

Parental Perspectives on Management in Auditory Neuropathy Spectrum Disorder

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From the Study

***‘Experiences of Parents Whose Babies
are Identified with Newborn Auditory Dys-
Synchrony through NHS Newborn
Hearing Screening Programme’***

Funded by the NDCS

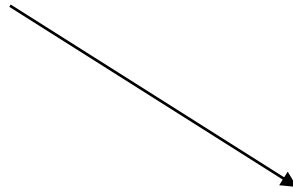
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ANSD in young infants

- **condition is characterised by:**
 - ABR absent or severely abnormal at high levels
 - OAEs present and/or CM present
- **normal cochlear function at the level of OHCs but dys-synchronous auditory brainstem responses**

At birth:
Normal OAEs
Absent ABR



Prognosis???

Normal
auditory
function

Total lack
of sound
awareness



Challenges in ANSD

- ABR cannot be used to estimate hearing thresholds
- Even if we obtain the hearing thresholds, they will be a poor predictor of perceptual abilities
- ANSD-specific changes: daily fluctuations, deterioration, improvement and even complete (?) recovery
- Management???:
 - Heterogeneity of the condition
 - Evidence base weak

Epistemology of the Study

- The study sought to place **parents in the centre of knowledge generation** about their own experience and set out to enable parents to construct to professionals what it is that should be known
- **Constructivist epistemology** was adopted because:
 - there was **no prior research** on parents with ASND on which to build;
 - it was assumed that previous studies of the experiences of parents of early identified deaf children (e.g. Young 2010) were fundamentally **not relevant** to particular circumstances of parents of infants with ANSD;
 - an objective of the research overall was to use its findings **to inform new professional guidelines for practice**, therefore we sought to invest authority in privileging parents' experience and understanding within that end process.

Methodological approach

- qualitative study, based on narrative storytelling
- a questionnaire to collect sociodemographic information and more detailed information about the child's medical and audiological profile

Sampling

- a purposive convenience-based sampling
- a sample size of **21 interviews** of parents of children identified with ANSD
- **no attempt at statistical analysis**
- instead, it is the **in-depth and qualitative comparison of individual case-studies which is valid here**

Analysis

- The **unit of analysis** in this study is the interview rather than the individual respondent
- Data were audio recorded and transcribed in full
- **Codes** were generated independently by members of the research team, then compared for consistency or deviance
- Accounts were then analysed using **thematic content analysis** using NVivo

Results

- **Visual communication**
- **Hearing aid fitting**
- **Cochlear implantation**

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- } Today's
talk

Conflicting information

All parents talked about getting ‘conflicting information’:

- *‘and she just said you know ..there’s obviously sign language and cued speech and.. and she did say there’s a whole school of thought around.. hearing aiding or not.....implanting or not ..and there’s a whole other school of thought around ...sign language or not....cued speech or not...both or not (laughs)....so you just sit there thinking...what do I do for the best.. ... I think we know what the options are so... you know we mightsee her with a hearing aid in ..we might see her with a cochlear implant ..she might get completely better and she might not.. it might get worse’ (F1)*

Hearing aid fitting

- from clinical experience with adults, the established approach in management and intervention when it came to hearing aid candidacy was: hearing aids are not beneficial (Starr et al, 1996)
- risk of damaging intact cochlear structures such as outer hair cells was raised (Berlin, 1999)
- this approach was challenged by Rance et al (2002) who showed that approximately half of the children diagnosed with ANSD benefited from hearing aid fitting evidenced by considerable improvement in their speech perception (Rance et al, 2002)
- the current guidelines recommend trial hearing aid fitting in these children (Northern, 2008)
- however the challenges remain

Hearing aid fitting

The **'intuitive' advice** from the extended family:

- *'my sister seems to think you know...put it's like wearing glasses erm... you can't see so you put your glasses on you can see all day every day and that's it, you're ok now' (F5)*
- *'the grandparents are a little bit more well....look he can hear us, it's gonna be ok he's gonna be alright, isn't he? He's going to be able to hear, he won't need the hearing aids ...' (F4).*

Hearing aid fitting

Managing parental expectations:

- *‘I burst out crying at the doctors ‘cos I saw all these little toddlers having a little chit chats....so I was kind of optimistic then about getting the hearing aids.... no never and this...and I was a little bit angry if I’m honest, so we got the call saying that the ear moulds had come in...so we went to go get them and I just smelled a big rat (laughs) then I knew something was... wrong because it was somebody else it was a different...’ (F13)*

Hearing aid fitting

Hesitation about **reinforcing** hearing aids in very young children:

- *‘he doesn’t like what he hears and it’s not helpful why should I or should I trust him...that he doesn’t want to wear it because he doesn’t it’s not good for him...or should I push...mmm it’s tricky’ (F9)*

Hearing aid fitting

Will hearing aid **damage** the perfectly functional **outer hair cells** whose integrity can be evidenced by normal OAEs and/or CM?

