Auditory Neuropathy Spectrum Disorder

To Parents from Parents

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

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NDCS uses the word ‘deaf’ to refer to all levels of hearing loss.
Introduction

This booklet was written following a study carried out by the University of Manchester. In the study we talked to 21 families who had a child identified with auditory neuropathy spectrum disorder through the Newborn Hearing Screening Programme. The families had used a range of options for their children, mainly including hearing aids, cochlear implants and sign language. Although parents and children experienced a range of outcomes, there were many common themes. This booklet describes what the families told us about their experiences and shares the useful advice which these parents would give to families who are in a similar position in the future.

It may be helpful to read the NDCS booklet *Auditory Neuropathy Spectrum Disorder: Information for families* alongside this booklet.
ANSD (auditory neuropathy spectrum disorder) is a term used to label a particular type of hearing problem. It is characterised by hearing test results that include absent or abnormal auditory brainstem responses (ABR), with cochlear microphonic (CM) and otoacoustic emissions (OAE) present.

For more information on different types of hearing tests read the NDCS booklet *Understanding your Child’s Hearing Tests*.

Sensori-neural deafness typically happens because the outer hair cells inside the cochlea (inner ear) are missing or damaged and are not working as well as they should. The cochlea looks like a snail shell and usually has two and a half turns in its spiral.

**Diagram of the ear**
The diagrams below show a cross section of the cochlea that you would see if the spiral could be pulled out into a long tube and sliced through. There are many thousands of hair cells throughout the length of the cochlea. In children with ANSD the outer hair cells are likely to work well and this is demonstrated by the presence of otoacoustic emissions. In ANSD the problem is elsewhere in the auditory system. Part of the auditory system that might be affected (sometimes referred to as ‘potential sites of lesion’) include the inner hair cells, the junction (synapse) between the inner hair cells and the afferent nerve fibres, auditory neurons in the spiral ganglion (all also within the cochlea), or the auditory nerve or brainstem auditory nuclei (part of the nerve pathway between the ear and the brain). It can also be a combination of areas affected in the auditory pathway.

Cross section of the cochlea showing the parts affected by typical sensorineural deafness

- Inner hair cell
- Junction (synapse)
- Outer hair cells
- Spiral ganglion
- Auditory (hearing) nerve
- Auditory nerve fibres
Cross section of the cochlea showing the parts affected by ANSD

- Inner hair cell
- Junction (synapse)
- Outer hair cells
- Spiral ganglion
- Auditory (hearing) nerve
- Auditory nerve fibres
How common is ANSD?

One in ten children diagnosed with deafness at birth have ANSD.

This means about 1 in 10,000 babies born each year have ANSD.
What do the hearing test results mean for the child?

Usually the auditory brainstem response (ABR) will closely match the levels at which a child responds to sound. However, when a child has ANSD, the ABR will not match the level at which the child responds to sounds. Very often, there will be no auditory brainstem response even though the child does respond to some sounds. When diagnosed with ANSD soon after birth, the information from hearing test results is more limited than for other types of deafness. The results only tell us that the child has normal OAEs and abnormal or absent ABR. More detailed information about how much a child can hear and make sense of is missing. This differs from children diagnosed with ‘typical’ deafness. So even the best expert knowledge in the world is not able to predict what the outcomes will be when the child is older. By outcomes we mean how much the child will hear, whether their hearing will change over time and how they will communicate in the future.
ANSD is a ‘spectrum disorder’ so that test results and functional outcomes are very variable between children with the condition.

- The ABR result may not change at all, or it may recover partially or fully.

- When the child is developmentally ready to perform behavioural tests of hearing (usually from about 6–9 months of age) their hearing threshold (the quietest sound they respond to) may be anywhere from normal hearing levels to profound deafness.

- When it is possible to assess their speech perception (how much the child is able to make sense of speech), it can range from no difficulties, experiencing difficulties hearing speech clearly in noisy conditions, or anywhere between mild to severe difficulties in quiet listening conditions.

