

The relationship between aetiology of hearing loss and outcome following cochlear implantation in a paediatric population

P.G. O'SULLIVAN, S. ELLUL, R.C. DOWELL,
B.C. PYMAN and G.M. CLARK

*Cochlear Implant Clinic
University of Melbourne, East Melbourne (AUS)*

SUMMARY

Eighty-eight children who underwent cochlear implantation at the University of Melbourne Cochlear Implant Clinic are reviewed. The aetiology of the hearing loss is classified and is compared to their best level of speech perception performance. The group whose hearing loss was not congenital in origin performed better than those who were congenital in origin. Of those whose hearing loss was congenital in nature those with rubella appeared to perform best.

INTRODUCTION

Many factors influence the performance of deaf children following cochlear implantation. This would include the age of onset and duration of their deafness before diagnosis, the severity of their deafness, the aetiology of their deafness and whether it results in associated cognitive defects. Other factors would include the level of speech development prior to cochlear implantation, age of cochlear implantation, technical aspects of the procedure e.g. depth of insertion of the electrode array. Lastly, the quality and availability of parental support and of social and educational services also play a vital role.

The classification of the aetiology of sensorineural hearing loss varies widely between different authors and is compounded by the difficulty in determining the aetiology retrospectively when hearing loss is eventually detected. The extent of investigation of a child newly diagnosed with sensorineural hearing loss also varies widely from centre to centre.

In an effort to further evaluate potential from cochlear implantation, and to anticipate the likely length of habilitation we have classified the aetiology of hearing loss of 95 children implanted at the University of Melbourne Cochlear Implant Clinic according to a preprepared format. The aetiology of hearing loss was then correlated to the best level of speech perception attained according to the grading system previously described by Dowell et al.⁴.

MATERIALS AND METHODS

In this study 88 children who received the Nucleus CI 22M multiple channel cochlear implant at the age of 18 or under and are currently being followed up by this department are reviewed. All children were grouped according to the aetiology of their sensorineural deafness as could be determined from a detailed medical history, examination and investigations performed either at the primary referring institution or at the Melbourne University Cochlear Implant Clinic following referral. Aetiology of hearing loss was known in 60.2% of patients and unknown in 39.8% of patients. Where aetiology is known these groups were then divided into those which were congenital in origin and those groups which were non congenital in origin. Genetic and non

Table 1.

KNOWN

CONGENITAL

Genetic	Syndromic		Non-syndromic	
	Waardenburg's syn	3	Dysplasia	6
	Usher's syn	1	Familial	7
	CHARGE	1	Consanguinity	1
	Klippel Fiel	1		

Non-genetic	CMV	6
	Rubella	6
	FDIU	1
	Coxsackie B	1

NON-CONGENITAL

	Meningitis	15
	Viral illness	2
	CSOM	1
	CMV	1

UNKNOWN

CONGENITAL	33
NON-CONGENITAL	2

genetic subgroups were then considered. Further sub-divisions as illustrated in Fig 1. were then applied as appropriate.

Table shows the breakdown of the population analysed

Speech perception scores for all children were then tabulated based upon the levels of speech perception achieved as previously formulated by Dowell et al, each subject being assigned a numerical score as illustrated below. This is the current method of standardising the speech perception used in the Melbourne University Cochlear Implant Clinic for patient follow-up. It facilitates the comparison of progress of speech perception regardless of the specific test used e.g PLOTT battery, NU-CHIPS closed set test, PBK monosyllabic word test, or the age of the child.

The levels are

1. Detection of speech sounds only.
2. Discrimination of suprasegmental aspects of speech in addition to 1.
3. Discrimination and recognition of vowels in addition to 1 and 2.
4. Discrimination and recognition of consonants in addition to 1 through 3.
5. Minimal open-set speech perception in addition to 1 through 4.
6. Open-set speech perception (>20% phoneme score for PBK words).
7. Good open-set speech perception (>50% phoneme score for PBK words).

RESULTS

The Student-t test was used to analyse the groups. Patients whose aetiology of hearing loss is known were compared to those where the aetiology was unknown. Patients whose hearing loss are congenital in origin were also compared to those whose hearing loss were not congenital in origin. Because of the small numbers in some of the subgroups statistically significant results were only found in comparing the Rubella, meningitis and familial groups. The results are shown in Table 3.

DISCUSSION

Our results suggest that those children whose hearing impairment was congenital in origin

Table 2. Mean and median speech perception scores according to aetiological group.

	N	MEAN	MEDIAN
Rubella	6	5.833	6.000
Meningitis	15	5.600	6.000
Dysplasia	6	5.000	5.500
Familial	7	3.714	3.000
CSOM	1	3.000	3.000
CMV	7	5.571	6.000
Unknown	35	5.257	6.000
Genetic syn.	5	4.200	4.000
Genetic non-syn.	2	7.000	7.000
Viral	3	6.333	6.000
Congenital non-genetic	1	1.000	1.000

Table 3.

	T value	P value	DF
Rubella vs. Familial	2.32	0.045	9
Meningitis vs. Familial	2.07	0.033	10
Acquired vs congenital	2.13	0.04	38

had a poorer performance than those whose hearing impairment was not congenital in origin. Of those children whose hearing loss was congenital in origin those resulting from Rubella appeared to perform best.

There was no significant difference between the group of patients in whom the aetiology of their deafness was unknown and the group of patients in whom the aetiology was known. This is not surprising as the unknown group most likely represents a heterogeneous population of many different aetiologies and so would approximate the average.

As mentioned previously there are many variables involved in assessing patients both before and after cochlear implantation. It is therefore difficult to draw definitive conclusions despite the trends we observed.

The authors feel that the aetiology of hearing loss is of significance and should continue to be an integral part of patient assessment and work-up prior to cochlear implantation

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Author/s:

O'Sullivan, P. G.; Ellul, S. M.; Dowell, B. C.; Clark, Graeme M.

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