A minority of parents (F1, F13, F16) pick up on that and express their worry about the damaging effect of the amplification:

- *I understand why he can't have...hearing aids and I think....I kind of came to that conclusion myself though from on the internet when I realised kind of properly that the inner ear's functioning fine then obviously if you put a hearing aid on then it would damage...but nobody really explained that on the first...kind of...meeting. (F13)*

Hearing aid fitting

Some parents express not having confidence in the **choice of hearing aid fitting strategy**:

- *I we did have a sort of discussion about how they were going to set her hearing aids 'cos some people believe to set it by the audiogram...and some people believe to set it conservatively...and then if you get no response increase it because at the end of the day you don't know what noise that child's getting. That were a concern and I did feel quite anxious at that point looking back 'cos I just thought god they really don't know you know... (F14)*

Hearing aid fitting

Out of 16 cases from our sample of 21 where hearing aids were trialled, 5 families found amplification very beneficial and 3 somewhat beneficial:

- *aids are good erm... (F5)*
- *erm and yet he responds so well (F16)*
- *I do think it does make a difference when he's got them in he seems to be more attentive when he's got them in... (F19)*
- *oh yeah massive...massive....the change in him is...yeah...his understanding of words is just brilliant...really brilliant... (F4)*
- *you can talk behind him and everything he'll understand everything he doesn't need to see your mouth but when you do that he knows it's a game he's trying to work out so.. (F11)*

Hearing aid fitting

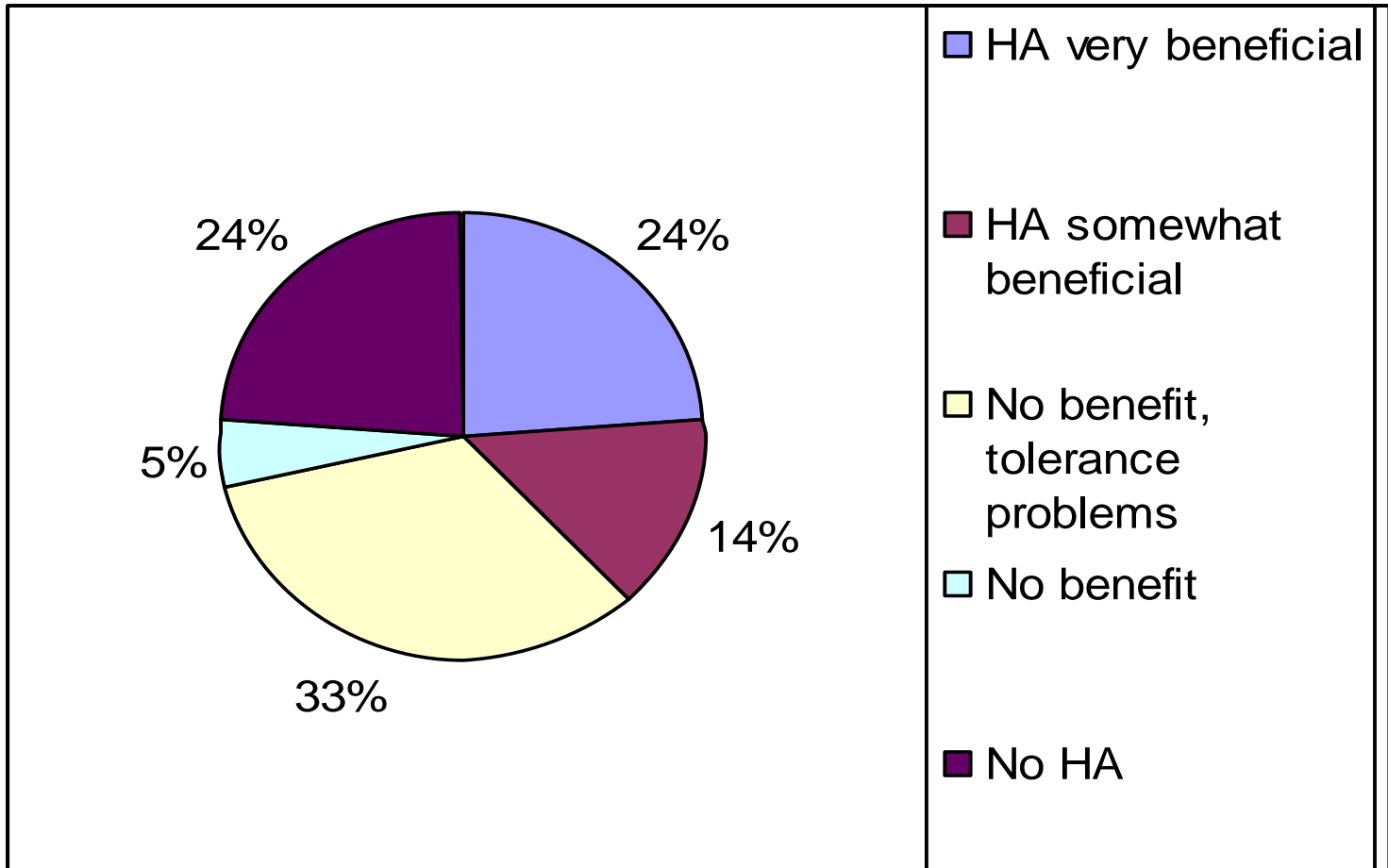
A few parents (N=8/21) said they did not find hearing aids beneficial, either because of apparent tolerance issues:

