

SHORT REPORT

Cochlear implantation for progressive hearing loss

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Arch Dis Child 2003;**88**:708–711

The concept of neural plasticity and the early natural abilities of hearing children to acquire speech and language without instruction have led many authorities to advocate cochlear implantation before the age of 5 years in congenital deafness. Older children therefore become lower priority for scarce public funds because they are perceived to have passed the “window of opportunity” to learn speech, even if hearing is restored, and continue to rely on sign language.

This paper shows that a subgroup of congenitally deaf children exists, who, having made good progress with conventional hearing aids, suffer a sudden or progressive hearing deterioration which arrests the speech development.

Sixty children have been implanted in the Cambridge Programme, half for meningitis or other acquired losses and half for congenital prelingual deafness. Six of this latter group were congenital but progressive; their progress, deterioration, and improvement after implantation are summarised.

Cochlear implants allow children who are congenitally deafened to develop auditory-aural communication rather than relying on sign language. This allows them to attend mainstream schools, often with some support. Not all children are suitable for implantation; for example, because their hearing is too good and hearing aids would be of sufficient help, or because the operation would be technically unfeasible. Those children who are appropriate candidates must be identified as early as possible as it has been shown that they will develop better understanding of speech and have better auditory-aural communication skills than those identified and implanted later.^{1,2} Due to the loss of neural plasticity in later development, children born with profound hearing loss do not develop useful hearing if implanted much over the age of 5. In the haste to implant these children we feel that other candidates, who are born with better hearing but suffer deterioration, may be neglected.

Children who are identified with sensorineural hearing loss which requires hearing aids are often referred to their local cochlear implant centre if they do not show good development of auditory-aural skills. Professionals concerned with their care are keen that they are assessed early for the reasons outlined above. Some children who complete their assessment and who are found unsuitable for implantation on audiological grounds may subsequently suffer deterioration in their hearing.

In children whose language and hearing skills deteriorate with good hearing aids it should be considered whether worsening hearing is the cause. These children must be identified and considered for reassessment. We wish to highlight some of those cases identified by the East Anglian Cochlear Implant Programme who show good development in spite of implantation occurring when older. Children who become profoundly deaf later have had cortical stimulation by sound and will have cortical development, so that the loss of neural

plasticity is no longer of such importance. We put the case for these children being good candidates for implantation, and show outcomes as satisfactory as other reported groups.³

METHODS

A subgroup of the children receiving cochlear implants in Cambridge was selected. Their parameters differ from the majority of implanted children in that they all suffered a progressive loss and were implanted late. The implants took place between 1996 and 1999. All received the Nucleus 22 device. All had full electrode insertions and all were rehabilitated with body worn speech processors.

RESULTS

Each child's results are illustrated graphically (fig 1); fig 2 summarises results for all children. The children had an average Bench-Kowal-Bamford (BKB)⁴ sentence score of 76.2% in the five who were measured (see fig 3). Those whose outcomes were measured using the Manchester Word List, had an average of 98% (see fig 4).

The most difficult speech hearing test uses single words, with no contextual or lip reading clues. This test was beyond the ability of the children in this series. Sentence tests (such as BKB sentence scores) are easier, as there are contextual clues, but these demand a significant pre-learned vocabulary. The Manchester Word Lists are easier, requiring a smaller vocabulary, such as that of a young or hearing impaired child.

Case 1 (FD) (deterioration 12 years after gentamicin)

This female patient was born prematurely at 26 weeks gestation and given gentamicin during the neonatal period. She was first given hearing aids at 2 years, and was able to speak 50 clear words at 2½ without signing. At 4 years of age, four frequency average audiometry was 72 dBA unaided. She was able to wear bilateral Phonak PPCL2 hearing aids. Her hearing loss was monitored, and was seen to deteriorate when checked at 12, 16, and 18 years of age. By 18 years old the four frequency average had deteriorated to 104 dBA unaided. Live voice BKB sentence scores with audition alone were 12%. Cochlear implantation was performed at 18 years of age. After implantation BKBs were 63% (97% with lip reading). The patient was able to undertake a university degree and obtain a Diploma of Education. She now teaches primary school children.

