children are skilled professional tasks and an appropriately trained and experienced audiologist is essential [1].

Hearing impaired people, children particularly, form a

significant part of the population of India. In the face of poverty and lack of adequate facilities, a significant number of children are deprived of early diagnosis & prompt intervention. These children fail to acquire normal hearing, speech and language. Hence, most resort to communication by sign language.

Sign language is different around the world. In a multi-linguistic country like India, the language differs in every part of the country, making communication by sign language even more difficult. At the present time, most hearing impaired people in India use a combination of body language and a type of sign language combined together. It doesn't take much thought to realize that this is hardly an effective means of communicating and the hearing impaired population remains isolated, even among the individuals who make up the hearing impaired groups.

A study was therefore conducted to study the various factors affecting the habilitation outcome in children with severe to profound hearing loss.

MATERIALS AND METHODS

A study was conducted at the ENT Department of Command Hospital (Eastern Command), Alipore, Kolkata, to evaluate the various parameters that affect the habilitation outcome in children with severe to profound bilateral hearing loss. The objective was to pinpoint the significant and important factors which substantially affect the habilitation outcome and the degree of impact of these factors.

It was a pilot study including 25 children with pre-lingual hearing impairment. The children were aged between 0-14 years, randomly selected, of both sexes, with severe to profound bilateral hearing loss. All children with associated co-morbidities like neurological disorders or multiple medical problems were excluded from the study. The study was conducted over a period of two years.

There are a large number of studies on multivariate predictors for cochlear implantation outcome in postlingually deafened adults, but so far there are no studies evaluating the factors that affect the habilitation outcome in prelingually hearing impaired children. Hence, we formulated a novel scoring system. The aim was to develop an integrated, qualitative and easy bedside method of assessing the prognosis prior to intervention, based on initial evaluation scores. Review of literature revealed that no such

scoring system had been used till date. The factors considered during the initial evaluation are the common factors that affect the habilitation outcome of these children and were decided upon, after thorough review of literature [3]. Factors included to assess the habilitation outcome, are the common parameters which help to assess the age-appropriate development of a child. These factors are also used in various protocols worldwide, to evaluate the level of performance in children following cochlear implantation [4] [5].

The initial evaluation was done on a total score of 14 and included the following parameters: motivation of the parents, age at first reporting or intervention, degree of hearing loss, adequacy of hearing amplification received, adequacy of speech and language therapy received and presence or absence of high risk factors or syndromic illness.

The children then underwent a complete evaluation which included a detailed history (with special emphasis on developmental history, presence or absence of risk factors and family history), ENT evaluation, radiological evaluation (MRI and HRCT Temporal bone) and audiological evaluation. Otoacoustic Emission and Auditory Brainstem Response was done in children aged 0-2 years, Behavioural Observation Audiometry and Auditory Brainstem Response done in children aged 3-4 years and Pure Tone Audiometry and Auditory Brainstem Response were done in children aged 5 years and above. Impedance audiometry was done in children suspected to have middle ear pathology. Auditory Steady State Response was done in children with severe degree of hearing loss prior to hearing aid fitting. This was followed by a thorough systemic, psychological, speech and language assessment. In older children, educational assessment was also done.

Thereafter, the type & nature of hearing amplification necessary was assessed. Adequacy of hearing amplification was checked in children who already had some form of amplification. The children received amplification, from our institution, in the form of digital, binaural Behind The Ear (BTE) hearing aids and suitable candidates underwent cochlear implantation, only after 3 months of hearing aid trial. The children were started on speech and language therapy and followed up at 3, 6, 9 and 12 months respectively.

During each follow up, they were evaluated on a total

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score of 17, on the following parameters: Aided audiogram, ability to identify Ling's 7 sounds, intelligibility of speech, attainment of age appropriate reception and expression, reading and writing skills and social development.

The results were then analyzed statistically. A chi square test was performed to test the significance. P-values of less than 0.05 were considered significant.

