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AUDITORY VERBAL OUTCOME IN PATIENTS WITH MONDINI DEFORMITY FOLLOWING COCHLEAR IMPLANTATION AT ONE YEAR POST-HABILITATION

Objective:

To assess the auditory verbal outcomes after one year of habilitation based on Categories of Auditory Performance (CAP) and Speech Intelligibility Rating (SIR) scores in Mondini deformity following Cochlear Implantation (CI).

Material and methods:

This is a retrospective comparative analysis of 13 implantees with Mondini deformity (Group A) and 13 implantees with normal cochlea (Group B) available at Department of Implantation Otology, Madras ENT Research Foundation, Chennai, India from November 2005 to December 2012. One year post habilitation auditory verbal outcomes of the selected patients were collected from habilitation centre run by the same institute. Data was analyzed using the Student t test.

Result:

The mean CAP score was 5 ± 1.35 (range 2-7) in group A and 5.23 ± 1.25 (range 5-6) in group B which was statistically not significant ($p=0.56$). Similarly the SIR scores varied from 1 to 5 in both the groups. The mean of SIR score was 3.08 ± 1.25 in group A and 3.54 ± 1.26 in group B. This was also statistically not significant ($p=0.36$).

Conclusion:

The auditory verbal outcome for the patients with Mondini deformity is similar to those with normal cochlea following CI. Hence CI is successful in patients with Mondini deformity, although a higher degree of variability is noted among these candidates.

Key words: Cochlear Implantation, Mondini deformity, CAP score, SIR score

INTRODUCTION:

The incidence of congenital hearing loss is increasing world wide. According to WHO report it is nearly 2 - 3 per 1000 live births. It is mainly because of failure of development of inner ear due to various causes like genetic, maternal medication, TORCH infection etc. Only 15% of the congenital hearing loss are due to radiologically demonstrable inner ear malformations¹. Mondini deformity is the most commonly described congenital cochlear abnormality and it has been classified under Incomplete Partition type II by Sennaroglu and Saatci². The abnormality is thought to result from a relatively late insult during the 7th week of embryological development, when most of the inner ear has already formed³. Earlier insults result in more severe abnormalities like Michel deformity, Cochlear aplasia and Cochlear hypoplasia. The use of cochlear implantation (CI) to treat hearing loss in patients with Mondini deformity has been increasingly successful. A lot of reports and results of cochlear implantation in this deformity have been published.

There are various methods of assessment of the result of cochlear implantation are available. Categories of Auditory Performance (CAP) and Speech Intelligibility Rating (SIR) scale were used to assess the outcome in current study. CAP score 4 consists of a nonlinear, hierarchical scale of auditory receptive abilities. (Table 1) Similarly, SIR scale 5 is a time effective global outcome measure of speech production, which might be recognizable to the listener (Table 2). The objective of the current study was to assess the auditory verbal outcomes after one year habilitation based on CAP and SIR scores in Mondini deformity following CI at Madras ENT Research Foundation, Chennai.

Tab. 1: Category of Auditory Performance (CAP)

Category	Criteria
7	Uses telephone with known speaker
6	Understands conversation without lip reading
5	Understands common phrases without lip reading
4	Discrimination of speech sounds without lip reading
3	Identifies environmental sounds
2	Response to speech sounds
1	Awareness of environmental sounds
0	No awareness to environmental sounds

Tab. 2: Speech Intelligibility Rating (SIR)

Category	Criteria
5	Intelligible to all listeners
4	Intelligible to a listener who has little experience of a deaf person's speech
3	Intelligible to a listener who concentrates & lip-reads
2	Intelligible speech is developing in single words
1	Unintelligible

MATERIAL AND METHODS:

The present study is based on the retrospective comparative analysis of record available at Department of Implantation Otology, Madras ENT Research Foundation, Chennai, India. All the records of CI from November 2005 to December 2012 were revised. The records of all the patients less than or equal to seven years of age with Mondini deformity operated for Cochlear Implant were analyzed (Group A). The records of 13 patients of matched age group with normal cochlear structure with prelingual profound deafness operated for CI were taken as control group (Group B). The preoperative reports of audiological tests and reports of CT/ MRI scan were obtained from the data-base recording of CI surgery. Post operative auditory verbal outcomes of the selected patients were collected from habilitation centre run by the same institute. The collected data was analyzed using the Student t test. Because of the retrospective design of the study and anonymously kept patients' data, it was exempted from approval by the institutional review board.

