MULTICHANNEL COCHLEAR IMPLANTATION IN USHER'S SYNDROME

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Summary

Four patients with Usher's syndrome have been implanted with the multichannel cochlear prosthesis developed by the University of Melbourne and Cochlear Pty. Ltd. All four patients have made good progress with the implant and use the device during most waking hours. One subject, an adult who developed speech and language before developing profound deafness, has learned to use the cochlear implant for auditory alone communication. Her results on speech perception testing compare favourably with other implanted adults. The other subjects were adolescents or young adults when implanted and had congenital profound or total hearing losses. These subjects retained adequate vision for lipreading but had lost hearing before the development of speech and language. Results for these subjects have indicated that the cochlear implant significantly aids lipreading and improves the detection and recognition of environmental sounds. However, they have not, as yet, been able to use the device for communication in the auditory alone condition. The ability to use the device in this condition would become important should vision deteriorate further. These preliminary results suggest that for patients with Usher's syndrome where deafness is progressive and speech and language have developed normally, the multichannel cochlear implant can provide an alternative auditory communication system when vision is no longer functional. When Usher's syndrome has caused a congenital profound deafness, it may be necessary for the cochlear implant to be integrated into the child's normal speech and language development from an early age to obtain optimal results.

INTRODUCTION

The multichannel cochlear prosthesis, developed jointly by the University of Melbourne and Cochlear Pty. Ltd., has now become an established clinical procedure for the treatment of profound or total deafness. The prosthesis has been used primarily in adults where the onset of the hearing loss has occurred after the development of speech and language (1). This surgically implanted device provides enough auditory information for many users to communicate normally in a face to face situation (i.e. using lipreading), and for some adults, communication using the implant alone is possible (2) (e.g., over the telephone). The application of the multichannel implant to deaf children and those who have lost their hearing early in life poses more difficult problems (3, 4). The development of speech and language is dependent on hearing and there appear to be crucial periods in a child's life after which the acquisition of speech becomes more difficult.

The simultaneous occurrence of retinitis pigmentosa and congenital deafness was first described in 1858 by Von Graefe (5) and in 1914 the familial nature of this condition was noted by the British ophthalmologist, Usher from whom the syndrome takes its name (6). Usher's syndrome has been shown to be an inherited autosomal recessive disorder with varying clinical expressivity. Four distinct clinical types (7) have been described with type 1 and 2 comprising approximately 90% of cases. In all clinical variations of the disorder, the visual symptoms follow a similar course. Night blindness is evident in the early years and the visual fields begin to restrict during adolescence. Central vision begins to deteriorate during the second and third decade and this may result in blindness by the fourth decade (8). Type 1 Usher's syndrome is characterized by profound congenital deafness and type 2 by a slowly progressive hearing loss (9). The less common types 3 and 4 are characterized by the additional symptoms of ataxia and mental retardation, respectively. The incidence of Usher's syndrome in the general population is between 1.8 and 3.5 per 100,000 (7) and between 3% and 10% among congenitally deaf children.

In cases of Usher's Syndrome where a cochlear implant may be considered, the visual problems associated with this syndrome provide an
additional complication. As the primary mode of communication for many profoundly hearing-impaired persons is visual (for example, the use of lipreading, sign language or cued speech), the person with Usher's syndrome may need to learn to communicate through an alternative modality when use of the visual system is restricted. These alternatives are tactile or auditory devices and it may be necessary to learn tactile signing codes. The assessment of the multichannel implant as a sensory substitute (to provide speech understanding without the assistance of vision) is necessary in patients with Usher's syndrome as well as the assessment of the device as a sensory aid (for use with lipreading), as the person may lose functional vision by the third or fourth decade of life.

DESCRIPTION OF THE PROSTHESIS
The current model cochlear prosthesis is shown in Fig. 1. All internal components are sealed under the skin during surgery which lasts approximately three hours. These internal components include the electrode array which is inserted into the cochlea, the stimulator that controls output to the electrode array, the receiver coil which receives the power and information from an external transmitting coil via radio frequency induction and the magnet, which holds the external transmitting coil in place.