The following formulated scoring system was used:

A) Initial Evaluation (Total Score: 2-14)

Factors affecting outcome	Total score (2-14)					
	0	1	2	3	4	
Motivation of the parents	No motivation	Intermediate motivation	High motivation			
Age at reporting or intervention		>7 yrs	5 to 7 yrs	3 to 4 yrs	0-2 yrs	
Degree of hearing loss		>90 dBHL	80 to 90 dBHL	70 to 80 dBHL		
Adequacy of hearing amplification used	No amplification	Inadequate amplification	Adequate amplification			
Speech and language therapy	Not received	Inadequately received	Adequately received			
Risk factors/ syndromic illness	Present	Absent				

B) Evaluation of Habilitation Outcome (Total Score: 0-17)

Habilitation outcome measures	Total Score (0-17)						
	0	1	2	3	4	5	6
Aided audiogram (location on speech spectra)	Located outside	Located in the lower part	Located in the middle	Located in the upper part			
Identification of Ling's 7 sounds	No detection of sounds	Detection of Ling's 7 sounds	Level 1 + identification of Ling's 7 sounds	Level 2 + discrimination of vowels	Level 3 + discrimination of consonants	Level 4 + recognition of closed set sentence test	Level 5 + recognition of open set sentence test
Reception, Expression (age appropriate)	None present	Any one present	Both present				
Intelligibility of speech production (age appropriate)	Speech not clear	Speech understood with difficulty by any listener	Speech understood by any listener				
Reading and writing skills (age appropriate)	Cannot read or write	Can either read or write	Can both read and write				
Social development (age appropriate)	Absent	Present					

Habilitation Outcome was classified as Poor: Scores 0-5, Average: Scores 6-11 & Good: Scores 12-17

RESULTS

Clinical profile:

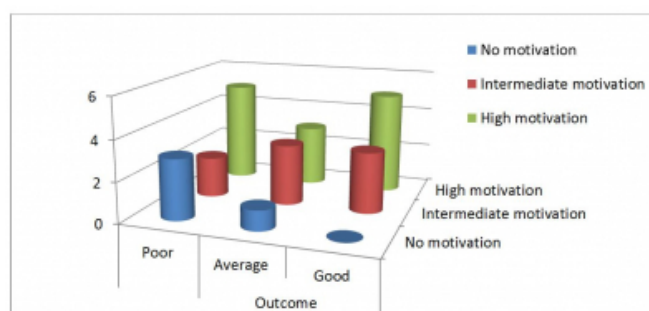
1. Age distribution: The maximum number of patients were in the age group 0-2 years (52%) followed by >7 yrs (20%). The youngest patient was 10 months old and the oldest was 14 years old.
2. Sex Distribution: Out of the 25 children included in the study, 12 were females (48%) as compared to 13 males (52%).
3. Degree of hearing loss: Majority of the children had more than 90 dB hearing loss i.e. 16 out of 25 (64%). 8 children had hearing loss between 80-90 dB (32%) and the least number of children had hearing loss between 70-80 dB i.e. 1 out of 25 (4%).
4. Nature of amplification provided: 17 out of 25 children (68%) were fitted with hearing aids while only 8 children (32%) who satisfied the candidacy criteria underwent cochlear implantation after 3 months of hearing aid trial.

1. Analysis of result:

a. Role of motivation of parents: Out of the 8 children with good outcome, 5 had parents with high motivation (62.5%). 4 children had parents with no motivation at all, of whom 3 had poor & 1 had average outcome. Fig. (1)

Figure 1

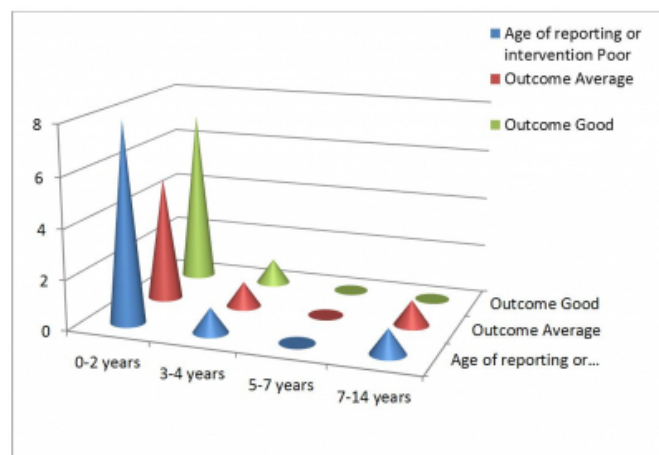
Role of motivation of parents on the habilitation outcome



b. Role of early reporting or intervention: Out of 20 children, who reported before 2 yrs, 35% had good, & 25% had average outcome, comprising a total of 60%. None of the children who reported after 7 yrs attained comparable results. Fig. (2)