Case 2 (EG) (deterioration 13 years after cytomegalovirus)

This female patient's hearing loss was due to cytomegalovirus. She was first provided with hearing aids at age 3. At age 12 she had aided hearing of 40 dBA average over four frequencies, and able to wear Phonak PPCL2. She was communicating in auditory oral fashion, with clear speech understood by everyone. Her hearing suddenly deteriorated at age 13. Audiograms showed average aided hearing of 85 dBA over four frequencies. On the Manchester picture test she achieved 10% only with audition alone. Cochlear implantation was performed at 15 years of age. After implantation she achieved aided four

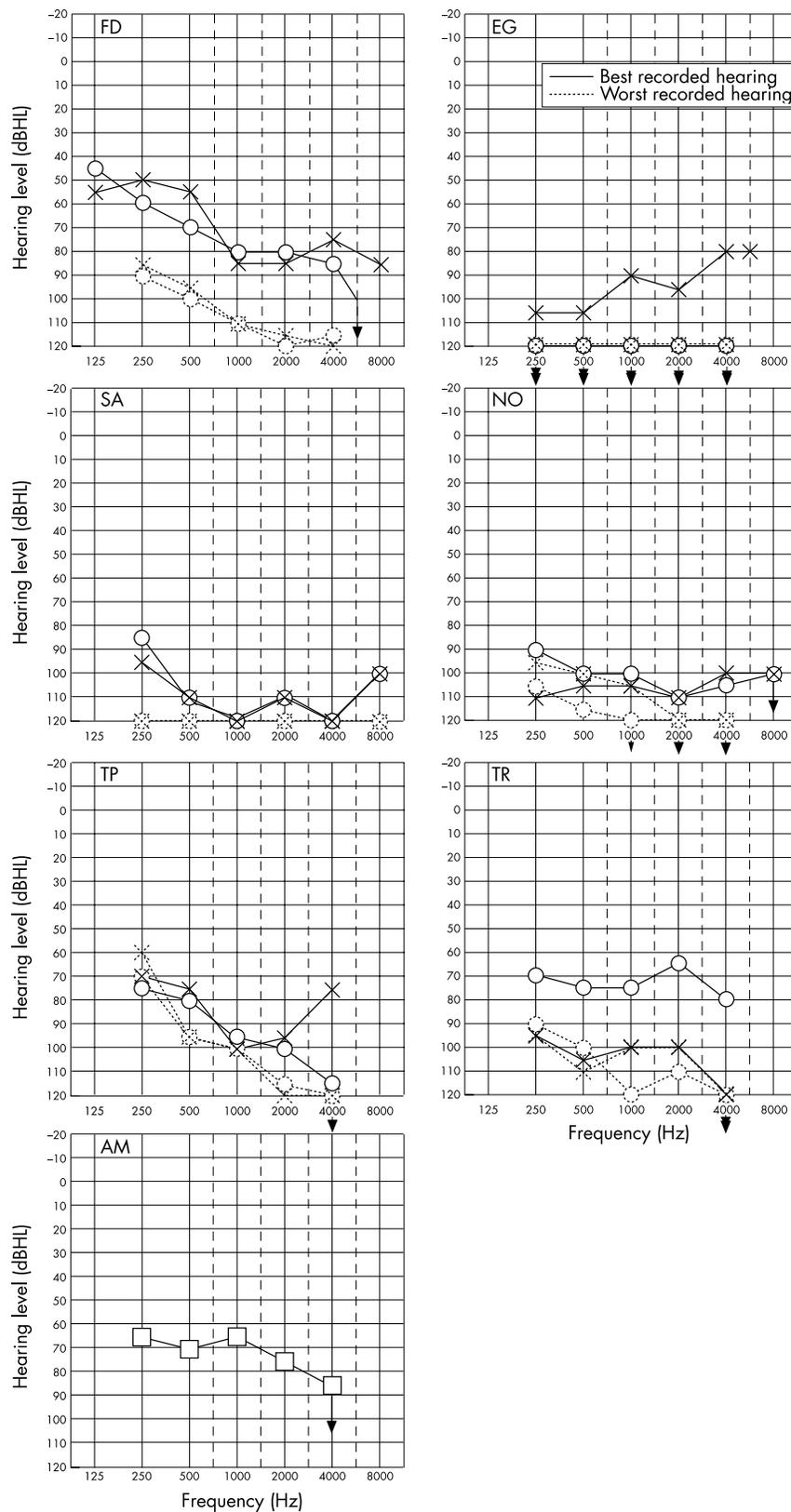


Figure 1 Unaided pure tone audiograms of the patients. Pre-deterioration audiograms not available for AM as too young.

frequency audio at 40 dBA, and a Manchester Word List (MWL)⁵ score of 100%.

Case 3 (SA) (deterioration 11 years after maternal influenza)

The mother of this female patient suffered influenza during the prenatal period. Hearing aids were first provided at age 2.