RESULTS:

Total 16 patients with Mondini deformity were implanted in-between November 2005 to December 2012. Three patients were excluded as they were more than 7 years. Out of 13 implantees with Mondini deformity 4 (30.77%) were male and 9 (69.23%) were female. In group A one implantee achieved category 7 and one achieved only category 2 after 12 months of habilitation, however all the implantees achieved more than 5 in group B. The mean CAP score was 5 ± 1.35 (range 2-7) in group A and 5.23 ± 1.25 (range 5-6) in group B which was statistically not significant ($p=0.56$). Similarly the SIR scores varied from 1 to 5 in both the groups. The mean of SIR score was 3.08 ± 1.25 in group A and 3.54 ± 1.26 in group B. This was also not significant statistically ($p=0.36$).

Tab. 3: CAP scores after 12 months of habilitation

CAP score	1	2	3	4	5	6	7
Group A (n=13)	0	1	1	0	8	2	1
Group B (n=13)	0	0	0	0	10	3	0

Tab. 4: SIR scores after 12 months of habilitation

SIR score	1	2	3	4	5
Group A (n=13)	0	4	1	3	1
Group B (n=13)	1	2	2	5	3

DISCUSSION:

Mondini deformity was first described in Latin by Carlo Mondini, an Italian anatomist in 1791. This is an anatomical deformity of cochlea with consists of a triad of abnormal cochlea with only one and half turns with cystic apex, an enlarged vestibule with normal semicircular canals and enlarged vestibular aqueduct. CI is considered as a treatment for profound hearing loss in patient with Mondini deformity. Miyamoto⁶ in 1986 reported first CI in 4 year boy with this deformity. Silverstein H⁷, in 1988, had claimed as the first case report of successful multichannel CI in an adult of 31 year old with the same deformity. At that point, due to the unavailability of various diagnostic modalities, the procedure was mainly experimental and outcomes could not be predicted. After that a series of studies has been published on it. Buchman et al⁸ retrospectively reviewed the cases of CI in abnormal cochlea. Out of 28 abnormal cochleas 9 were of Mondini deformity and found the excellent result as in normal cases. Almost similar to current result was published by Li Yx et al⁹ in 15 cases of Mondini deformity among 300 patients, which showed no significant difference in the result of cochlear implantation in Mondini deformity in comparison with normal cochlea group. Similarly Daneshi A,¹⁰ in 1997, reported successful Cochlear Implantation in 5 patients with Mondini deformity. Category of Auditory Performance (CAP) consists of a nonlinear, hierarchical scale of auditory receptive abilities, with the lowest level describing no awareness of environmental sounds and the highest level being represented by the ability to use a telephone with a known speaker. The CAP is not a closed-set laboratory type test but a measure of everyday auditory performance and thus reflects the “real-life” progress of children in developing the use of audition. Speech Intelligibility Rating (SIR) is used to measure the speech intelligibility of the implanted children. The SIR is a five-point hierarchical scale describing various degrees of speech intelligibility ranging from unintelligible speech to speech that is intelligible to all listeners. SIR measures progress in speech intelligibility from before the first words until connected speech is established. The inter-observer reliability of CAP scale and SIR scale has been formally confirmed, and is being used widely for the assessment of auditory performance and speech and language development. That is why CAP and SIR were used in current study to assess the outcome of CI.

Because of loss of neural plasticity, a child shows poor speech development after the age of seven years. As the age at implantation is the proved variable affecting the outcome of CI, only younger children were included in current study. Although the outcome of CI in Mondini deformity and normal cochlea has no significant difference, CAP score of two implantees in group A achieved below category 3 and none of the implantee in group B achieved below category 5 after 12 months of habilitation training. This reflects that there may be the chance of poor outcome in Mondini deformity following CI. Therefore development of speech perception and subsequent language development in patients with Mondini deformity may vary so the parents must be counseled regarding this issue before implantation.

CONCLUSION:

There is no significant difference in auditory verbal outcome after one year of habilitation training in cochlear implantees with Mondini deformity in comparison with normal cochlea. Hence cochlear implantation could be performed successfully in patients with Mondini deformity, although a higher degree of variability is noted among these candidates.

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