Each of these three dimensions (ABR, hearing thresholds and speech perception) is relatively independent of one another. Just because one is affected in one way does not mean the others will be affected in similar ways. Also all of them can fluctuate on a daily/weekly/monthly basis.
Definitions from parents

Factual descriptions of ANSD similar to those on previous pages were ones that the parents we spoke to had routinely encountered and were learning to make sense of slowly. But they had also developed different ways of describing ANSD themselves. They found it easier not to worry so much about where the ‘problem’ was in the auditory system, but what and how the child was likely to be hearing. Parents valued those descriptions that likened ANSD to something with which everyone is familiar. Many compared ANSD to ‘faulty wiring’ and ‘radio static’. These explanations were useful because they described what the child might be hearing, their actual experience, rather than what the problem might be.

“imagine a speaker. Everybody’s had a broken speaker at some stage where the wire doesn’t quite connect properly... that’s the one I use and everyone says ‘I get what you mean’”

“and then she explained to me about the fact that... everything seems out of synch... a bit like when you’re playing a song and all the parts of the song are all jumbled up and...to me that was the clearest... way of explaining... something like that”
Family experiences

ANSD is a spectrum disorder, meaning children with ANSD may have a range of very different outcomes. These summaries come from the experiences of three different families who took part in our research. A few details have been changed to preserve confidentiality.

The Jones family

The Jones family’s son, Sam, was born six weeks early and spent three weeks in the Special Care Baby Unit. He was identified with what looked like profound deafness very soon after birth and then one month later with probable ANSD. At four months old the family were offered hearing aids but they turned them down, scared they would do more harm than good as it was so unclear what exactly Sam could hear. At this point they felt like they had enough to deal with and that the professionals did not seem sure about it. When he was not responding to sound and speech at one year old they decided to go for the hearing aids. It took a few months for him to get used to the hearing aids and to build up the time he kept them in, but they are now working very well. Occasionally he has ‘hearing days’ when he chooses not to wear them because it seems he hears fairly well without them. But his hearing levels do not fluctuate as much as they used to. The family chose to use Makaton with their son before he was aided, preferring not to learn British Sign Language (BSL) as he might not end up needing it. They also worried that signing would cut their son off from ‘speaking society’. He is now four years old and his speech is improving. The family has never considered a cochlear implant for Sam.
The Blake family's daughter, Tara, was born six weeks early and spent six weeks in the Special Care Baby Unit. She struggled to put on weight and was very small when she left hospital. Her parents were told that she had a profound deafness and she had ear moulds taken for hearing aids when she was just six weeks old. In the end her parents decided not to have them because they wanted to give their daughter some time to grow and just be a child. This decision was against medical advice and the family found this period very stressful. Over time Tara started to babble and the hearing test results started to show that she had more hearing. Now Tara is two years old and is speaking normally for her age. There is no suggestion that she will need hearing aids or a cochlear implant in the future.
The Khan family’s son, Ali, was born two months prematurely and spent two weeks in the Special Care Baby Unit. He needed an operation for a hernia not long after he left hospital and some of his appointments with audiology were delayed because his operation was more of a priority. His hearing levels have not changed very much since he was first diagnosed and his hearing responses seem to match his behavioural test results. Ali’s parents were offered hearing aids as something to try after his behavioural tests seemed to confirm the diagnosis. He was fitted with two hearing aids just before he was one. They had a very hard time with them for the first few months because Ali wouldn’t keep them in. His mother was very keen for him to understand Deaf culture and both his parents learned British Sign Language (BSL). Later, they decided to undergo assessment for a cochlear implant. There was some delay as he needed grommets fitted because of glue ear and this made it difficult to accurately test his hearing. He received his cochlear implant just after he turned three years old. The family were told that the cochlear implant would only make a small difference but it has worked really well for them. Ali is happy with his implant and is developing his speech. His mother was unhappy that going on the cochlear implant programme meant ‘de-signing’ him, as she put it, and is keen that BSL should be a second language for Ali and a part of his Deaf identity.
No parents in our research study were deaf themselves although we tried to recruit deaf families. It may be that deaf parents who already use British Sign Language at home have different experiences of having a child with ANSD. Concerns about how much a child with ANSD might ‘hear’ and how this might change are likely to have a different significance for culturally Deaf families. Further research is needed to help us understand their information needs.

