



Auditory Neuropathy Spectrum Disorder in Infants; Experiences of Parents whose babies were identified with Auditory Neuropathy Spectrum Disorder through the Newborn Hearing Screening Programme

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Citation for published version (APA):

Uus, K., Young, A., & Day, M. (2011). *Auditory Neuropathy Spectrum Disorder in Infants; Experiences of Parents whose babies were identified with Auditory Neuropathy Spectrum Disorder through the Newborn Hearing Screening Programme*. National Deaf Children's Society.

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Auditory Neuropathy Spectrum Disorder in Infants

Experiences of parents whose babies
were identified with Auditory Neuropathy
Spectrum Disorder through the Newborn
Hearing Screening Programme

Final Report – April 2011

Kai Uus, Alys Young, Marianne Day



The University of Manchester



Our vision is of a
world without barriers
for every deaf child.

The Research Team

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Acknowledgements

The authors thank the families who participated in the study, the Teachers of the Deaf who supported the study and helped with recruiting families and National Deaf Children's Society for their support.

Funding

This work was funded by the National Deaf Children's Society.

The views expressed in this report are, however, those of the authors and not necessarily those of the National Deaf Children's Society.

Ethical Approval

Ethical approval was granted by the Ethics Committee of the School of Psychological Sciences at the University of Manchester (Ref MHS/80470). As sampling was partly done through Teachers of the Deaf within Children's Services Departments, ethical approval was also obtained through the Association of Directors of Children's Services (Ref RGEo8 11 29).

¹ The University of Manchester is a member of Manchester Academic Health Science Centre, a partnership between six NHS organisations in Greater Manchester and The University of Manchester.

Executive Summary

Experiences of parents whose babies were identified with Auditory Neuropathy Spectrum Disorder through Newborn Hearing Screening Programme

Twenty-one hearing families whose children had been identified with Auditory Neuropathy Spectrum Disorder (ANSD) through the Newborn Hearing Screening Programme participated in the first ever study looking at parents' experiences within this context.

The insight into parental perspectives is vital. It has potential to help the professionals who are used to working with families of children diagnosed with 'typical' hearing loss, to appreciate the similarities but also the marked differences between those two conditions. It serves also to recognise the specific needs of the families of ANSD children and to give parents a voice in the debate about how best they and their children should be supported.

The research was funded by the National Deaf Children's Society, carried out at the University of Manchester and was approved by ADCS and the Ethics Committee of the School of Psychological Sciences, University of Manchester.

ANSD in the wider context

In England the protocol of the Newborn Hearing Screening Programme only facilitates the identification of ANSD in babies who spend 48 hours or more in the Special Care Baby Unit. It is very common that these children have been born from complicated pregnancy and/or labour, experience a stormy perinatal period and a considerable proportion will end up with serious medical and developmental difficulties. Parents typically found themselves overwhelmed by the medical complications and at the point of diagnosis hardly ever saw hearing as an important issue, *let alone* a priority. The predominant feeling was that ANSD was yet another thing on top of everything else.

Information and lack of information

All parents talked about information with regard to ANSD, about how to get it and how to make sense of it. The main quandary was the general lack of information. There were different levels to that lack of professional information: firstly, as ANSD in infants is a relatively newly described, low-prevalence condition with a wide range of phenotypic manifestations, there is a genuine gap in knowledge that hopefully will be bridged through further research in future years; secondly, there is a lack of expertise, or more accurately the patchiness of expertise within audiologists, teachers of the deaf, ENT surgeons, speech and language therapists and other hearing-related professionals who the parents in general expect to have more expert knowledge; thirdly, parents described an almost complete lack of awareness amongst general health care professionals (e.g. GPs, paediatricians etc).

Parents also reflected on how they wanted the information to be given to them. They were understanding about the lack of information in professionals as they realised that ANSD in infants was a rare and relatively new entity, but they wanted the professionals to be honest about what they did and did not know. Parents sought family-friendly information that was given to them honestly, sensitively

and competently. This included being honest about what was uncertain, what was unpredictable and the limits of current knowledge. Parents had contrasting views on whether or not all information should be given to them at once or should it be delivered gradually. They also differed in how optimistic or pessimistic the prognosis ought to be, given that at the point of diagnosis it is impossible to give accurate predictions on the hearing thresholds as well as future perceptual abilities. The outcomes range from normal or near-normal hearing thresholds with little difficulty in speech perception to severely distorted speech perception and/or profound deafness. In general parents did not find emphasising, or rather over-emphasising, one extreme of the continuum of the possible outcomes, helpful.

Making sense of the diagnosis

ANSD is a perplexing condition. The label captures the actuality that it is really a range of different conditions with a host of different aetiologies, sites of lesions and most importantly, a continuum of different outcomes. There are important aspects of ANSD that differ from 'typical' sensorineural hearing loss. Most important of these is the paradoxical discrepancy between the hearing threshold and speech perception. The child may seem to hear relatively well and react to environmental sounds, but have disproportionate difficulty in understanding speech. The other great paradox is that auditory function may change (improve as well as deteriorate) over time and, in many cases, even fluctuate considerably from day to day.

Hence, it was not surprising that parents struggled making sense of the diagnosis, explaining it to their families and friends particularly as the child's reactions to sounds were inconsistent and did not follow the pattern of what people expect in a deaf child. Many parents found comparisons with either deaf children or hearing children quite unhelpful, which of itself was a cause of stress. Wishing one's child was just typically deaf, was not uncommon.

Wait and see

As mentioned above ANSD in infants is a relatively newly described, low-prevalence condition which is considerably complicated by the fact that it manifests in a spectrum of phenotypes which are impossible to determine in a newborn or very young infant. Additionally, a proportion of children are diagnosed with ANSD at birth due to delayed maturation and may experience a considerable improvement in their perceptual abilities, but electrophysiologically (i.e. normalisation of their auditory brainstem response) as well.

Parents were usually told to 'wait and see'. Whilst a 'wait and see' approach is clinically justified and hasty intervention following the same timelines commonly seen in 'typical' sensorineural hearing loss can be potentially harmful, parents had differing attitudes to waiting and seeing. Families differed in their acceptance to wait before management decisions were made: i) happy to wait; ii) willing to wait with some reasoning to support this stance; iii) unhappy to wait for various arguments behind it. Generally speaking, these differences reflected varied abilities or willingness to tolerate uncertainty.

Making decisions with regard to communication/support/management

Following on from the real diagnostic and particularly prognostic difficulties, choosing the right communication/support/management pathway for the child with ANSD is very challenging.

Parents described their various journeys through decision making. Deciding whether to choose a visual form of communication/visual language, hearing aids, cochlear implants or a combination of these was obscured by diagnostic uncertainties. These decisions were further confounded by the lack of knowledge and expertise of the professionals as well as misinformation from the internet, low levels of awareness and overall confusion of their own extended family and close community.

Parents who understood that there was no one-size-fits-all miracle answer for all ANSD children, seemed to cope better with these difficult choices.

Principal Conclusions

ANSD in infants is a relatively newly described, low prevalence condition with a continuum of phenotypic manifestations. When identified through Newborn Hearing Screening Programme, most parents attach relatively low priority on the diagnosis as predominantly their children have experienced or are experiencing other more pressing medical problems. It is only later when the child's overall health has stabilised when parents begin to prioritise hearing. The lack of knowledge, expertise and awareness of ANSD is common. Parents in general are understanding of that, they just want the professionals to be honest about what they do and what they do not know. They want the information to be given competently and sensitively. Parents differ in when they want to get the information and how 'hopeful' the information should be. Families find making sense of the diagnosis challenging and struggle in explaining it to people around them. They also find determining the identity of their child very challenging. After the diagnosis the parents have to endure a period of 'wait and see' before it is possible to determine the outcome of their auditory function. Parents can be split into three different groups in their attitudes towards this waiting phase, with some happy to wait, some willing to wait and some very unhappy to wait, mainly as they are worried the waiting will be detrimental to their child's development. Parents have to make difficult decisions with regard to the communication and management choices. These choices are particularly complicated by the disagreements in expert opinions and even more importantly by the range of phenotypic manifestations requiring different approaches.

Introduction

This research project concerns parents of children who were screened through the national newborn hearing screening programme in England (<http://hearing.screening.nhs.uk/>) and who subsequently were told that their child had an ANSD. Although ANSD is not a new condition it is one that thanks to newborn hearing screening is now routinely detected at an early age. As such, it is a diagnosis which is becoming much more commonplace. Therefore new questions and challenges arise about how best to support parents in making sense of the implications of ANSD for their particular child. It cannot simply be assumed that what we know about parents with deaf children will hold true for parents with a child with ANSD. For professionals who routinely have formed parts of the early intervention matrix for families with deaf children, ANSD is likely to be largely unknown territory that stretches the boundaries of their experience and expertise.