Unaided audiometry at age 8 showed an average of 109 dBA. At that time she was using bilateral post-aural hearing aids successfully. She communicated auditory-orally, although speech was only understandable by sympathetic adults. She began to learn sign language at age 11, and suffered sudden hearing loss at that time. At age 15 unaided audiometry showed four frequency average of 120 dBA, and BKBs were

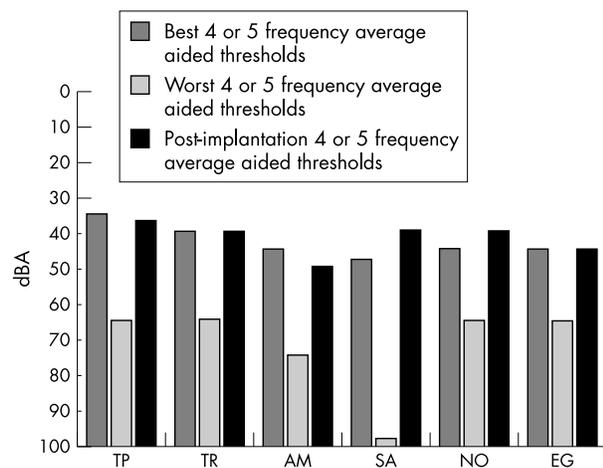


Figure 2 Bar chart showing improvements for aided threshold average when at worst, and implant aided threshold average.

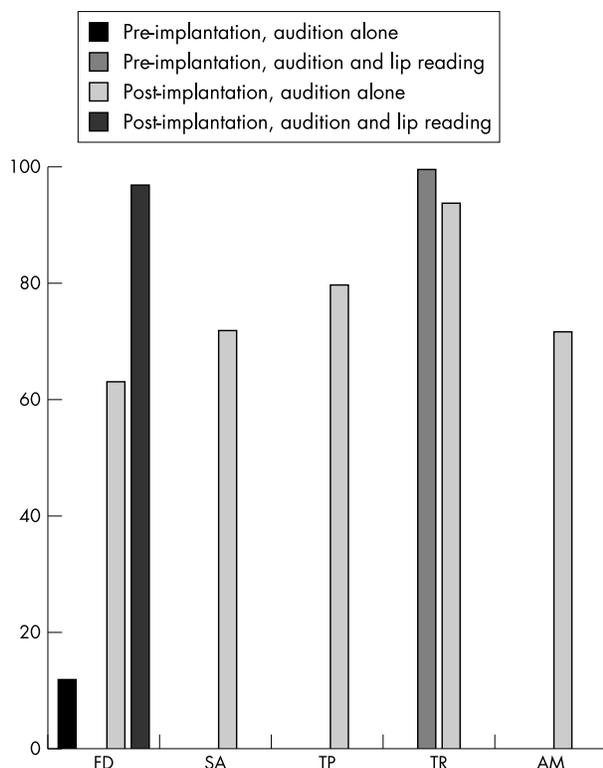


Figure 3 Bar chart showing results of BKBs pre- and post-implantation.

0%. Cochlear implantation was provided at age 15. Following implantation she had aided threshold average of 30dBA, and live voice BKBs of 72%. She was able to cope as a college student and to use the telephone to speak to family.

Case 4 (NO) (deterioration 10 years after congenital idiopathic deafness)

This female was born at term. She was diagnosed as having congenital idiopathic hearing loss, and first provided with hearing aids at 2 years old. At age 6 she had unaided average thresholds of 103 dBA. She used Phonak PPCL4+ aids, and her speech understood by a sympathetic listener. She gradually progressed to profound hearing loss by age 10, and began to learn sign language at age 14, although she remained reluctant to use it. By age 14 her aided threshold average was

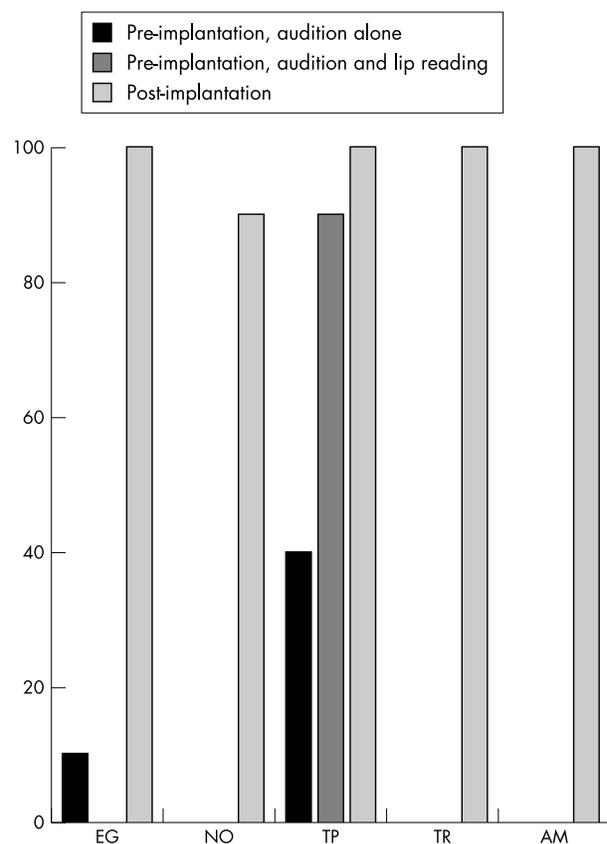


Figure 4 Bar chart showing results of MWLs pre- and post-implantation.