Deaf written with a capital D usually refers to people who consider themselves to be members of the Deaf community and to be part of a cultural or linguistic minority. Most members of the Deaf community will use British Sign Language as their first or preferred language.
Common experiences

Although parents’ situations were very different, they described to us seven key experiences that were common to many of them. These include the issues which families found challenging and what has helped them in these situations. You may also experience some of these same issues, although each family will be different.
ANSD and other health problems
Many children who are diagnosed with ANSD will have been born following complicated pregnancies and will have had difficult births. Some parents will have had times when they feared losing their child. Many parents told us that, in the beginning, concern about their child’s hearing was simply not their highest priority if their child had serious health problems. Your child may or may not have other health problems apart from ANSD. This means that some parents may have many professionals to see and a lot of appointments to go to which can be overwhelming, especially when the child is very young.
Parents’ advice to other parents:
“This feeling of being overwhelmed will get less with time.”

“It is helpful to think about whether you need to prioritise certain appointments at certain times, rather than trying to do everything all the time.”

“You may need to concentrate on health problems that are more urgent, or you may need time to come to terms with the things you and your child have gone through. Don’t worry if the ANSD comes lower on your list of priorities than other things in the early days. Your child’s development will not suffer if you put off thinking about it for a little while. But, if possible, do keep the audiology services informed of your decision and together come up with a plan that works for your child and your family.”

“Try not to postpone it for too long though. The potential effects of ANSD on hearing and language are important, so it is vital to make sure your child is being monitored.”
Information
Parents emphasised that it could be hard to find the right kind of information about ANSD. Although there is a lot of information available on the internet for example, it can be hard to know what to trust. Not all information is written with parents in mind. Not all is relevant or helpful for ANSD in very young babies and infants. Not all is factually correct.

When looking up information, some parents realised that there were a great many differences in expert opinion about ANSD and this could be disconcerting and confusing.
Parents’ advice to other parents:

“Make sure the way information is given to you matches your needs. For example, some parents want to read everything they can about ANSD. Others might prefer to talk to families in the same situation. Some want to talk with a professional about their questions rather than having to read lots of information. Tell your professional how you would prefer to access this information.”

“Let people know when you would prefer to receive and use information. You might be happy to have all the information given to you in the beginning or you might find this too much and put it away until it seems more relevant.”

“Not everyone wants the same amount of information at the same time. Let people know what you want.”

“Information that is shared or presented in a way that you prefer, and information that comes from or is linked to a trusted person works best for parents.”

“Don’t be afraid to ask for different kinds of information. Some people might want suggestions about good medical research to read. Some might want to have contact with other families with a child with ANSD.”
Making sense of the seemingly contradictory diagnosis

Infants with ANSD may often seem to ‘hear’ and react to sounds. A diagnosis of a ‘hearing problem’ can therefore be extremely difficult to come to terms with. This is very common with ANSD. Differently from ‘typical’ deafness, the child may hear sounds but may not be able to understand speech: like somebody who is listening to a car radio that starts to go off station with lots of interference, and is unable to understand the words. One family described it as ‘dyslexia of hearing’. Not surprisingly, some parents find it very upsetting when the audiologists started talking about fitting hearing aids and cochlear implants before they had time to take in that there might be a problem.
Parents’ advice to other parents:

“Get written information from professionals so you don’t have to take everything in at the time.”