In this context, the National Deaf Children's Society in the UK commissioned a piece of research² that set out to ask families, who had been told their child had ANSD following routine hearing screening, about their experiences. They discussed what had happened to them, their hopes and fears, their coping mechanisms and their advice to professionals and other families who find themselves facing similar challenges in the future. The results of this study are an important missing piece in helping to move forward the agenda of what counts as good practice in supporting families whose children have an ANSD and in setting the agenda for future research and intervention studies.

2.1 Background

Auditory neuropathy is a term widely used to label a spectrum of auditory dysfunctions that are typically observed on audiological testing as the presence of normal evoked otoacoustic emissions and/or cochlear microphonic (implying relatively normal outer hair cell function in the cochlea), with an absent or severely abnormal auditory brainstem response³. Though not a new condition per se, it is only thanks to the recent routine use of otoacoustic emissions and auditory brainstem response in the clinical setting that differentiating this condition from 'typical' sensorineural hearing loss has become widespread. Possible sites of lesion include the inner hair cells, the inner hair cell synapse to the afferent nerve fibres, auditory neurons in the spiral ganglion, auditory nerve, brainstem auditory nuclei or any combination of these (Starr *et al.*, 1996; Berlin *et al.*, 1998). In June 2008, a panel of experts met in Como, Italy at the Newborn Hearing Screening 2008 Conference to develop *Guidelines for the Identification and Management of Infants and Young Children with Auditory Neuropathy* (Northern, 2008). The panel reached a consensus to refer to the condition as **Auditory Neuropathy Spectrum Disorder (ANSD)**.

Older children and adults with ANSD display a wide range of clinical features with behavioural hearing thresholds ranging from within normal limits to profound hearing loss, and typically poorer speech perception than would be predicted from the behavioural audiogram (Rance *et al.*, 2009). Speech perception ability in children with ANSD varies greatly. While some perform at levels similar to children with comparable degrees of 'typical' sensorineural hearing loss, others show very little

² This study was undertaken jointly by staff from the Audiology and Deafness group (<http://www.psych-sci.manchester.ac.uk/research/groups/audiologyanddeafness/>) and the Social Research with Deaf People group (www.manchester.ac.uk/sord) at the University of Manchester.

³ Evoked otoacoustic emissions and cochlear microphonic are techniques to assess the function of outer hair cells of the cochlea in the inner ear; auditory brainstem response is a method to measure the electric signals that are induced by the auditory nerve as it responds to sound.

or no speech understanding despite sufficient sound detection (Rance *et al.*, 2002). In England, each year approximately 600 babies are identified with congenital permanent bilateral moderate or greater hearing loss. Around 10% of these present with the combination of audiological test results characteristic of ANSD (Uus & Bamford, 2006).

ANSD is found in well-baby populations, most commonly due to autosomal recessively inherited genetic mutation (Varga *et al.*, 2003; Delmaghani *et al.*, 2006) however its prevalence in the well-baby population is believed to be low. Consequently, many screening programmes around the world, including that in England, are not suited to identify ANSD in healthy babies because they rely exclusively on otoacoustic emissions as a first-stage test, which are typically normal in subjects with ANSD. In England, babies identified with ANSD are, therefore, found through high-risk screening. Prevalence figures for ANSD in high risk populations are varied: 0.2% (Rance *et al.*, 1999) to 24% (Berg *et al.*, 2005). The variation is likely to be a result of the disparity in diagnostic approaches as well as characteristics of the underlying population.

The routine identification of newborns with ANSD poses a new set of challenges in providing appropriate parent support and clinical management. Whilst clinical management is attracting international attention as evidenced by the international guidelines (Northern, 2008), there is little research evidence available to inform how clinical and early intervention professionals should best support families whose children have been diagnosed with ANSD. There are no studies that have sought to address this point from the perspective of parents themselves, nor to place them in the position of the generators of new knowledge for this new circumstance. Although there is an increasing body of work that has sought to understand parents' experiences of early identification of deafness (Young, 2010), it cannot be assumed that this evidence is relevant to the experiences of parents of children with ANSD, just because the mechanism and timing of identification is coincidental.

2.2 Research Aims

The overall study was designed to answer the following research questions:

- 1) How do parents construct and frame their experiences of having a child with ANSD?
- 2) What are their experiences of diagnosis and early intervention, both in terms of the care pathway and the personal impact in relation to their particular family?
- 3) What evaluations do they make of these experiences?
- 4) What advice or suggestions would they give for the improvement of this process for others?

2.3 Research Methods and Sample

2.3.1 Methods

Participants in the study were all family members of babies who had been identified through Newborn Hearing Screening Programme in England. More specifically, all babies had been screened according to the Special Care Baby Unit protocol that incorporates both otoacoustic emissions and auditory brainstem response.

The definition of ANSD was based on absent or abnormal auditory brainstem response and presence of otoacoustic emissions and/or cochlear microphonic on a diagnostic assessment following the screen, consequently cases of transient ANSD were included.

Data were collected between 26 February and 10 September 2009.

An exploratory qualitative approach was adopted based on narrative. Parents were invited to tell their own stories, in their own words within the broad framework covering the experiences of the pregnancy, labour, early perinatal period through to hearing screening and identification with ANSD; the experience of early intervention and professional support; their advice to other parents and professionals engaged in the same process. The interviewer's role was to clarify points in the narrative as it progressed to ensure information was collected about comparable events across all interviews undertaken; to support the narrative telling through empathic engagement with the teller; and to record the interview for later analysis. In this way, parents did not respond to a set of predefined questions in which to fit their experience but rather were given the opportunity to make decisions about what is meaningful and important in their experiences and to set the criteria by which they would want their experience to be understood and evaluated.

Parents completed a narrative-based interview lasting on average 60 minutes (ranging from 37 to 80 minutes). All interviews were conducted in English in the homes of the families. The interviews were audio recorded for later analysis. In addition, parents completed a simple questionnaire to collect sociodemographic information and more detailed information about the child's medical and audiological profile.

Data were transcribed verbatim and thematically analysed. An initial open coding procedure produced 20 codes which were later collapsed into 17 final themes for analysis. A list of thematic codes and their initial working definitions can be found in the Appendix (Table 1). For a more detailed description of the research methodology see Uus, Young & Day (in press).

2.3.2 Sampling

A purposive convenience-based approach to sampling was used in order to recruit those parents likely to have the most relevant experiences to the study's aims and in order to capture as broad a range of participants as possible.

One hundred and ten Teachers of the Deaf in England were contacted by the researcher and research

packs (containing parent letter, consent form, information sheet and detailing the study) were sent to them. Teachers of the Deaf were asked to forward the study packs to the parents whose children had been identified with ANSD. At this point, the name and address of the family were unknown to the researcher. Families who wished to be involved in the study were asked to complete a response sheet with their name and address and send it directly to the researcher; an interview would then be arranged. Additionally, information of the study was available from the National Deaf Children's Society, the funders of the research, inviting volunteers to contact the researcher in order to participate in the study.

Thirty-two families initially made contact to volunteer for the study. Of those 21 were interviewed. Out of 11 that were not interviewed: 4 did not meet the inclusion criteria as their child's ANSD had not been identified through newborn hearing screen; 4 had to cancel the interview for a number of reasons; 3 did not respond to follow up contact by phone or email.

2.3.3 Characteristics of the sample

Ten families were recruited through Teachers of the Deaf and 11 families through the National Deaf Children's Society advertising. Respondents were encouraged to choose who should be present at the interviews. Consequently, 21 interviews equated to participation from 25 parents/caregivers/extended family. In 16 interviews, only the mother was present, in 1 only the father was present and in 4 interviews, both mother and father were present. The unit of analysis in this study is the interview rather than the individual respondent. All participants were hearing parents.

The demographics of the interviewees are given in Appendix (Table 2).

3.1 ANSD in the wider context

Considering the complicated pre- and particularly perinatal history (see Table 3 in the Appendix for details), ANSD was a relatively insignificant part of that bigger story which encompassed experiences such as complicated pregnancy; a stormy perinatal period due to premature labour; very serious medical deterioration soon after birth including the child's dramatic fight for survival and, in case of multiple births, loss of one of the babies.

Without exception, all parents in our sample experienced extreme worry, fear and/or loss due to their child's or children's serious medical problems well before ANSD was identified. This wider health context remained a significant part of their experience of having a child with ANSD and predated the unexpected audiological diagnosis. As such, it created a particular backdrop that was dramatically different from that of most 'typical' sensorineural hearing loss children.

