Additional Difficulties Associated with Aetiologies of Deafness: Outcomes from a Parent Questionnaire of 540 Children Using Cochlear Implants - Short Review

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Introduction
Since 1996, the Nottingham Auditory Implant Programme (NAIP) has been providing cochlear implants to children with a wide range of additional difficulties, some of which are known prior to implantation, whilst others are diagnosed months or years afterwards. Therefore we were aware that our database required updating, so that the child's care pathway could be tailored to their individual needs.

In the early 2000s, some members of the team were being asked whether or not electrical stimulation from using a cochlear implant could trigger or exacerbate epileptic seizures. NAIP could not review which children had a diagnosis of epilepsy, as many of these children did not have seizures at the pre-implant stage, so it was difficult to audit implant use and occurrence of seizures in these patients.

Audit
In 2010, NAIP sent a short written questionnaire to the parents of 590 children who had received cochlear implants in Nottingham, asking them to inform us of any diagnosed additional difficulties so that we could update our records. We did not ask parents about speech and language or behaviour, as we thought that this could lead to a high proportion of subjective positives and/or few cases of diagnosed specific speech and language or behaviour difficulties [1,2].

The questionnaire asked about the following diagnosed additional difficulties:

- Visual impairment (not corrected by glasses); cognitive delay; epilepsy; autism; movement difficulties; any other health issues.

The results of 540 returned questionnaires allowed NAIP to analyse the information in relation to the proportion of severe/profoundly deaf children with additional difficulties, to the child's aetiology of deafness and specifically to those which seemed to be associated with epilepsy.

The proportion of our paediatric population having at least one diagnosed additional difficulty, likely to affect listening and spoken language outcomes following cochlear implantation was found to be 47% of the total questionnaires returned. This is higher than Fortnum and Davis' large epidemiological survey of children in 1997 and higher than a recent survey by the National Deaf Children's Society, UK.

The distribution of additional difficulties in cohorts of children with the same aetiology was examined and trends were identified. Those children who were deafened by congenital Cytomegalovirus (cCMV) infection, acquired Meningitis (or other cerebral infections) or those whose deafness appeared to be suggestive of Auditory Neuropathy Spectrum Disorder (ANSD) appeared to be at greatest risk of suffering epileptic seizures either prior to or after cochlear implantation. Two cases, deafened after acute menigitis, suffered multiple seizures prior to having a cochlear implant and their parents made a decision not to continue with implant use as they could not rule out that their seizures were exacerbated by wearing the processor. However there was no evidence from other children with a diagnosis of epilepsy that their fits were triggered or made worse by using the processor [3-5].

Children whose deafness was associated with cCMV, Meningitis infection and ANSD also had the highest number of additional difficulties, including a higher proportion of autism than the general population, whilst those children whose deafness was caused by Connexin 26 had the fewest difficulties.

Over one third of NAIP's paediatric population at the time of the audit had no known cause of deafness, yet amongst these children there were 11 with autism, 4 with epilepsy and 15 with visual problems.

Conclusion
This audit provided new information about additional difficulties in children with severe/profound deafness who use cochlear implants. It also challenged the common belief that between 30- 40% of deaf children have an additional difficulty.

Knowing that certain aetiologies of deafness can cause additional difficulties allows parents to be counselled with evidence-based research, which can help them to have realistic expectations following cochlear implantation.

This audit did not investigate how many children have specific speech and language difficulties or behaviour disorders and it is hoped that NAIP will be able to address this in a future study.

It should also then be possible to look at speech, language and listening outcomes in cohorts of children with the same aetiology of deafness to counsel parents and advise education and health services to provide specialised rehabilitation and learning programmes to support these childrens' needs.

References