“Take written questions to appointments so you remember what you wanted to ask.”

“If you feel like professionals are rushing ahead let them know that you need time to think about what they have told you.”

“Try to get contact details so that you can ask questions later. There is no need to try to understand everything about the diagnosis or every possible outcome at these early appointments.”
**Wait and see**

You will be told that you need to wait and see and that the child needs monitoring. It may sound frustrating, but it is a very wise approach for ANSD for a number of reasons:

- a child with ANSD has abnormal or absent ABR. This means that your audiologist cannot use the hearing test to estimate the hearing level, the way it is usually done in ‘typical’ deafness

- even after the child is old enough to be tested behaviourally (usually around 6–9 months of age), the audiogram gives very little information about how the child will receive and develop speech

- hearing status may go through significant changes. Not only will it change from day to day (many ANSD children have ‘good hearing days’ and ‘bad hearing days’), in some cases, hearing may recover. This is because ANSD is linked to maturational (developmental) delay in some children and, once their development catches up, their hearing may recover. If that is the case, it will usually happen within the first 18–24 months.

For these reasons, professionals might delay fitting hearing aids or set them cautiously at a lower level than they would if your child had ‘typical’ deafness. Professionals will not recommend cochlear implantation to your child before 18–24 months of age because of the possibility that hearing may recover. This can seem frustrating and it might make you wonder why they bothered to make the diagnosis if they cannot do anything about it. It is easy to think that professionals just do not know enough about ANSD, which is why they might appear hesitant. Really, it is because they cannot know how your child is going to develop.
Parents advice to other parents:

“Make sure professionals explain why they cannot do things that it might seem obvious to do if a child is deaf, for example fitting a hearing aid.”

“Even though your child’s development might be uncertain, it will help if you feel that there is a plan in place and that you know who will be working with you and your child and when they will be involved.”

“You might like to start looking into the options available for your child in the future even though not all children with ANSD will go on to need hearing aids or cochlear implants or use visual communication (such as British Sign Language, Makaton or Cued Speech).”

“You might also find it useful if you have suggestions for activities you could be doing to help your child’s communication and overall development in the meantime, or ways that you can monitor your child’s communication and development.”

“A lot of parents felt that ANSD became more of a priority around the age when children usually began to talk. As this time approaches, you may feel that you want something to be done if your child is not developing speech. Professionals should have a much better idea of your child’s hearing levels by this point. You may wish to try amplification through hearing aids once the hearing levels are established or you may wish to consider being referred to a cochlear implant programme. You may also decide you want your child to use sign language over the long term, or it may be best suited for your child to use a combination of these options.”
Changes in the child’s hearing tests and hearing function
It may seem very strange to you that children with ANSD can go from being what is thought to be ‘profoundly deaf’ to having ‘normal hearing’. Professionals will probably tell you that some children’s ANSD is caused by delayed maturation (development) of the nerve involved in hearing and that over the first 18 months to 2 years of a child’s life this could improve on its own. But professionals have no way of telling whether this will happen with your child or not. In the meantime, it can be hard for families to decide how to think about their child’s needs and identity. For some, they think of their child as deaf although their hearing may fluctuate. For others, their child is thought of as a child whose hearing is uncertain. Every family and every parent is different in what they find helpful to cope with the puzzling diagnosis of ANSD and the uncertainty that follows.
Parents’ experiences

• Some of the parents who took part in this study told us that it was helpful to know about the possibility that ANSD would not affect their child’s hearing later on. This possibility made it easier for them to cope with the present. They found it useful to look out for signs that their child was responding more to sound.

• Other parents had the opposite experience. They found this hope made it more difficult to cope. They would have preferred a definite prognosis. Some said they would have found it helpful to know that their child was ‘properly deaf’.

• Many parents told us that it was hard to accept that even if their child’s hearing seemed to be improving, there was no way to be sure about their final hearing status before 18 months to 2 years.