Results suggested that after the shock of agonising over the stormy perinatal period, the realisation of the potential long-term health problems added another dimension to the context in which parents discovered their child's ANSD.

Expressions of being overwhelmed were a commonly recurring theme amongst parents when talking about their reactions to the diagnosis of ANSD. However, being overwhelmed could mean different things to different parents and had a range of origins. For some, it was not that the diagnosis of ANSD was overwhelming but that it happened within a much larger experience of being inundated with new and rapidly changing information about their child's varying health issues, a multitude of medical appointments and interactions with a vast constantly expanding array of various health professionals.

The common feeling of 'deafness on top of everything' was articulated by almost everyone in our sample. Many parents also described how their broader experience of the many health conditions and problems of their children was also useful in helping them put the diagnosis of ANSD in perspective. As one parent (F2)⁴ pointed out, dealing with hearing problems actually felt to them to be more tangible than many of the other unknowns which seemed to surround their child's health.

Most parents reminiscing about their experiences of having found out about the diagnosis, recalled that they had not seen ANSD as a high priority concern. Most parents expressed very explicitly that, compared to what the child and the family had been through, hearing loss had seemed '*kind of minor*' (F17), '*we wasn't that fussed*' (F11); '*didn't seem like a big deal to me at that point in time*' (F19), '*being deaf was a tiny little thing*' (F10); '*after what we'd been through it was like I don't care (laughs)*' (F12)

The comparison with both past traumatic medical experiences and potential future health uncertainties was a key context in which parents formed judgements about the extent to which they should be concerned about their child's diagnosis of ANSD. As one parent (F16) emphasised, for example, it was a 'relief' that their child has auditory dysfunction and '*... erm at least he's not blind.*'

⁴ An alphanumeric code was used to refer to the members the participating family, e.g. F1 denotes any interviewee from the first family we interviewed etc.

The extent to which ANSD was a priority concern for parents was also mediated by time; both through the resolution of other difficulties and the gradual manifestation of the effects of ANSD as their child matured. For some parents, their child's medical problems either became defined with more certainty and/or parents became more confident in their knowledge of the likely consequences of them. This greater degree of certainty created space for their consideration of the consequences of the ANSD diagnosis and its implications gradually came to the fore. Also with the passage of time and their child's maturity, the implications of ANSD became more obvious and more apparent. One parent (F1) summarised the sentiment of the majority in the sample: *'turned out to be the worst of our worries... in the long run...'*

In Summary

Parents of children identified with ANSD through newborn hearing screening do not in general prioritise hearing very highly at the point of diagnosis because of other competing and more serious medical problems they have experienced and/or are still experiencing. Initially, the impact of the diagnosis is fundamentally mediated by these experiences and the uncertainties of their child's future rather than by a consideration of the potential impact of deafness. ANSD is not seen as a standalone diagnosis but as a part (in some cases insignificant part initially) of a much bigger picture.

3.2 Information and lack of information

The most common concern all parents emphasised was the need to improve knowledge, expertise and information about this perplexing condition. In emphasising this point, parents were reflecting the current state of knowledge. ANSD in infants is a fairly recently described, low-prevalence condition with a wide range of phenotypic manifestations and there are genuine gaps in knowledge. Establishing the phenotype of ANSD for an infant diagnosed soon after birth and giving a prognosis for auditory, communication and developmental outcomes is very challenging, practically impossible in most cases. Parents certainly picked up on the need for more research however, only one parent (F3) felt that identifying a condition, that so little was known about, was of questionable value:

there's something amiss somewhere and really they...they should research...yeah if they're going to diagnose a condition they should know about the co...there's no point diagnosing it and then saying actually we know nothing about that

Parents also drew attention to the gaps in knowledge which were related to practice rather than to research. Many highlighted the need for more continuing education amongst professionals. Whilst there was some sympathy for the challenges the relatively rare condition presented for those professionals seeking to support parents, parents did not appreciate professionals trying to hide their lack of knowledge and expertise:

cos I think it is quite easy for people to fob you off when they don't know about it themselves in the profession you do find that. yeah I would rather than they were honest and said I don't really know a great deal about it but I'll try...to get some information for you and see what I can find... yeah you can yeah I mean when you've been through it a few times you start to see the signs (laughs)... because they just change the subject and they'll start talking about something else...erm but I think you do have to be persistent (F19)

On the contrary they valued honesty and welcomed the professionals' attempts to seek more information to plug their own gaps in knowledge:

but er...(name)'s just erm...I think it was not last week but the week before she went to a conference down in London.. for auditory neuropathy I don't know if you've been or not but she says it was fantastic for her 'cos she got to mix with teachers...you know in a similar position to her...what do you do with them what works what doesn't work...she came back and she was going oh we're going to try this with (child's name) ooh we're going to try this with (child's name) and you can see the excitement I think there needs to be more of that I think there really does need to be...you know..(F6)

However, single professionals skilling themselves up was not seen as a universally helpful solution, if the whole support system which surrounded parents of children with auditory neuropathy was not equally skilled. Without systemic understanding, the enhanced knowledge of one professional was of little use:

and even GPs...they could have something you know...actually (---) a point yeah the GPs still don't know really what's going on...she's deaf. First thing that happens with (child's name) if she has a problem with her ear it's straight up to the (hospital name) and I don't think it's necessary half the time...it's just a case of she needs a nice dose of antibiotics and sort her out (F6)

In this respect, some parents suggested that perhaps it would be better to establish specialist centres who deal with children with auditory neuropathy where parents could feel confident that all professionals they came into contact with were up to date and knowledgeable.

The dissemination of useable information to parents was also a crucial issue to all the parents we spoke to. It was identified as fundamental to providing better services to this population. Parents' emphasis was on information that was family-friendly and conveyed in accessible language⁵:

just...a parent friendly bit of information with someone saying just the basic facts in layman's terms whereas there isn't doesn't seem to be anything like that (F15)

don't use so big words really or erm....give information on...things and stuff what it actually means and other things that could happen and what...the stages that you go through and...stuff like that what support's out there and...(F17)

so it wasn't too technical it was just not...you know just not what you needed you just needed a few facts sometimes and they just weren't...good at doing and then following it up with information afterwards you know (F14)

For the information to be useable, parents suggested that it contain explanations that were illustrated with examples, or analogies that they could easily relate to:

I'd use the speaker...broken speaker cable one... as an example of what your child's hearing (F6)

Although facts were important, information that was sensitive was important too. This sensitivity also encompassed getting the language level right as much as speaking with honesty and care within the text.

When it came to how much information to give and when, there was no consensus. Some parents wanted all the information right away:

er trust me...believe what I'm saying...er...give me as much information as possible...er give me contacts or useful links or papers to read....er I know everybody's different and there are some parents that don't want that but I do...er...(F9)

⁵ As a result of this study the research team in collaboration with NDCS has produced a companion publication for parents to plug this information gap ANSD: *To Parents from Parents*.

Whereas others found it overwhelming and appreciated a more gradual approach:

I would have preferred it to be more gradual I think if they'd have eased... 'cos it like I said in the beginning there was nothing and then it was...all these people started coming all of a sudden...I think they should ease you into it do a step at a time rather than pushing all these things on straight away (F19)

As to what format the information should be in, most parents expressed the need for leaflets, but also for relevant regularly updated websites that give UK-specific information:

yeah and it would be lovely if there was one that was sort of more relevant as in like what we do in the UK...erm and I mean I do know that people do go on that erm...that Yahoo one obviously from the US but there's even people going on from India and the UK and different countries...it would be nice if there was something that was more relevant to us 'cos you know obviously we've got...erm you know the PCT regulations the NICE regulations and things like that (F14)

Naturally leaflets and websites alone were not considered enough. More interactive options such as the chance to ask questions from experts and/or families with children diagnosed with ANSD, forums and networks were also suggested.

Because of the unfeasibility of predicting hearing thresholds and perceptual ability in very young infants with ANSD, a very specifically ANSD-related issue that a few parents raised was the need to balance how 'optimistic' or 'pessimistic' the information given to the family should be. In general parents did not find emphasising, or rather over-emphasising, one extreme of the continuum of the possible outcomes, helpful.

just be honest with people from the start but...maybe give them hope as well (F15)

so it was best we found out so early I just don't...think it should have been so black and white right from the start.... all they had to say was ..yes in some cases this does happen but...you know it can go either- it's just this it's just this barrier the whole time of...you know how bleak the picture was.. (F5)

In Summary

Parents understood the reasons for lack of knowledge, expertise and awareness about ANSD in professionals, but insisted on honesty about what the professionals did or did not know. They wanted to get information about ANSD delivered to them sensitively and competently in an accessible language and format.