Figure 2

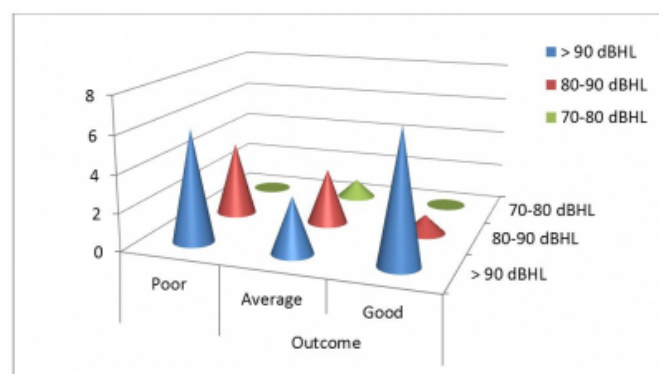
Role of age of reporting on the habilitation outcome



c. Role of degree of hearing loss: 1 child, in spite of having 70-80 dBHL and fitted with hearing aid, had average outcome while 43.75% of the children with profound hearing loss did very well. These children were all implantees. Fig. (3)

Figure 3

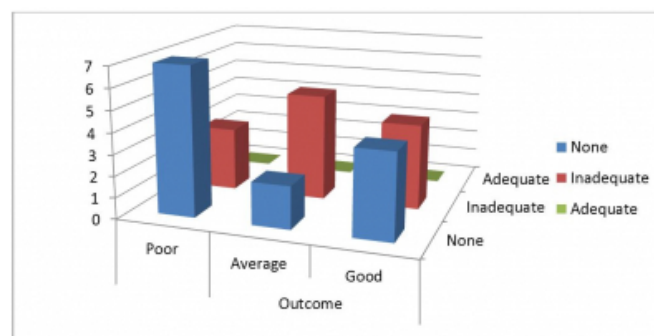
Role of degree of hearing loss on the habilitation outcome



d. Role of adequacy of hearing amplification: This parameter directly influenced the outcome. Out of the children with no amplification at the beginning of the study, 53.84% had a poor outcome while all the children with some form of amplification, even if inadequate, had better outcome. Following intervention, implanted children had the best outcome. Fig. (4)

Figure 4

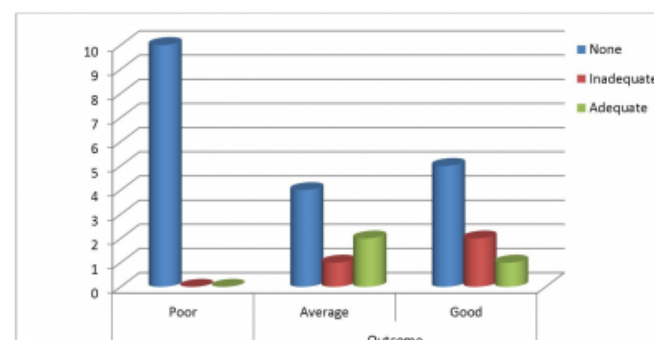
Role of adequacy of amplification on the habilitation outcome



e. Role of speech and language therapy: The earlier the initiation, the better the results. Out of the children with no form of therapy, 53.62% had a poor outcome. Those with some form of speech & language therapy had average or good results. (Fig. 5)

Figure 5

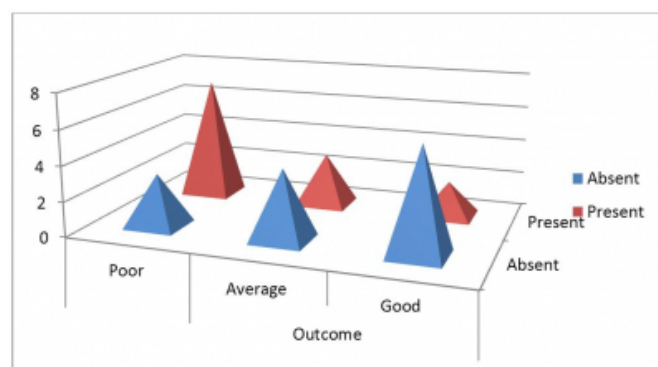
Role of speech & language therapy on the habilitation outcome



f. Role risk factors or syndromic illness: Children with some high risk factor had a poorer outcome compared to children who did not. 46.15% children with no risk factors did very well & 30.76% did averagely compared to 16.66% & 25% of those who had a positive history. The result was statistically significant (p-value: 0.04735). (Fig. 6)

