

# Auditory Neuropathy Spectrum Disorder (ANSD)

Information for BC Families



## BC Early Hearing Program

A service of BC Children's Hospital  
and the Provincial Health Services Authority



This booklet was created by the BC Early Hearing Program to help provide you and your family with accurate information. Thank you to the parents who shared their personal experiences. We have included some of their quotes.

This booklet has information about:

- ▶ Your child's unique hearing status and how to help your child (see sections: [What is Auditory Neuropathy Spectrum Disorder \(ANSD\)?](#) [How will I communicate with my child?](#))
- ▶ Who provides help going through the process (see section: [Who will help me support my child's development?](#))
- ▶ The different types of Audiology tests that are used to identify this hearing loss (see section: [What tests identify ANSD?](#))
- ▶ How the ear works in children identified with ANSD (see section: [What is hearing loss?](#))

*“As new parents, this world is so new to us and we want to educate ourselves and absorb everything we can in order to better help our little bundle of joy.”*

*“As parents we need to be advocates for our children and help others understand how ANSD affects our child uniquely.”*

Share this booklet with your family and team of service providers who may also want to learn more about ANSD.

*Remember you are the expert  
on your own child!*

**BC Early Hearing Program**

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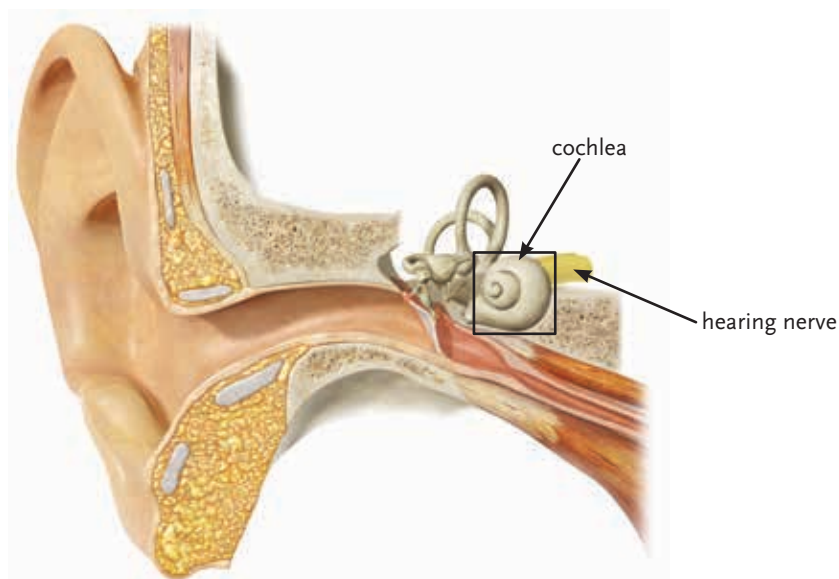
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## What is Auditory Neuropathy Spectrum Disorder (ANSD)?

Auditory neuropathy spectrum disorder (ANSD) is a rare type of sensorineural hearing loss where sound is not transmitted properly from the cochlea (inner ear) to the hearing nerve or from the hearing nerve to the brain.



It is important to know if your child has ANSD because this type of hearing loss is managed differently from the more common form of sensorineural hearing loss.

ANSD is called a *spectrum* disorder because its effects are unique for each child. Some children with ANSD will experience mild hearing difficulties that are only a problem in background noise. For most children with ANSD, however, the ANSD leads to significant hearing difficulties and challenges understanding speech.

*“As I have met other parents who also have children with ANSD it is very interesting to learn that we all have different stories. Sometimes there are common elements but every child seems unique.”*

Common features of ANSD include:

- ▶ **Hearing loss of some degree.** The hearing levels in children with ANSD vary from near-normal to profound deafness. Your child's hearing level is how loud a sound needs to be for your child to "detect" the sound or know that the sound is present. Hearing levels do not tell us anything about your child's ability to understand or make sense of the sound.
- ▶ **Speech understanding difficulties.** Speech understanding abilities for children with ANSD are highly variable and cannot be predicted by their hearing levels. Children with ANSD may have more difficulty understanding speech than would be expected based on their hearing levels. Even if soft sounds are heard, the sound information may be distorted or disorganized by the time it reaches the brain. For example, think of a radio that is slightly off-station; you can hear that someone is talking, but it is too fuzzy to make out all of the words clearly. Turning up the volume does not help; it makes the fuzzy talking louder, but not clearer.