Parents differed greatly in the finer details of their information needs, such as whether information ought to be given gradually or all at once and, even more importantly, in the face of wide prognostic range, how to balance the too optimistic/too pessimistic message given to them at the point of diagnosis.

3.3 Making sense of the diagnosis

All families whose babies were diagnosed with ANSD struggled to make sense of the diagnosis. Very importantly, and not surprisingly, no-one in the sample had heard about the diagnosis.

Parents were often told that ANSD was either a 'new', 'rare' or 'unusual' diagnosis and that it was very difficult to manage. In addition, they were often told that very few other children were diagnosed with it. On top of the uncertainty of diagnosis this was interpreted by parents as a lack of knowledge and interest from professionals as well as sometimes leading parents to feel like isolated cases who could not draw on other family support. As one mother said she didn't think anyone else had it. (F5)

What parents recalled about these initial explanations is that the structures of the inner ear were working but that the messages were not reaching the brain in a coherent way. This set up a strange relationship between being able to hear and not being able to hear which was very difficult to come to terms with. In an attempt to make sense of it parents tended to find useful explanations which related to concrete and familiar experience such as the idea of faulty wiring or a song that has been jumbled up.

and then she explained to me about the fact that...it's everything seems out of synch with aud- a bit like when you're playing a song and all the...all the parts of the song are all jumbled up and...to me that was the clearest...way of explaining...something like that (F19)

imagine a speaker everybody's had a broken speaker at some stage where the wire doesn't quite connect properly... and that's the one I use and everyone says I get what you mean (F?)

The uncertainty of an ANSD diagnosis was a defining factor for all of the families in the study and it is experienced in a number of different ways. As previously discussed, a definitive diagnosis of ANSD did not imply a definitive prognosis. Consequently one family questioned the helpfulness of an early diagnosis at all.

and I was angry that they couldn't tell me anymore about it because I thought well how can you give somebody a diagnosis if you don't know yourself what it is (F19)

However, 20 out of 21 the interviewed families said that they were glad that they found out about their child's ANSD at the newborn hearing screen because it had allowed them to put support in place and because many believed that they would not have noticed that there was a problem until the child was much older as most of the time they appeared to be hearing.

Even amongst those parents who found the early diagnosis of ANSD to be helpful, it could still seem counterintuitive. The evidence of their everyday experiences of living with their child seemed to contradict the diagnosis, even when they knew that ANSD could imply a constant change of hearing abilities.

but also we were still very cagey about the whole thing because we were rattling things and he was turning round to them...we were calling him from downstairs upstairs he was turning

round...erm...one erm... thing that really sticks in my mind we were playing round in here in the living room...his granddad arrived tapped on the window...jumped...we're thinking...they're telling us lies (laughs) at this hospital we don't get it (F4)

Another aspect of confusion for some of the families whose children had milder cases of ANSD was the spectrum nature of the condition. Two families in particular questioned whether their child's lack of a hearing problem in everyday life meant that the diagnosis was wrong especially in a case where the family had contact with another child whose ANSD was manifestly worse than their child's.

that he's not quite doesn't seem quite right but other than that he certainly doesn't seem to have the problems that erm...(name) has for example.... and...you know I mean maybe they're two different extremes maybe they have both got auditory neuropathy and maybe (child's name) is very very mild and (other child's name) is very very....bad (F12)

As well as finding ANSD difficult to understand or to come to terms with themselves parents were faced with the necessity of explaining it to others. A number of the families said that they tended to tell people their child was deaf rather than go into any details. However, this has led to situations for some parents where they felt like frauds for using this explanation when their child appeared to be hearing.

if they then see him respond to sound and you've told them he's deaf they're like but but... but...I...I...it's not worth it you know ...close friends I try to explain it to them...and you know every now and again you'll say something and...you kind of think yeah.. it's ...it is slowly sinking in with people who see him a lot...erm...but when you just meet people for the first time it's just too... hard.... (F2)

Not surprisingly this strategy did not work with extended family, close friends and neighbours and many parents talked about their frustration of being asked the same questions over and over again:

yeah and you start to feel like a stuck record after a while (laughs)...yeah having to go through it all again and then still I don't think they get it...what it is actually (F19)

oh god...they oh they just wind us up every time they go ...oh what's that what's that they just expect an answer you know every time you go to an appointment they expect it's like that's it you know the answer and...and you can't explain it to them they don't listen and they don't understand it and it's just like you can't be bothered to explain 'cos they won't understand it... so...you just say well you know we've got some results and it looks like maybe this...and then they ask questions that you know they're not gonna...follow (F17)

Many parents discussed having to insist to family members and friends that their child indeed had a hearing loss because people often assumed that their child could hear because he or she was so obviously responding to sound.

Family will look at him and say oh he's not deaf there's nothing wrong with him... but then you

get.. to the point where I don't want to go around being negative 'cos obviously I don't want him to be deaf (F13)

I mean honestly his Granddads' the worst...when he doesn't have his hearing aid in well he's hearing that alright he's talking to me alright...yeah I know but he's not...you know you have to kind of.. it's hard...it's really hard (F4)

The majority of parents said that when they talked about ANSD no one had heard of it, including GPs and other parents of deaf children. This meant that the parents were having to give explanations in a context of total unfamiliarity using familiar concepts from 'typical' sensorineural hearing loss which can then be at odds with the child's actual behaviour. A number of parents were more comfortable in talking about their child's hearing loss later when there was an outward indicator of deafness such as a hearing aid, cochlear implant or even a definite diagnosis or audiogram to support their explanation.

...it has changed I think now I just say he's got he's deaf...by his hearing aid and an implant and his deafness is auditory neur- is a kind of deafness called auditory neuropathy...so that's what I describe it as that's how I...it's easier now because...he's got a hearing aid he's got a and he's got an audiogram.. (F10)

A number of parents talked about how it might have been easier for them if their child had had a *straightforward diagnosis of profound deafness* (F1) even though this could mean that the child's hearing would be worse than it actually is.

it would in a way it perhaps would be easier for us sometimes I think if someone would say she's profoundly deaf she's never gonna speak she's never gonna hear...and then we could almost....get over it...and move on..... it's like a bereavement you can bereave for her hearing loss but as it stands now I can't because she might get better this is what we've been told she might she might not...so it's (F15)

Because infants and children with ANSD often react to relatively soft sounds, a diagnosis of a 'hearing problem' can be extremely difficult to come to terms with. Differently from 'typical' hearing loss, the child may hear sounds but cannot understand speech. Like somebody can hear the sound of a radio, but because it is off the channel, is unable to understand the words. One family described it '*dyslexia of hearing*' (F2).

In summary

Parents in general had a difficult time making sense of the diagnosis. They had never heard about ANSD before. Because of their often paradoxical auditory experiences with their child, they struggled with the comparisons with 'typical' sensorineural hearing loss and found it hard to establish an identity for their child in relation to both hearing and deaf children. Families appreciated explanations that they could relate to in their every day life both in understanding the conditions themselves as well as providing explanations to their relatives, friends, and neighbours.

3.4 Wait and see

For parents of children with ANSD, this significant unpredictability was a major challenge to decision making about best support for their child. Part of this challenge arose from the fact that the usual pathways of intervention and support for a child with a diagnosis of ‘typical’ sensorineural hearing loss, do not easily fit the uncertainty of a child with ANSD. For example, many parents were impatient that their child be fitted with hearing aids or implanted before the window of opportunity for speech development, as they saw it, closed. Whilst this is an established approach in ‘typical’ sensorineural hearing loss, it is not automatically transferrable to ANSD. Diagnostic challenges (difficulty in establishing a hearing threshold coupled with the relative clinical meaninglessness of the hearing threshold as it does not correlate with speech perception ability) as well as short-term and long-term changes in the auditory abilities all postpone the start of auditory management considerably.

Therefore for many parents, the fitting of hearing aids or the decision to implant appeared delayed or postponed. Even when the reasons for this waiting were understood, the wait could be agonisingly difficult because parents felt that no progress was being made.

A few parents in the study had also had a diagnosis of profound deafness before the ANSD was identified. They had, therefore, already begun to imagine the future in these terms. A diagnosis of ANSD had changed again the parameters of their expectations and what it was they felt they were adjusting to. Furthermore, the uncertainty of prognosis meant that for all of the parents in the study, there seemed to be little solid ground from which to make any kinds of decision because the very terms on which they might make decisions (e.g. about language, communication) had the potential to change. Indeed it is a striking finding in this study that some parents stated they would have preferred a certain diagnosis, be it with a perceived negative outcome, to one of persisting hope for a perceived positive outcome. There was a kind of comfort in knowing what one was dealing with, even if it felt like it had serious consequences for their child, in preference to accepting uncertainty at the price of a potentially less serious outcome.