• Some parents who had become involved with professionals and support groups for deaf children worried about how relevant these would be if their child ‘became hearing’, but they still valued the support.

• Others found it difficult to fit into or use support for families with deaf children because their child was not ‘deaf’ or ‘hearing impaired’ in the usual way.
About professionals not knowing
Parents told us that they had often had experiences of professionals who had very little or no experience of ANSD. Some felt they were being treated as a ‘guinea pig’. This is because ANSD is rare and each child is so different, so even the most experienced ANSD expert in the world will treat each child with utmost care and caution. It is not unusual to feel professionals are being very cautious about treating your child. It can be stressful for parents if they do not have confidence in the people who are working with them.

Parents told us the following:
- they found it easier to cope if they had identified a professional who they trusted as a source of advice or just as a main point of contact.

- it was important that professionals were open to finding out about ANSD and were open with parents about what they knew and what they did not know. This openness built trust.

- having contact with professionals who believed in parents’ observations and trusted that parents knew their own children well was also important.
Making decisions
You might find it difficult to make decisions which will affect your child’s future when professionals cannot give any guarantees about what is going to work. You may also come across a lot of conflicting advice about the best approaches for children with ANSD and you might find yourself wanting to go against some of this advice. Some parents have very clear ideas about the route they want to take but many are happy to try things and if they don’t work go onto the next thing.

Parents told us the following:
• it was helpful for professionals to acknowledge that parents might change their minds and that there was not one right approach for a child with ANSD.

• it was important to feel that professionals discussed the various options in a fair and unbiased way.

• it is helpful to meet with other parents of children with ANSD to discuss why they made the choices that they did.

• it might be helpful to actually meet children with ANSD who use hearing aids or have a cochlear implant, or who use sign language, as well as those who manage well without any extra support to find out more about the range of outcomes for children with ANSD.
Details of the study

The full details and findings of the research are available to download from the NDCS website *Auditory Neuropathy Spectrum Disorder in Infants; Experiences of parents whose babies were identified with Auditory Neuropathy Spectrum Disorder through the Newborn Hearing Screening Programme* (April 2011) at [www.ndcs.org.uk/professional_support/external_research/](http://www.ndcs.org.uk/professional_support/external_research/)

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For further information on ANSD, please contact Dr Kai Uus at: kai.uus@manchester.ac.uk
NDCS provides the following services through our membership scheme. Registration is simple, fast and free to parents and carers of deaf children and professionals working with them. Contact the Freephone Helpline (see below) or register through www.ndcs.org.uk

- A Freephone Helpline 0808 800 8880 (voice and text) offering clear, balanced information on many issues relating to childhood deafness, including schooling and communication options.
- A range of publications for parents and professionals on areas such as audiology, parenting and financial support.
- A website at www.ndcs.org.uk with regularly updated information on all aspects of childhood deafness and access to all NDCS publications.
- A team of family officers who provide information and local support for families of deaf children across the UK.
- Specialist information, advice and support (including representation at hearings if needed) from one of our appeals advisers in relation to the following types of tribunal appeals: education (including disability discrimination, special educational needs (SEN) and, in Scotland, Additional Support for Learning (ASL)); and benefits.
- An audiologist and technology team to provide information about deafness and equipment that may help deaf children.
- Technology Test Drive – an equipment loan service that enables deaf children to try out equipment at home or school
- Family weekends and special events for families of deaf children.
- Sports, arts and outdoor activities for deaf children and young people.
- A quarterly magazine and regular email updates.
- An online forum for parents and carers to share their experiences, at www.ndcs.org.uk/parentplace.
- A website for deaf children and young people to get information, share their experiences and have fun www.buzz.org.uk
NDCS is the leading charity dedicated to creating a world without barriers for deaf children and young people.

NDCS Freephone Helpline: 0808 800 8880 (voice and text)

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www.ndcs.org.uk