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Speech production in deaf implanted children with additional disabilities and comparison with age-equivalent implanted children without such disorders

Thomas P. Nikolopoulos^{c,*}, Sue M. Archbold^b, Casper C. Wever^b, Hazel Lloyd^a

^a ENT and Audiology Directorate, Nottingham University Hospital, Nottingham NG7 2UH, United Kingdom ^b The Ear Foundation, 83 Sherwin Road, Lenton, Nottingham NG7 2FB, United Kingdom ^c Athens University, 2nd ENT Department, Attiko Hospital, Athens 124-62, Greece

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Cochlear implant; Deaf; Speech; Intelligibility; Children; Multiple; Additional; Disorder; Handicap; Disability; Language; Result; Outcome; Communication	Objective: To assess the long-term speech intelligibility in implanted children with additional disorders and compare them with age-equivalent implanted children without such disorders. Patients: 175 profoundly deaf children 5 years following cochlear implantation; 67 children with additional difficulties and 108 children without such difficulties. All children were implanted under 5-year-old. Main outcome measure: Speech intelligibility rating (SIR) scale that can be readily applied to young deaf children irrespective of their performance and is reliable between observers. Results: Five years following implantation, 47 (70%) children with additional difficulties developed connected intelligible speech versus 104 (96%) in the control group. However, the quality of speech was quite different between the two groups, as only 11 (16%) children with additional difficulties achieved the two higher categories (intelligible to all or to people with little experience), whereas 66 (61%) children in the control group did ($P < 0.000001$). The total number of additional disorders had the strongest correlation with the outcome. Language and communication disorders were the most important contributing factor, followed by physical, cognitive, and autistic spectrum disorders.
	nected intelligible speech 5 years following implantation; however, a significant

* Corresponding author at: 125 Anakous Street, New Philadelphia, Athens 143-42, Greece. Tel.: +30 210 2519641; fax: +30 210 6994904. *E-mail address*: Thomas.Nikolopoulos@Nottingham.ac.uk (T.P. Nikolopoulos).

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proportion do not develop any speech at all. Thus a third of this group did not realise one of the most important objectives for parents of implantation. Benefit from implantation should not be restricted to speech production alone in this specific population.

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1. Introduction

Many cochlear implant centres have been conservative about implanting children with additional needs, although they represent a significant proportion of deaf children. However, as implant teams have become more experienced and confident, children with complex needs have increasingly been considered. Several studies have reported benefit from implantation in these children [1-5], although many naturally only have small numbers.

Intelligible speech is often the primary and most important objective for parents of profoundly deaf children to seek cochlear implantation. Several studies have demonstrated that intelligible speech is a realistic expectation for many implanted children but not for all [6,7]. However, the situation becomes more complex in profoundly deaf children with additional disorders and the respective literature is extremely limited [8].

The aim of this study was to assess the traditional outcome measure of long-term speech intelligibility in a significant number of implanted children with additional disorders and compare it with the respective intelligibility of an age-equivalent group of implanted children without additional disorders.

2. Materials and methods

The present study assessed the speech intelligibility of 67 profoundly deaf children with additional difficulties and compared them with 108 implanted children without such difficulties, 5 years following cochlear implantation (from 1997 until the writing of the present study). All children were implanted under 5-year-old. Prior to implantation, all subjects had a bilateral profound sensorineural hearing loss (greater than 95 dB) across the speech frequency range. Care was provided free of charge on the UK's National Health Service, and hence ability to pay did not determine access to the service; as a consequence, the children in the study were drawn from the full range of social, educational and communication environments and were drawn from throughout the UK. Children with additional disorders were considered for implantation at an individual basis. All children received the Nucleus multichannel cochlear implant. They were programmed with the speech processing strategy recommended at the time and upgraded with new encoding strategies as they became available.

The programme's database (BCS system) contains the following categories of additional difficulty:

- Autistic spectrum disorders.
- Behaviour difficulties, which are over and above what one would expect as a result of deafness.
- Cognitive difficulties, which are likely to prevent normal progress.
- Language and communication difficulties, where children have significantly delayed or deviant language, or aberrant communication patterns.
- Oro-facial.
- Vocal tract anomalies.
- Physical difficulties.
- Visual impairment.
- No additional difficulty suspected.

These difficulties are in addition to those arising purely from deafness. They are only entered on the database when they have been identified, or when they are suspected and suspicions have been fully discussed with the child's or adult's parents/ carers, or adult/child themselves. It would be unethical to easily put a label of an additional disorder to a deaf child, even if this hesitation might have underestimated the additional disorders in these children.

From the 67 children with additional difficulties, 39 (58.2%) had one additional difficulty, 15 (22.4%) two additional difficulties, 6 (9%) three, 5 (7.5%) four, and 2 (3%) five additional difficulties. In detail (Table 1) 44 children (66%) had language and communication difficulties, 18 (27%) cognitive difficulties, 21 (31%) visual impairment, 8 (12%) physical difficulties, 13 (19%) behavioural difficulties, 7 (10%) vocal tract anomalies, 4 (6%) autistic spectrum disorders, and 2 (3%) oral/facial disorders.

In the additional difficulties group, 31 children (46.2%) had congenital deafness of unknown origin, 19 (28.3%) were deafened by meningitis, 6 (9%) CMV, 6 (9%) syndromal deafness (of which 3 had Usher's syndrome), and 5 (7.5%) children were deafened from other causes. From the 6 children with CMV as etiology of deafness, four had more than one additional disorders (2 had vocal tract anomalies).

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