This attitudinal paradox experienced by some parents was also reflected more widely in the common experience of many parents of service trajectories just not fitting their children. Many parents expressed the view that it would be a lot easier if their child was just ‘*straightforward deaf*’ (F1). Even when parents attended support groups with other parents of deaf children, they did not necessarily feel supported because their child and their expectations and concerns were not those of other parents around them. This was a particularly difficult problem for those parents who might be waiting for their child’s hearing to improve, or who saw evidence of those episodes when their child could hear. These kinds of hopes and expectations created very different terms of engagement with support services than those of many parents of deaf children.

Although most parents understood the uncertainty associated with prognosis and the establishing of hearing thresholds, they were divided in their *willingness* to wait before the decision of the communication/support/management route was made. There were three kinds of response to this situation: i) happy to wait; ii) willing to wait with some specific reasoning to support this stance; iii) unhappy to wait.

Those families who were happy to wait were more often than not simply hoping that their child's hearing would improve or become what they considered to be normal. For others, the willingness to wait was done with clear awareness that although waiting might result in delayed development, waiting gave them time to come to terms with everything and allowed them *'that time to treat him as a baby... (...)... we've dealt with it a lot better than say...if they'd just told us he needs hearing aids from the day one...'* (F8). For other families the willingness to wait arose because they had picked up how unsure the professionals were about the decision to start intervention and they were not prepared to 'experiment' on their young child (F2).

However many parents were unhappy to wait as they were concerned that it might be detrimental to their child's development. Families (F4, F16, F17) very clearly stated that their discomfort with waiting had been mainly due to the information they had read and/or heard about the need to start intervention as early as possible. Often this information was from sources dedicated to 'typical' sensorineural hearing loss.

look I'm getting really nervous now that time's marching on and nothing's happening...I've you know been told and I've read you know that we need this hearing aid thing and we need to get going on all this 'cos he's getting older...(F4)

One family (F5) articulated a very specific concern that waiting before starting intervention was going to hurt their child's speech development, particularly as they did not have full confidence about the diagnosis of ANSD and suspected that their child had 'typical' sensorineural hearing loss after all.

Another family (F10) did not like waiting not just because of its potential detrimental effect on a child's speech development, but because of other practicalities such as having the child keeping their hearing aids in once she is older.

In summary

All parents spoke about the uncertainty that surrounded the prognosis of ANSD in infants and about the rather long period of having to 'wait and see'. Families had different takes on that approach from welcoming the breathing period to enjoy their young baby to being very frustrated and worried about the potential negative effects this waiting could have on their child.

From the data it is not possible to say whether these different perceptions had to do with the way the clinicians had explained the condition to the family or with specific characteristics of the child and/or the family or the combination.

3.5 Making decisions with regard to communication/support/management

As already noted, choosing the most appropriate communication, support, management and intervention for a child with ANSD is yet another challenge for everyone involved. Parents extensively talked about frantically searching for answers and trying to make sense of the intervention dilemmas by talking to the professionals as well as doing internet searches only to realise that when it came to ANSD, even the so-called experts did not agree. Most parents talked about getting conflicting information from everywhere:

and she just said you know there's ..there's obviously sign language and cued speech and (?)... I didn't even know there was anything...so we wouldn't...I don't think we would have actually found it ... had we not.. had she not mentioned it ..to us and then we obviously started looking a little bit more into it .. and she did say there's a whole school of thought around.. hearing erm..... sorry....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 ...what do I do for the best.. ... I think we know what the options are so there might be... you know we might put her....see 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.. it might get worse (F1)

you know even if it's a you know...this is what it is and there is nothing that can be done... so...do your sign language do whatever go on with it you know or this is what it is and if he wears hearing aids it'll be lovely you know and he'll be able to hear everything and it'll be beautiful and you can go and singing and dancing and...you know la de dah...everything's fine you know whereas with this it's this is auditory neuropathy and one day he might hear this and another day he might hear that and there could be nothing and hearing aids might work hearing aids might not....cochlear implant might work cochlear implant might not you know and you kind of hear different things from different people because we've had people saying to us....cochlear implants... no good...and then we've had people saying to us ...that could be the way forward....and hearing aids no good hearing aids they might work you know and you're like woah... (F16)

Most families who participated in this research had opted for a combination approach to communication (Table 4 in the Appendix). However, in what follows we will look at the three main trends (visual communication, hearing aids and cochlear implants) separately, focussing on why some of these choices are essentially different from what we commonly see in the choices that are being made with children diagnosed with 'typical' sensorineural hearing loss.

3.5.1 Visual communication

Sixteen out of 21 families, had at least attempted some form of visual communication. This ranged from BSL to Makaton and included also some home signs and gestures. However, parents found choosing visual communication challenging and struggled with issues. Some of those are very similar to those of the families of children with 'typical' sensorineural hearing loss, some entirely different and ANSD-specific altogether. In addressing these challenges it is worth remembering that all of the parents in our sample were hearing and none had any previous experience or contact with Deaf people who used Sign Language.

In some cases the child's additional needs made adopting sign language difficult from a physical perspective, however in the following example the parent focuses on problems of expressive communication, rather than on receptive:

we started almost immediately after the diagnosis we had been lucky we had a support worker coming home and teaching me the basics...so we could introduce it...erm but (child's name) didn't really erm...take to it er...probably you know considering his hemiplegia and his visual problems...no wonder....(F9)

Because of the nature of ANSD, parents were sometimes advised against signing because as one parent reported being told "we're not sure what we're dealing with" (F4). What that comment actually implied, was unclear to the parent, but the underlying message to avoid sign language at that stage was clear. Other parents said they found it hard to be motivated to make a full commitment to use sign language if there was hope that the child's hearing would improve and he or she would start speaking. In this sense the use of signs or gestures was seen as an interim rather than a long term measure:

well the signing thing erm I was told by the...educational audiologist...that he should I should be signing I should be signing...and I suggested Makaton because...British sign language (laughs) ...I mean that's a language in itself it's like...saying to me you'll have to learn Portuguese to speak to your child and...I'm like I can't learn that I can't do it and I'm not prepared to do it I think we should...see whether he's gonna speak...and if not obviously and they kept going on and on and on about this signing and...I mean...everybody does it to their baby do you want a drink do you want something to eat where's the ball...you know everybody does that's basic gestures so I started using the Makaton (laughs) ..which they weren't happy about but I think...basic sign language like Makaton is so simple...it's what they need at their age and they use it now in his nursery anyway with normal children...(F5)

Some parents, very likely for the same reasons highlighted above, also expressed interest in learning cued speech but found it difficult to get access to the cued speech programmes.

Parents, who in the making sense of ANSD process assumed that their child would have a Deaf identity, found adopting sign language the easiest:

I feel we've done the right thing in assuming he was...deaf and carrying on down that route I

mean we're learning sign language....erm and have been using sign language with him...to some...degree...erm...from more or less when we found out....erm... (F2)

However, the acceptance that their child was deaf and in some cases would have a Deaf identity could be very hard for extended family or close friends to accept or understand. This was because the child's auditory behaviour, in particular responses to sound, seemed to challenge parents' assumptions and acceptance that their child was deaf. This kind of seeming contradiction could lead to those around some parents not being supportive of their choice to sign:

and then signing and then there's hello and goodbye and happy.. she learnt happy she doesn't do the sign but she learnt happy through...the singing that we do on a Tuesday morning and then I sing it to her every morning as well I say how are you today and she says happy...so she's happy so she can through that it's helped her a lot you don't realise sometimes you think my mum says why are you doing that sign language she's not deaf (F8)

There were practical difficulties also such as finding the time to attend courses as well as lack of funding for families whose child did not have an audiogram to evidence deafness:

Funding the early years because of funding...that is for deaf children and deaf children only (F12)

Also much like in some families of 'typical' deaf children, there were a few parents who were worried that sign language was not going to solve the child's communication difficulties:

erm and you know and I almost thought of looking at the bigger picture as in like I say you know you can't just walk into your corner shop and...can I have a 10p mix or whatever you know and...'cos people not everybody signs very few people sign really unless they need to.... (F14)

In spite of the challenges and hesitation, with hindsight, the overwhelming advice these parents in our study who had tried sign language would give to other parents of ANSD Children, was to seriously consider sign language. Sign Language, in these parents' view, had its place in supporting children with ANSD:

I would....I would say to them do as much signing as you can because if that child can hear... just a little bit he might have difficulty in speaking so you're still going to need sign language to communicate with him anyway....so do and learn as much sign language as you can (F12)

yeah yeah everything sign language anything that would you know make life better for them you know (F20)

3.5.2 Hearing aids

From clinical experience with adults whose late-onset auditory dysfunction is usually, not always, part of systematic neurological disease, the established approach in management and intervention was: in auditory neuropathy hearing aids are not beneficial (Starr *et al.*, 1996). Furthermore, the risk of damaging intact cochlear structures such as outer hair cells has been raised (Berlin, 1999).