Figure 6

Role of risk factors or syndromic illness on the habilitation outcome

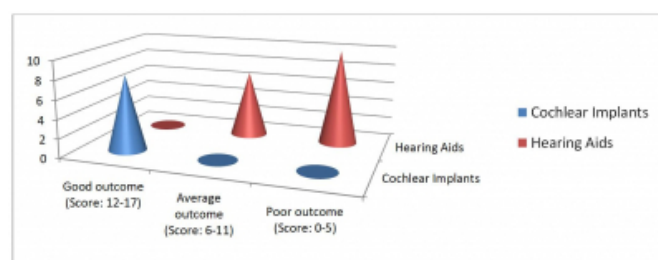


g. Final result: When the role of cumulative effect of the factors studied was evaluated, a statistically significant relationship (p-value: 0.01360) was seen between the factors being evaluated and the habilitation outcome. Majority of the children with high initial scores went on to have good outcome scores as well. However, the final outcome depended on the cumulative effect of all the factors.

h. Role of nature of amplification: The most important influencing factor was the adequacy amplification. Cochlear implantation significantly affected the habilitation outcome and the children showed marked improvement in all the parameters. The result was statistically significant (p-value: 0.00009345) (Fig.7)

Figure 7

Effect of adequacy of amplification on the habilitation outcome



DISCUSSION

Approximately one child in 1000 is born with a bilateral permanent childhood hearing impairment (PCHI). This is defined as confirmed bilateral hearing impairment exceeding 40 dBHL (hearing level) (average of pure tone thresholds at 0.5, 1, 2 and 4 kHz in the better hearing ear [1].

About 60 percent of these children have a moderate (41-60 dBHL) hearing loss, while the remainder have a severe

(61-80 dBHL) or profound (>81 dBHL) loss [6].

The prevalence of PCHI increases with age, suggesting that a further one in 1000 children develop acquired or progressive hearing impairment [7].

It is now known that the critical language learning period of a child is from birth to about three and half years of age. A landmark study demonstrated that children whose hearing losses were identified by 6 months of age demonstrated significantly better receptive and expressive language skills than did children whose hearing losses were identified after the age of 6 months [8].

Language acquisition is very complicated. The complexity of learning a language arises from a synthesis of many influences and activities that enable a child to become linguistically engaged. Children learn language by developing and assembling together four systems of skills. The pragmatic, phonology, semantic and syntax are separate but inter related systems that comprise the foundation of language acquisition [9, 10]. Except for the semantic system, acquisition of each of these systems is subject to a critical period after which full mastery of language is unlikely [10].

Luterman (1999) [11] maintained that the self-esteem of the parents, particularly, the mother is the crucial key to the child's success, and that all clinical endeavors should be devoted to empowering and increasing parents' self-confidence. Our study also reflected the role of parental motivation in the outcome of the children. It was evident that parental motivation was required for a good outcome. Of the children with best outcome, 62.5% had motivated parents. However, the p-value was calculated to be 0.1755 and the association was not found to be statistically significant.

N. Daneshmandan and P. Borghei et al [12] conducted study to emphasize on the importance early intervention following early detection. 9 severe to profound hearing impaired children below 2 years old were selected and given aural habilitation and speech therapy. Speech intelligibility assessment showed acquisition of language skills but a delay of 2 to 3 years in comparison with normally hearing children of the same age. Our study also had similar results. Out of the 20 children who reported before 2 years of age, 65% had good or average outcome scores. Early reporting was essential to provide auditory stimulation but outcome was best only when supported by adequate intervention, i.e. cochlear implant in our case. However, a p-value of 0.8 was obtained showing no

statistical significance.