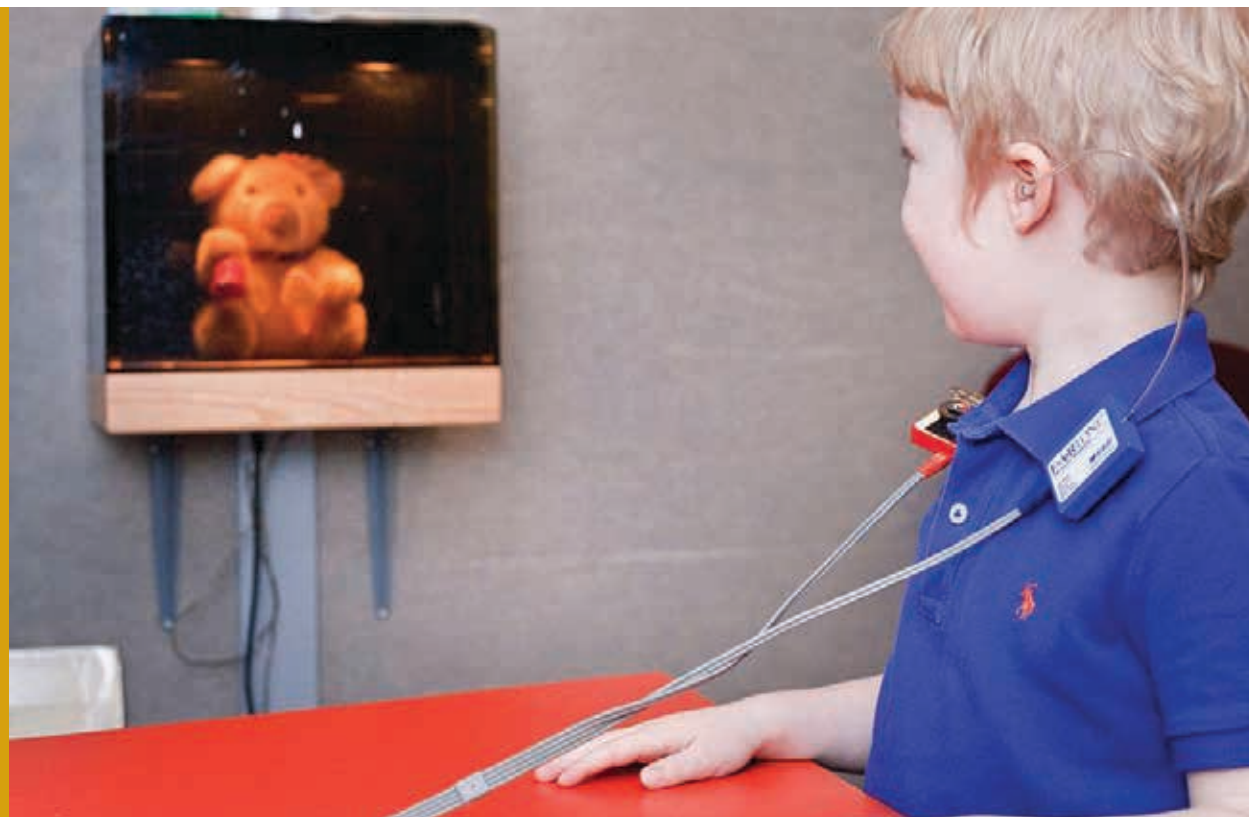


The early stages of hearing testing in infancy can clearly *identify* ANSD. However, a child's hearing levels cannot be predicted until they are old enough or developmentally able to participate in behavioural testing. This is different than the more common form of sensorineural hearing loss, where hearing levels are predicted from early testing in infancy.

Behavioural tests are usually attempted as early as 6-9 months, but for some children may not be possible until later. In these tests, behaviours in response to sounds help determine a child's hearing levels and speech understanding.