All other components of the cochlear prosthesis are worn externally. Sounds are received at an ear level microphone, sent to the speech processor via the cord and are processed according to the individuals 'map'. The 'map' is a program, held in memory within the speech processor, which is individualized for each implant user. Each implant user undergoes extensive postoperative testing to measure the required data for their map. The coded signal is sent to the transmitter coil and the receiver/stimulator converts this code into electrical signals which stimulate the hearing nerve fibres.

The speech processor selects the important acoustic features of the speech signal and presents them electrically to the auditory nerve in an appropriate form. The features of speech currently being coded are amplitude, the fundamental frequency and first and second formants. Amplitude is coded as the current level, giving the percept of loudness. The fundamental frequency, which conveys aspects of voicing and pitch, is coded as the pulse rate and gives the auditory percept of pitch. This provides important information regarding the rhythm and intonation of speech. The first formant determines the position of the lower pitch electrode and the second formant determines the position of the higher pitch electrode. These two formants provide important spectral information. The cochlear implant user is able to use this loudness, voice pitch and spectral information to assist in understanding speech (10).

POTENTIAL RECipients
Persons who may be considered for a cochlear implant need to satisfy four main criteria:
1. Profound bilateral sensorineural deafness.
2. Hearing aids provide only minimal benefit. (Persons obtain chance scores on tests using audition alone.)
3. No radiological/medical contra-indications.
4. Well motivated, but with realistic expectations.

Candidates for the cochlear implant procedure can be divided into three groups. The first group includes adult clients who suffer hearing loss through disease or injury after they have acquired speech and language.
They can demonstrate significant benefit from the device in terms of speech perception with and without lip-reading (1,11).

The second group includes the young children whose loss is congenital or acquired pre-linguistically (that is, prior to complete speech and language development). Preliminary research data has shown encouraging results for this group (3,4) and it appears that the cochlear prosthesis will be of considerable benefit for developing auditory skills at a time of optimum cognitive, social, speech and language growth.

The third group includes adults and adolescents who acquire profound hearing loss prior to speech and language development. In these cases there is no auditory memory for speech and language and the optimum time for development of normal speech and language (prior to puberty) has passed. At present 6 adolescents and 4 adults with congenital or pre-linguistic hearing loss have been implanted. Results at present indicate improved detection of environmental sounds and assistance in communication with lipreading. However, understanding speech without lipreading has not been possible with this group as yet and only modest improvements have been achieved in communication skills, even with extensive training.

SUBJECTS AND RESULTS

Four patients with Usher's syndrome have undergone the implant procedure at the University of Melbourne. All four are different in terms of their degree of visual impairment, and their onset and degree of hearing loss. These are summarized in Table 2.

Subject 1, an adult, acquired speech and language before her hearing deteriorated to profound deafness. (Type 2 Usher's syndrome). She was unable to use her poor vision to lipread, so prior to receiving the cochlear prosthesis her communication was extremely restricted. Subjects 1's speech perception results, using audition alone, show that in the year following her operation she could understand approximately 30% of words in sentences. This score is close to the average for adult patients with acquired deafness using the cochlear prosthesis. (see Table 3).

The remaining three subjects were adolescents with congenital deafness but with sufficient vision to be able to lipread. These three use the device as a sensory aid in conjunction with lip reading. Subject 2 has reported significant benefits from the implant and speech processor, with the ability to detect and recognize some environmental sounds, recognition of vowels from closed sets and discrimination between different speakers. No data is yet available to demonstrate subject 2's audition alone skills, however the implant has been found to significantly aid his lipreading.

Subjects 3 and 4 when tested with sentence material obtained results that were significantly better when the device was used with lipreading compared with lipreading alone (Table 4), demonstrating the effectiveness of the implant as an aid to communication. However, at the time of writing, subjects 3 and 4 were not able to communicate using audition alone.