This approach was challenged by Rance *et al* (2002) who showed that a considerable proportion of the children diagnosed with ANSD benefited from amplification evidenced by considerable improvement in their speech perception. The current guidelines recommend trial hearing aid fitting in children with ANSD (Northern, 2008). However the challenges remain.

The 'intuitive' candidacy advice from the extended family varied from getting hearing aids being an obvious choice '*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) to the reassurances that the child did not need amplification '*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). The latter was most likely to be caused by the discrepancy between relatively good hearing threshold and poor speech perception and/or fluctuations in auditory functions as repeatedly discussed above.

However hearing aids candidacy was highly controversial in ANSD management. Parents often felt they were left to carry the consequences of this controversy on their shoulders, they felt that though the overall consensus had long been that hearing aids did not benefit children with ANSD, the amplification was used when the parents were pushing for it. One family (F3) summarises it: '*she said...it was unlikely that he would have hearing aids erm and that usually with auditory neuropathy it's only when parents really push for hearing aids that they get hearing aids...*'

In some cases even through all the difficulties, parents actually found making this choice empowering.

Some parents found the rationale of using hearing aids at odds with what they had been told about ANSD, particularly as they have been told that sounds were distorted. One parent (F9) wondered: '*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*'. The other parent (F12) was equally puzzled by '*if it was the nerve that was the problem and he could actually hear ok... then by giving him a hearing aid...you might be amplifying the sound too much for him and he might not be able to tolerate that even and he still won't be able to hear any better erm...*'

Some parents however expressed that they understood the individual differences in ANSD children and how it related to hearing aid candidacy.

All in all, parents in general found the conflicting candidacy issues very frustrating:

it's really hard again but you know if he was deaf and he was going through the...erm...

hearing aids or the cochlear implant and he you know he just had a normal hearing impairment (laughs)...erm I could see it would be even more...useful and the support would be phenomenal because...they're all going through that you know but with (child's name) he's going through something slightly different to most of them because we hardly ever see the other children with auditory neuropathy it's usually the...the ones with the hearing aids and the cochlear implants you know and sometimes you do feel a bit sort of...on the edge if you like (laughs) (F12)

Parents told us about their expectations with regard to hearing aids as well as their positive and negative experiences once the child was finally fitted with hearing aids. A few expressed how hopeful they had been about hearing aids:

yeah but I still think like once he gets his speech he's responding and if he gets his speech and his hearing aids are there....hopefully fingers crossed we'll be... (F21)

keep them in and...his hearing will be...his hearing will be alright as long as he keeps the aids in....(F7)

However, others reflected on the importance of parents' expectations needing to be managed before their child was fitted with hearing aids:

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 because nobody had mentioned auditory neuropathy at all 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)

As seen from Table 4 in the Appendix, once the children were fitted with hearing aids, out of 16 cases from our sample of 21 where hearing aids were trialled, only five families found amplification very beneficial and three 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)

Even the families, who reported great benefits, described the pressure to make the child wear hearing aids:

(mimics strict voice) you've got to wear these aids the doctor says you've got to wear your hearing aids...if he doesn't want to wear it that's fine but he does...and he'll ask for them now and...he doesn't pull them out or...throw them out in the garden he knows that they're special and...we take care of them (F5)

I'm speaking for myself here I know you might be different (indicating husband)... for me he doesn't wear his hearing aids a lot...he pulls them out he's a pain in the backside (laughs) with his hearing aids he'd much rather eat them than wear them.. (F16)

Some parents, rightly or wrongly, were not quite convinced it was the hearing aids that had really helped the child. One family's (F11) account was particularly fascinating:

so it took you know...in all this time of pressing it was only the last time that they've admitted they've actually seen a difference with them in sort of thing erm so you kind of go...well yeah this is what we've been saying you know...now will you believe us sort of thing...erm (laughs)...I said... it was even a daft one that...he just wouldn't do anything at all but as soon as we put them in and he'd do everything absolutely correct but...it felt as if...it was such a large jump it wasn't the hearing aids it was just the fact that he clicked into it and decided to start doing the task...so even then it's...it's be wary...I don't necessarily think it's the hearing aids that allowed him to understand the speech I think it's more that just kicked him into life to...to actually do the task...it's not that he does everything but

Other parents (F21) were also not convinced their child even needed hearing aids as their hearing appeared to be relatively good anyway. In some families, particularly due to the fluctuations in hearing (F16), two parents had contrasting views on whether the child needed and benefitted from hearing aids.

A few parents (N=3/21) said they did not find hearing aids beneficial, either because of tolerance issues: '*dreadful...he hates them...*' (F2), '*she wasn't tolerating them*' (F12), '*no...she wouldn't keep them in anyway (laughs)....*' (F15) or simply because they did not appear to work for the particular child '*they clearly don't make any difference...*' (F3). The last family added (about Audiology professionals who were clearly pushing for the amplification) '*but again they weren't listening to parent you know*'.

Even if hearing aids did not work for their child, a few 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).

3.5.3 Cochlear implants

Quite similar controversies surround cochlear implantation. Again, based on the clinical experiences with adults with ANSD who predominantly suffer from a neurodegenerative disease, there was a lot of hesitation expressed by professionals about whether cochlear implants even worked for ANSD. Parents picked 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)

In reality, latest research has shown that the success of cochlear implantation may be linked to the site of lesion (McMahon *et al.*, 2008), whereby in cases of pre-synaptic lesions cochlear implantation is likely to produce better outcomes.

Parents were understandably very aware of the potential harm of cochlear implantation, particularly considering the uncertainty surrounding ANSD in infants:

I still...we still think about it sometimes but...it's like...if we took his coch- if we did that now and then he couldn't hear anything which it could be and it's a big operation...and then he couldn't hear anything he's lost what he's got and at least he's got a bit of something now...so we could be better (F21)

Hence, the decision to opt for cochlear implantation was extremely hard for the family as there really was no knowing whether it would work or not. One family described the common situation these families tend to find themselves in:

they actually didn't know if it was going to make a difference or not and they said that to us didn't they when we went we don't know if it'll make she's...he said (child's name) would be an ideal candidate if she was profound- if ...she was a normal profoundly deaf child..(...)...but they added auditory neuropathy into it and that's when that's why we had to make the decision we had to... (F6)

Cochlear implantation candidacy was even more complicated than that of the above-described hearing aid fitting. Whilst some parents felt frustrated by the pressure from the professionals to choose the cochlear implantation route:

and for the ENT consultant perhaps to have appreciated that we quite clearly didn't want to go down the cochlear implant route and not at every bloody appointment go ... so do you want that referral yet... like she just wants us out the door especially as we've sat and waited an hour and a half you know in a waiting room with a screaming baby who's hungry you know just for a.... (F2)

Other families had to fight for their choice of cochlear implantation. Outdated and inappropriate candidacy criteria that took account the pure-tone audiogram instead of functional hearing tests such as

speech tests were applied. Paradoxically, a few families told about their relief when they found out that their child's hearing had deteriorated and finally met the criteria acceptable for cochlear implantation candidacy:

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)

But this discrepancy between pure-tone audiogram and perceptual abilities was not just problematic when it came to cochlear implantation candidacy criteria. Not surprisingly, some parents themselves found it impossible to contemplate cochlear implantation when their child seemed to hear, albeit not to perceive speech:

... 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)

Some parents who were convinced that cochlear implantation was the best choice for their child, found themselves having to fight for it:

and I really had to fight and I can remember crying and saying you're just not listening to me you know I've done my research I know what I'm talking about...you know I wouldn't put her through it if I didn't think that she'd benefit...and I felt like I had to fight them in the end and... (F14)

Sadly, a few parents also told about the unnecessary pressure they had to endure from different schools of thought. On the one hand, one family talked about the pressure and false information against cochlear implantation:

I think you know right from the very beginning we went to the deaf club....and got and got the absolutely clear message that cochlear implants were not....were not an appropriate choice..... they're of the devil yeah....(F3)

On the other hand, a couple of families also told about their frustration because of the pressure to stop signing once implanted:

yeah I mean and then I thought well actually you know they can't take his implant off him now then can they can't say well I'm sorry we saw you signing yesterday (laughs)...erm... (F10)

In our sample, 5 out of 21 had received a cochlear implant⁶ for their child and all expressed that they felt it had been a good decision. Their descriptions of the benefits varied greatly which is very common in cochlear implantation in this particular population. It illustrates truthfully the range of outcomes in ANSD:

it's absolutely made such a gigantic difference.. (F3)