The habilitation outcome significantly is affected by the degree of hearing loss, the nature and adequacy of hearing amplification provided. Compared to hearing aids, use of a cochlear implant has a dramatic impact on the linguistic competence of profoundly hearing-impaired children. The outcome also depends on the duration of time after the initiation of amplification use. Studies have shown that children with profound hearing loss achieve unprecedented levels of speech perception skill 4 to 7 yr after cochlear implantation. Ann E Geers, Johanna G Nicholas, Allison L Sedey et. al [5] in a study investigated the factors contributing to speech perception outcomes in children with prelingual deafness after 4 to 7 yr of multichannel cochlear implant use. More than half of the children in this sample with average learning ability produced and understood English language at a level comparable with that of their hearing age mates. Such mature language outcomes were not typical of children with profound hearing loss who used hearing aids. In our study too, age-appropriate auditory skills were all achieved by implanted children in spite of more than 90 dB hearing loss. The children with less severe hearing loss, but with inadequate amplification in the form of hearing aids could not attain comparable results as their implanted peers in the outcome.

Our study also revealed the importance of speech and language therapy in the outcome, but only after provision of adequate amplification. A prospective longitudinal study conducted to assess the habilitation outcome in a group of consecutively implanted children over 10 years after implantation indicated that cochlear implant centres need the structure and funding to provide long-term support, counselling, audiologic follow-up, rehabilitation, and device monitoring to implanted children [13]. During initial evaluation, 12% children had received adequate, 12% inadequate and 76% had received no form of speech and language therapy. After intervention, it was found that all the children with age appropriate auditory, speech and language and academic skills had been implanted and received regular speech and language therapy. Speech and language therapy in the absence of adequate amplification didn't have any significant effect. Statistical analysis gave a p-value of 0.16, i.e. there was no statistical significant effect of mere speech and language therapy on the final outcome unless the child received adequate amplification.

Kaga, Kimitaka; Shindo, Mitsuko; Tamai, Fumi; Tanaka, Yoshisato [14] conducted a study consisting of hearing-impaired infants with and without multiple handicaps. The auditory behaviours of the hearing-impaired children with no other problems showed constant changes with age after hearing aid fitting while majority of the children with multiple handicaps, showed considerable improvement in auditory behaviours. The authors recommend early diagnosis of deafness and the early fitting of hearing aids in multiply handicapped children even if a child's neurological or mental status is poor. Our study showed a statistically significant relationship between the absence of risk factors or syndromic illness and the habilitation outcome (p-value of 0.04735). 75% of the children with aided audiogram in the upper part of speech banana & recognition of at least closed set sentence, 66.66% of those with age appropriate reception and expression, 87.5% of children with intelligible speech, 100% children with both reading and writing skills had no history of any risk factor.

The final analysis was the comparison of the total habilitation outcome score with respect to the total initial evaluation score. Children with high initial scores performed better than children with low scores who had a poor outcome. Statistical analysis gave a p-value of 0.01360. Thus it was concluded that a statistically significant relationship exists between the cumulative effect of the factors studied and the habilitation outcome.

However, the individual role of each factor could not be statistically proven, due to small sample size, short follow up period and presence of confounding factors like maternal education, nature of hearing aid and its adjustments, and difference in speech and language therapists. The study however indicated that they definitely have a significant role to play.

The other significant result of the study was that in the presence of similar initial factors, children with cochlear implants showed very fast improvement as early as 1 year after implantation, compared to their hearing aid using peers. Their results were even comparable to their normally hearing peers. In this short span of 1 year, 2 children were even admitted to a regular school. Strong statistical significance was proved with a calculated p-value of 0.00009345.

CONCLUSION

Our study therefore evaluated the factors affecting the

habilitation outcome of children with bilateral severe to profound hearing loss and their individual role.

The results were as per the literatures, with amplification used being the most important factor affecting the habilitation outcome. However all the factors played a cumulative role in the final habilitation outcome.

Our unique scoring system also showed promise to evolve into a tool to assess prognosis of children following intervention based on their initial evaluation scores.

Our results clearly point out the importance of universal neonatal hearing screening, early diagnosis and earliest possible adequate intervention fortified by parental care & training by skilled audiologists.

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