CONCLUSION

The primary aim for hearing impaired persons fitted with the multichannel cochlear implant and speech processor is for successful communication combined with lipreading. Approximately one third of adults with acquired deafness can use the device to communicate with out lipreading. Where visual skills are compromised the aim is for satisfactory communication using the implant alone. Our preliminary results with Usher's syndrome patients indicate significant improvements for detection of environmental and speech sounds as well as communication with lipreading, but if the implant is required as a sensory substitute (that is, vision is too poor for lipreading) then the training to use the
implant should coincide with early speech, language and cognitive development. The three implant users with congenital hearing loss (described above) had developed speech and language through vision. However, their vision had significantly deteriorated in the second decade of life and they received the implant after puberty. Results at a perceptual level have been very encouraging considering the lack of auditory input before implantation and evaluations using lipreading indicate the prosthesis does aid communication in the auditory-visual modality. However, the data collected at this stage does not indicate the ability to use audition alone for communication.

This has implications for professionals and habilitationists working with children with Usher's syndrome, particularly with regard to early intervention programs. The multi-channel cochlear prosthesis should be integrated into the young child's speech and language development and normal communication at the optimum developmental period, before vision has deteriorated significantly. Training to use the auditory input at this early age would assist the child in reaching his/her potential in audition alone communication with the multi-channel cochlear prosthesis.

REFERENCES
Fig. 1  Cochlear implant package and electrode array. These components of the implant system are surgically placed under the skin behind the ear.

Fig. 2  External components of the cochlear implant system.
FIGURE LEGEND

Fig. 1: Cochlear implant package and electrode array. These components of the implant system are surgically placed under the skin behind the ear.

Fig. 2: External components of the cochlear implant system.
### TABLE 1

**Coding Strategy for the Multi-Channel Cochlear Prosthesis (POF1F2)**

<table>
<thead>
<tr>
<th>Acoustic Parameter</th>
<th>Electrical Parameter</th>
<th>Auditory Percept</th>
</tr>
</thead>
<tbody>
<tr>
<td>Amplitude</td>
<td>Current Level</td>
<td>Loudness</td>
</tr>
<tr>
<td>Fundamental</td>
<td>Pulse Rate</td>
<td>Rate Pitch</td>
</tr>
<tr>
<td>1st Formant</td>
<td>1st Electrode</td>
<td>Place/Pitch (Low)</td>
</tr>
<tr>
<td>2nd Formant (High)</td>
<td>2nd Electrode</td>
<td>Place/Pitch</td>
</tr>
</tbody>
</table>

### TABLE 2

**Subjects**

<table>
<thead>
<tr>
<th>Subject</th>
<th>Audition</th>
<th>Vision</th>
<th>Age at Implant</th>
<th>Results</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Gradual loss.</td>
<td>Tunnel vision.</td>
<td>35:0</td>
<td>Audition used as sole communication channel.</td>
</tr>
<tr>
<td></td>
<td>Profound by age 27.</td>
<td>No lipreading.</td>
<td></td>
<td></td>
</tr>
<tr>
<td>2</td>
<td>Congenital profound loss.</td>
<td>Poor peripheral vision.</td>
<td>23:0</td>
<td>Discrimination of environmental sounds. Used with lipreading.</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Night blind.</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>Acuity 6/18.</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>Functional lipreading.</td>
<td></td>
<td></td>
</tr>
<tr>
<td>4</td>
<td>Congenital total loss.</td>
<td>Tunnel vision.</td>
<td>14:9</td>
<td>Improved detection and discrimination for speech and environment sounds. Used with lipreading.</td>
</tr>
<tr>
<td></td>
<td>&lt; 10 degrees.</td>
<td>Night blind.</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>(R) 6/18 (L) 6/9</td>
<td>Poor lipreading.</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
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