...picks up her speech we didn't know we still...re- I guess really don't know at this stage...it looks like I think it's improving it feels like it's going the right way...(F6)

no she's doing really language work with them er so er when (child's name) got implanted er it was like the basic things attention to sounds you know difference in sounds and...is there a sound there is no sound and then different pictures and stuff and now we are working on...er actually getting some more vocabulary some more language some words and him and babble work and things like that...(F9)

... and then you're brought in and it was very much erm...almost well you realise that you won't you'll have to de-sign him when you stop with when your child has the implants so he underst- and you....ok so you've changed tracks from last week then and... the teacher of the deaf was in the room who didn't ...say hadn't told me about that before it was just like in that room and suddenly...it was very much as if and I felt as if...I that was what it was they were indicating that's why he hadn't developed so much speech up to that point and I know it's complete absolute nonsense and the only reason his vocab is so good is because we had done signing as well but....(F10)

her audiogram has changed so much (...) ... erm whether it's because you know her auditory nerve is being stimulated consistently because of the implant I don't know I mean I have read a few parents on the forum have said about that that they've noticed...where their children did have a little bit of residual hearing beforehand...they've said that that has improved since they've had the cochlear implant and they notice more you know like when they've got the times when they've not got the implant on say swimming and baths...that they're perhaps responding that little bit better...and that's why I'd sort of said to them at (place) you know just for the fun of it can we just see if her audiogram has changed...erm.. .and we went last (month) and it was the same...but then when we've gone again this year...there's been a massive difference.... so why that is I don't know ...whether she's just better at testing....you just it's just one of those unknowns (F14)

In Summary

Choosing an acceptable and appropriate route to auditory management and/or language development in neonatal ANSD was a challenge. Parents discussed their concerns, misconceptions, their efforts to make sense of controversial expert opinions, their realization that there is no size that fits all. Perhaps most importantly, parents articulated their need to be heard and appreciated that they were the experts in their own right when it came to their child.

⁶ At the time of interviewing, no children were bilaterally implanted.

4. Implications and recommendations

ANSD in very young infants is a relatively newly described condition and particularly in the context of newborn hearing screening poses clinicians with immense clinical challenges as well as ethical dilemmas.

Whilst not attempting to disregard the potentially considerable rewards associated with early identification of ANSD, it is essential to acknowledge that screening for ANSD does not currently meet many of the criteria outlined in the seminal work of Wilson and Junger (1968) that forms the foundation of the screening philosophy in the United Kingdom:

- (1) the condition sought should be an important health problem in the society concerned;*
- (2) there should be an accepted and effective treatment for the cases identified;*
- (3) facilities for assessment and treatment should be available;*
- (4) there should be a recognisable latent or early symptomatic stage;*
- (5) there should exist a simple predictive test or examination suitable for screening;*
- (6) the test should be acceptable to the population;*
- (7) the natural history of the condition should be understood;*
- (8) there should be an agreed policy on whom to treat as patients;*
- (9) the cost of case-finding (including further assessment and treatment of patients confirmed to have the target condition) should be non-wastefully balanced in relation to possible expenditure on medical care as a whole;*
- (10) case-finding should be a continuing process and not a 'once and for all' project.*

In the light of these criteria, having a young baby identified with ANSD as a result of routine screening for another condition (deafness) can be regarded as problematic from a professional perspective. From a parent's perspective it is also particularly challenging because they have to cope with a considerable amount of uncertainty with regard to their child's diagnosis, prognosis, support, intervention, management and choice of communication. Indeed professionals are often just as perplexed by the condition as the parents themselves.

Whilst diagnosis and clinical management in ANSD have attracted considerable research interest to date, the complex emotional and indeed ethical issues surrounding the condition in this particular population have hardly received any attention at all. Parents' voices have been largely silent. This is

the first study that has explored with parents' their unique and diverse experiences of having a child identified with ANSD through Newborn Hearing Screening Programme. Through those experiences, professional, ethical and clinical dilemmas become framed in a different way and new issues and complexities become identified.

The original premise of this study was to explore whether screening for a condition that we do not completely understand and do not really know what to do with is seen as a beneficial or indeed, harmful thing by families. This study provides evidence that in spite of the struggles and challenges that parents experience, in general, the families see early identification of ANSD as a positive thing. Having said that, one family in our sample felt that screening for ANSD was unjustified and detrimental.

Firstly, to better appreciate the experiences of parents whose babies have been identified with ANSD through newborn hearing screening, it is essential to realise the wider context in which these families find themselves. In England all children identified with ANSD have been in Special Care Baby Unit 48 hours or more. Granted, 41% of children identified with 'typical' sensorineural hearing loss also come from the Special Care Baby Unit population (Uus & Bamford, 2006) and thereby it is somewhat speculative to suggest there is anything different in the early parental experiences in ANSD and 'typical' sensorineural hearing loss. However there is evidence to imply that babies identified with 'typical' sensorineural hearing loss through the Special Care Baby Unit protocol are born on average at a higher gestational age and higher birth weight, have lower bilirubin levels, and are administered smaller doses of gentamycin (e.g. Dowley *et al.*, 2009). So it is perhaps justified to suggest that parents of ANSD babies do in general experience a more traumatic perinatal period and more anxiety and worry about the child's survival and long-term health and developmental sequelae than parents of children with 'typical' sensorineural hearing loss identified through the Special Care Baby Unit protocol. This study, through engaging parents in the telling of those experiences, provided greater detail about what those differences might be and their implications.

Parents participating in the present study had indeed gone through an excruciatingly difficult time because of the very serious medical worries and consequently did not in general prioritise hearing very highly at the point of diagnosis. ANSD was not seen as a separate diagnosis rather as a part of a much bigger medically complicated picture. This kind of contextualisation by parents challenges all professionals who work with children with ANSD to look beyond and around that condition and not just to see it in relation to other aspects of the child's health/development but to communicate its relationship with that wider context. Parents in this study were trying to understand their whole child's needs of which the consequences of ANSD were often not the most immediate issue. They were making choices about their child's and their own support needs within this bigger understanding of their child's condition, which ANSD fitted into, rather than totally defined.

An important theme in the parents' stories was centred on information and lack thereof. Parents sought information and typically struggled with getting it or getting it in a way that was meaningful to them. Parents in general were rather understanding of the reasons for lack of knowledge, expertise and awareness about ANSD amongst professionals because they quickly became aware that ANSD in infants was a fairly recently described, low-prevalence condition with a wide range of clinical manifestations.

They did however insist on honesty about what the professionals did or did not know. They wanted to get information about ANSD delivered to them sensitively and competently in an accessible language and format.

Knowing how parents understand and explain ANSD will help us to generate better explanations at the point of diagnosis and beyond to address parents' particular informational needs. It will also preempt and suggest how to respond to the many misunderstandings which can arise from analogies of deafness. In general, parents preferred imaginative explanations that illustrated the main manifestation of the condition, auditory temporal processing difficulty, in a way that they could really relate to in their every day lives, such as a broken speaker for example. For parents of children with ANSD, explanations were also important because they were so often put in the position of having to account for the seeming contradictions in their children's auditory behaviour to others.

When it comes to explanations and making sense of ANSD, a difficult conundrum, quite different from that of the challenges seen in 'typical' sensorineural hearing loss, is meaningfully introducing a spectrum approach when talking about the possible future outcomes in ANSD in infants and children. With the present knowledge, it is difficult, if not impossible, to predict where the outcome will lie in a meeting point of the following three dimensions: 1) electrophysiologically ABR may undergo full recovery, partial recovery or no change at all; 2) once the child is developmentally ready to perform the behavioural assessment (usually not before 6 months corrected age) the pure tone threshold will show anything from normal hearing sensitivity to profound hearing loss; 3) once it is possible to assess speech perception, it will range from no difficulties to difficulties only in noise to disrupted speech perception in quiet, the latter ranging from fairly mild to severe degree. These three dimensions will be relatively independent of one another. Furthermore, all these dimensions may go through daily/weekly/monthly fluctuations. Not overemphasising, yet incorporating all the possible scenarios to give the family an accurate view of this spectrum disorder without distorting the expectations in either direction, should be part of support and counselling. Parents in the study often felt that information given to them was either overly optimistic or, conversely, too pessimistic, yet the majority clearly demonstrated how adept they have become at living with uncertainty, unknowability and unpredictability. Perhaps a key lesson from this research is for professionals to believe that parents can be supported to do this, rather than to assume comfort and hope would arise from being definitive in their communications with them.

Hence, in infants with ANSD, with the present knowledge, the 'wait and see' approach is clinically justified, yet it can be potentially difficult for the families to endure. Explaining why this approach is warranted; providing opportunities to discuss it when more is known of the different 'dimensions' of the child's auditory function; making a management plan with clear development-appropriate milestones in mind and putting in place support for the family through the 'wait and see' period could possibly alleviate the frustration.