65 dBA with no responses above 1500 Hz. Cochlear implantation was carried out at age 14. After implantation she achieved aided audiometry of 37 dBA average with MWL of 90%, and was able to return to auditory oral communication.

Case 5 (TP) (deterioration five years after gentamicin)

This male patient was born at 32 weeks gestation. The cause of hearing loss was thought to be gentamicin used during neonatal period. He was first given hearing aids at 1 year. His average aided hearing at age 3 was 35 dBA. He was able to usefully wear Oticon 380P hearing aids with good speech production and without need for signing. He gradually progressed to profound hearing loss by the age of 5. By the age of 6 he had average thresholds of 107 dBA unaided, and 60 dBA aided. The Manchester picture test results were 90% auditory and visual, but only 40% on audition alone. Implantation was carried out at 6 years of age, after which he achieved implant aided average thresholds of 38 dBA over five frequencies, live voice BKBs of 80%, and MWL of 100%.

Case 6 (TR) (deterioration nine years after congenital idiopathic deafness)

This male was born at term. The aetiology of his congenital hearing loss was thought to be idiopathic. He first received hearing aids at 3 years old. At age 8 he had four frequency average thresholds of 40 dBA, and wore Phonak PPCL4 aids, achieving clear speech production, and not using sign language. He suffered gradual progression to profound hearing loss at age 9, by which time he had average threshold of 105 dBA unaided and 73 dBA aided. On BKB testing he achieved 100% when tested by lip reading with audition, but 0% on audition alone. He was implanted at 10 years of age, and after implantation achieved a four frequency average of 40 dBA, live voice BKBs of 94%, and MWL of 100%. He enjoys

mainstream schooling with support and relies only on auditory oral communication now.

Case 7 (AM) (deterioration with septicaemia, aged 2 years)

This boy had congenital hearing loss associated with Ehlers Danlos syndrome. He was first given hearing aids at age 1. At age 14 months he had aided two frequency average thresholds of 40 dBA, and was able to use 20 words with Phonak Pico PPCL aids. He suffered septicaemia at age 2, causing sudden deterioration in his hearing. By age 3 he had four frequency average thresholds of 119 dBA unaided, and 82 dBA aided. At that time he was able to differentiate "quack quack" from "moo" with the aid of lip reading. He underwent cochlear implantation at age 3½. Afterwards he had implant aided thresholds of 42 dBA five frequency average, with live voice BKBs of 72% and MWL of 100%.

DISCUSSION

Over the years the ideal age for implantation suggested by authors has been getting younger.⁶⁻⁹ Children with a moderate to severe congenital sensorineural hearing loss which is progressive do not necessarily fit into a general protocol. They are not suitable candidates for implantation at an early age, but may become so later than the ideal age for children born with profound hearing loss. We are concerned that these children may have their implantation delayed in preference to those children born with profound sensorineural hearing loss.

Children who have developed predominantly auditory and vocal style of early communicative behaviour are predicted to have relatively high levels of speech and language skills after implantation¹⁰; therefore one would expect children who initially had hearing amenable to a conventional hearing aid should have good results from cochlear implantation. This is borne out by our results. These children had an average BKB of 76.2% in the five who were measured. The five whose outcomes were measured using the MWL had an average of 98%.¹¹ This compares well with an article using BKBs in post-operative assessment.¹² The lower score in BKB testing is probably because children require a larger vocabulary to perform the more difficult BKBs.

Duration of deafness has been found in one adult study¹⁰ to be an important negative predictor of results of implantation. Children such as these should therefore be implanted as soon as deterioration in hearing and speech becomes evident.

These children make good candidates for cochlear implantation, especially if they have had good auditory oral communication and good conventional hearing aids before progression to profound sensorineural hearing loss. We hope that we have shown that children such as these should be carefully considered for referral to cochlear implant centres for assessment and not overlooked because of their older age.

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Previously presented as an abstract at CI 2000 6th International Cochlear Implant Conference, Miami, Florida, USA, 3-5 February 2000

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Accepted 15 December 2002

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