- *‘dreadful...he hates them...’ (F2), ‘*
- *‘she wasn’t tolerating them’ (F12),*
- *‘no...she wouldn’t keep them in anyway (laughs)....’ (F15)*
- *‘pulls them out he’s a pain in the backside (laughs) with his hearing aids he’d much rather eat them than wear them..’ (F17)*

or simply because they did not appear to work for the particular child

- *‘they clearly don’t make any difference...’ (F3)*

Hearing aid fitting



Hearing aid fitting

Even if hearing aids did not work for their child, many families would still recommended trying them to other families with children identified with ANSD:

- *‘it’s worth trying yeah it’s got to be worth trying but...obviously with (child’s name) it’s not’ (F12)*

Cochlear implantation

- Controversies surround cochlear implantation in ANSD
- Based on the clinical experiences with adults with ANSD who predominantly suffer from a neurodegenerative disease, hesitation by professionals about whether cochlear implants even worked for ANSD.
- Many parents pick up on this:
 - *no I mean well someone said that wouldn't work for auditory neuropathies is that right would you know... yeah somebody told us it's not an option for us to be honest.. (F15)*

Cochlear implantation

- Electrical stimulation can result in synchronised neural impulses sufficient for auditory stimulation in **some** patients with ANSD (Shallop et al 2001, Madden et al 2002, Sininger & Trautwein 2002, Mason et al 2003, Peterson et al 2003, Rance & Baker 2008, Teagle et al 2010).
- CIs effective in **some** children with ANSD (Teagle et al 2010)
- Performance poorer than in typical 'SNHL' (Rance & Baker 2008)
- 20-30% of kids able to perform open-set speech perception testing scored <30% correct (Myamoto et al 1999, Rance et al 1999, Gibson & Sanli 2007, Teagle et al 2010)
- Success of CI likely linked to the site of lesion (McMahon et al., 2008):
in **pre-synaptic lesions** CI likely better outcomes
- Outcomes affected by CNS pathology

Cochlear implantation

Balancing the timing of CI:

- *‘Our implant centre told us they had children before... who recovered actually almost completely...so they didn’t implant and there was one little girl from who was implanted there at the age of 1... but then they found out that actually the other ear is good and she is able to talk without her implant on just with the left ear so they were very very cautious about proceeding with the implantation’ (F8)*
- *‘Actually the hardest time was when we felt he was getting 2 years of age and we felt there was still no development of speech whatsoever.....it was quite stressful because I felt I need to be now the pushy parent...but I was quite sure I know what I’m doing...and so eventually we they did agree to implant (child’s name) he was 22 months then...’ (F8)*

Cochlear implantation

Some parents struggle considering CI because it has been stressed to them throughout the whole process that ANSD children have an **intact cochlea**:

- *‘although at one stage they were offering (child’s name) a cochlear implant which...I was horrified at first because it was like...but his cochlear works ok and you’re gonna have to destroy that cochlear....and to me that was like illogical but then when I thought about it and I thought well if it’s gonna help get the message through and if he’s never gonna hear because of the problem but a cochlear implant is gonna help that then obviously....it’s got to be done’*
(F12)

Cochlear implantation

Some parents find it difficult to consider CI when their **child's relatively good sound detection**:

- ... and we're physically you know and mentally ready to kind of start off down the.... you know monthly journeys down to (place name).....erm....'cos it's not a ...I don't feel like it's gonna to be an easy process and it's not going to be an easy decision.. to make 'cos we know...we know (child's name)'s got hearing in both ears and the thought ofturning that... you know...turning that off....is really hard... (F3)

Cochlear implantation

Outdated and inappropriate candidacy criteria taking account the PTA instead of functional hearing tests such as speech tests were applied:

- *when he lost his hearing to be honest it was the best thing that could have happened to him I think....because he's now he's got...far better hearing with his implant than he ever had...without and there was a point where...we went for...a years worth of...erm...assessments at (area name for cochlear implant centre) and then in the end they like went actually his hearing isn't bad enough and I felt really...like we were just stuck...you know he wasn't bad enough to have a cochlear implant and he wasn't good enough to.... hear without it (F3)*

Cochlear implantation

In our sample, **5 out of 21** had received a cochlear implant for their child and all expressed that they felt it had been a good decision.

Cochlear implantation

- *it's absolutely made such a gigantic difference.. (F3)*
- *...it looks like I think it's improving it why feels like it's going the right way...(F6)*
- *when (child's name) got implanted er it was like the basic things attention to sounds (...) getting some more vocabulary some more language some words and him and babble work and things like that...(F9)*
- *I think I've made the right decision and he loves his implant and he says I like my implant sometimes (laughs)... `cos it helps me listen (laughs) (F10)*

In summary

- **This parent summarises the whole experience succinctly:**
 - *‘so actually auditory neuropathy to me was a real pain in the *** (laughs)’ (F4)*
- **Better understanding what parents are going through will hopefully help the clinicians working with these families support them better and make the whole experience more tolerable for them**



Thank you!