Nonetheless, despite such uncertainty of prognosis parents are required or at least they feel they are required to make decisions about communication, support, intervention and management, which in case of neonatal ANSD, is a challenge. Discussing parents' concerns, clarifying misconceptions and paying real attention to their observations are not miraculously going to exonerate the difficulties, but

it is nevertheless fundamental to better and more rewarding collaboration between families and the audiology team, and, hopefully, ultimately better outcomes for the child and the family in whatever form they will be measured. As parents in this study demonstrated, be it from differing starting points, there were real benefits to be gained in trying out different kinds of communication support and in exploring differing approaches to auditory management. Although for some there was a clear need to feel a decision had been reached for others certainty of management felt like a temptation. To fix on sign language, to decide for a cochlear implant, to be sure that hearing aids would suit best, could turn uncertainty of prognosis into a manageable course of predictable steps to development. However, the cost of that certainty could be to pre-empt the natural course of changes in their child's ANSD. For some, certainty was worth the risk, for others, living with uncertainty was worth the risk.

In conclusion

This is the first ever study focusing on the experiences of the parents whose babies have been identified with ANSD. It is evident from the data that not only do the manifestations of ANSD vary enormously; every child and every family is different as well. Yet the authors feel that there are significant commonalities in parents' experiences that are indeed generalisable.

Having said that, it is important to bear in mind that since some of the children in the study were identified with ANSD, there has been, and continues to be, significant progress made in the area of ANSD in infants. There is intensive research into finding better diagnostic tools in the attempt to reduce the uncertainty and cut down the 'wait and see' period. Indeed, techniques such as cortical auditory evoked potentials show a lot of promise in doing just that. Also, it is encouraging to know that there is ongoing research into the epidemiology, aetiopathogenesis and, very importantly, support and intervention of ANSD that will hopefully fill the gap, or at least some of the most vital gaps, in the knowledge in the near future. Consensus statements and guidelines have been produced and disseminated amongst clinicians, considerably diminishing the confusion in the profession.

Last but by no means least, largely due to the above-mentioned developments, the information and support available to families is gradually improving as well. Hopefully the accompanying document produced by the authors in collaboration with NDCS, *'Auditory Neuropathy Spectrum Disorder: To Parents from Parents'*, will furthermore contribute to addressing some of the common queries that families have.

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Appendix

Table 1. List of codes generated by team members.

Code	Inclusion criteria
AN in wider health context	To include references to ANSD within the wider health context of the child, including its relative significance
Hearing aids	To include all references to hearing aids
Cochlear implants	To include all references to cochlear implants
Sign language	To include all references to sign language
Outcomes and development	To include all references to the outcomes and development of the children including achievements and comparisons
How parents explain AN?	To include all examples of parents explaining ANSD including its relation to deafness and the formation of public narratives
Advice/improvements	To include all advice/suggestions made by parents for how they would improve services and parental experience including that to professionals and other parents
Positive support/ information	A sweeper category to make sure everything positive is collected. To include all references to positive aspects of parental experience
Newborn hearing screening and identification	To include all references to hearing screening and the identification of deafness
Pathways	To include all references to the pathway of service for ANSD children in terms of mapping what has actually happened and also references to process/stages/ANSD as a journey
Ignorant professionals	To include references to professionals who have negatively affected parental experiences due to ignorance of ANSD/ child/ parental experience
Certainty/ uncertainty	To include all references to certainty/uncertainty including professional and parental uncertainty. This will include references to trust and risk
Information and support	To include all references to information and support; what has been given/used and how it has been used (user strategies/ preferences)
Time	To include all references made to time to include speed and the passing of time
What kind of parent do you have to be?	To include all references to the strategies and personality types associated with managing their child's ANSD e.g. being a pushy parent, being proactive
Choices and decision-making	To include all references to what choices are made, how they are made and the emotional effects of making these decisions for their children
Child identity	To include all references to the child as normal/deaf/hearing.

Table 2. Details of the interviewee and their child identified with ANSD.

	Interviewee	Highest educational qualification of interviewee	Child's age at interview	Child's siblings
1	Mother	Postgraduate	24wks	twin
2	Mother	Postgraduate	19 mo	no siblings
3	Both parents	NVQ level	6 yrs	older siblings
4	Mother	Degree	2 yrs	twin
5	Mother	A level	4 yrs	no siblings
6	Both parents	A level	4 yrs	no siblings
7	Mother	GCSE	4 mo	no siblings
8	Mother	NVQ level	2 yrs	no siblings
9	Mother	Postgraduate	3 yrs	triplets
10	Mother	A level	4 yrs	older sibling
11	Mother	Degree	3 yrs	no siblings
12	Mother	Degree	4 yrs	?
13	Mother	Postgraduate	7 mo	twin
14	Mother	A level	6yrs	sibling
15	Mother	NVQ level	3 yrs	younger sibling
16	Both parents	Postgraduate	13 mos	twin
17	Both parents	Postgraduate	11 mos	twin
18	Father	?	6 yrs	older sibling
19	Mother	A level	18 mo	twin
20	Mother	NVQ level	4 yrs	no siblings
21	Mother	NVQ level	3 yrs	older sibling

Table 3. Medical history as declared by the families during the interviews.

	Gestation weeks	Birth weight	Perinatal medical conditions	Persisting medical conditions	Multiple pregnancy	Delivery
1	34	1.7	RD, anaemia, VCC	No	twin	EC
2	36	3.6	HP, RP, RD, seizures, intracranial haemorrhage	global DD, athetoid CP		EC
3	37	3.9	RD, anaemia, HP	No		
4	24	0.8	RD, hernia	No	twin*	
5	34	2.8	RD, RP	No		
6	26	0.7	GI, cephalopathy, RD	NP, syndrome?		
7	37	3.2	RD, anaemia, hyperbil hypoxic-ischaemic	global DD, syndrome? global DD, CP		
8	34	1.4	encephalopathy	(spastic hemiplegia)		EC
9	26	1.2	RD	CP (spastic hemiplegia), RP	triplets*	
10	32	2	inguinal hernia, seizures	CP?		
11	27	0.8	RD, dysphagia	dysphagia, CP?		
12	25	0.8	RD, sepsis, hyperbil	global DD, RP, nystagmus		
13	25	0.9	RD, VCC, brain haemorrhage, hyperbil	CP	twin	
14	24	1	RD, hyperbil	no		EC
15	30	0.9	RD, sepsis	Turner syndrome?		EC
16	31	1.6	NP, RD, hyperbil	No	twin	
17	29	1.5	RD, hyperbil, acute hydrocephalus	No	twin	
18	37	3.2	hyperbil	global DD, condition?		EC
19	30	1.5	sepsis, hyperbil, VCC	???	twin	
20	37	2.8	hyperbil	global DD (kernicterus?)		
21	29	1.5	RD	CP		
	30.7	1.8				

RD: respiratory distress;

HP: hepatopathy;

NP: nephropathy;

CP: cerebral palsy;

VCC: congenital heart defect;

RP: retinopathy; hyperbil: clinically significant hyperbilirubinaemia;

DD: developmental delay;

EC: emergency caesarean section

Table 4. Distribution of communication/management options in our sample.

	HL	HA benefit	CI	Visual communication
1	???	No amplification	considering	Cued speech, some BSL
2	Moderate	No benefit, tolerance problems	considering	BSL
3	Profound	No benefit	YES	BSL
4	Moderate	Very beneficial	No	No
5	Moderate-severe	Very beneficial	No	Makaton
6	Profound	Somewhat beneficial	YES	Total communication
7	Normal	No amplification	No	No
8	Normal	No amplification	No	Makaton
9	Severe	No benefit, tolerance problems	YES	BSL
10	Severe	Somewhat beneficial	YES	BSL
11	Mild	Very beneficial	No	Makaton, BSL
12	Mild	No benefit, tolerance problems	No	BSL
13	Moderate	Professional decision against HA	No	Baby sign
14	Profound	No benefit, tolerance problems	YES	Baby sign, lipreading
15	???	No benefit, tolerance problems	No	BSL
16	Profound	Very beneficial	No	Some BSL
17	Moderate-severe	No benefit, tolerance problems	considering	No
18	Normal	No amplification	No	No
19	Moderate	Very beneficial	No	No
20	???	Somewhat beneficial	referred for	Some BSL
21	Moderate	Not sure about the benefit	No	Some BSL

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Published by the National Deaf Children's Society © NDCS July 2011
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NDCS is a registered charity in England and Wales no. 1016532 and
in Scotland no. SC040779.
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