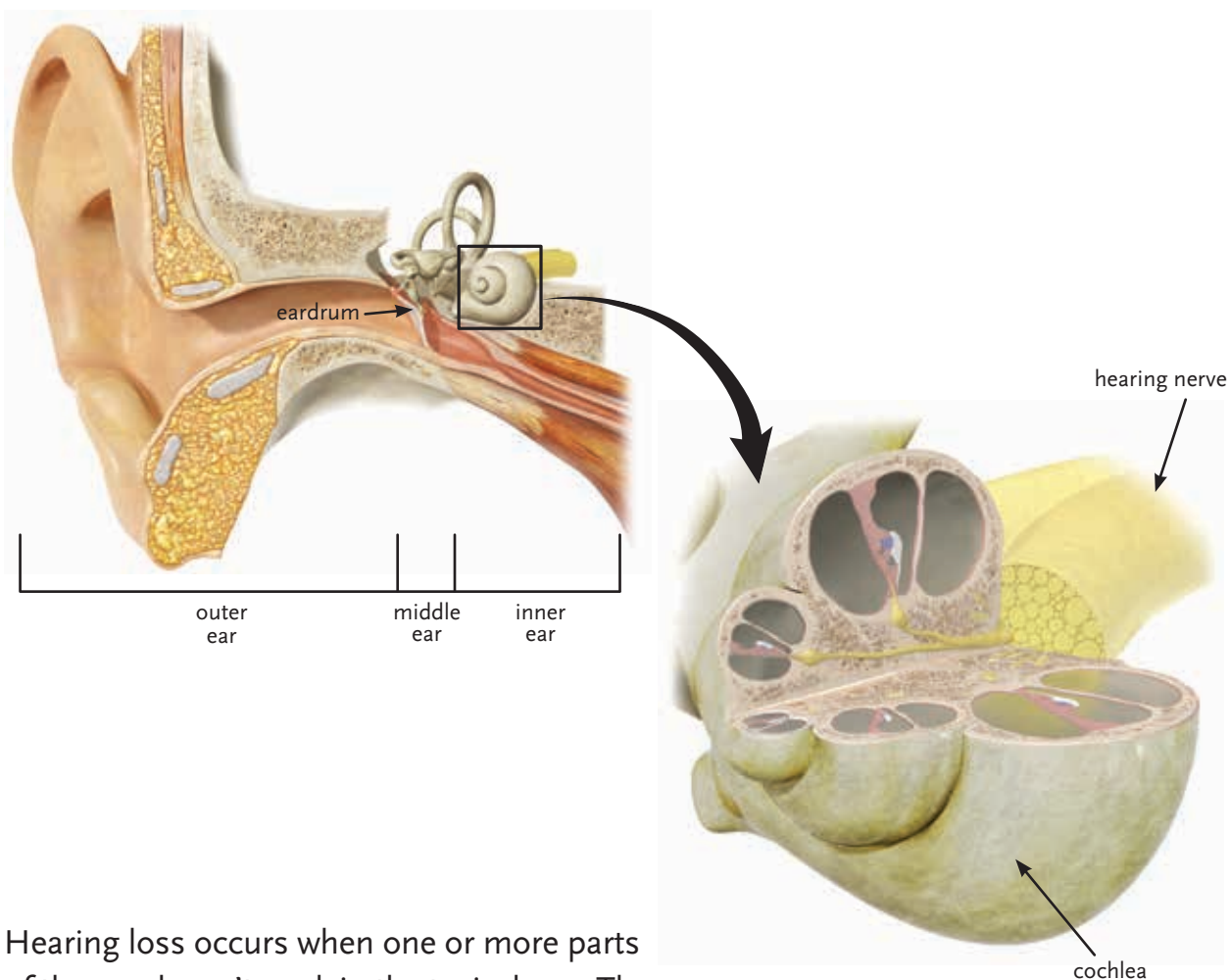
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***ANSD does not affect all children in the same way and its effects range from mild to severe. Each child with ANSD differs in how they understand sounds. One way ANSD is different from other types of hearing loss is that a child's hearing levels and speech understanding abilities cannot be predicted from infant hearing tests.***



## What is hearing loss?

The ear is made up of three main parts: the outer ear, the middle ear and the inner ear.



Hearing loss occurs when one or more parts of the ear doesn't work in the typical way. The part of the ear involved determines the type of hearing loss. Sensorineural hearing loss occurs when the inner ear or the hearing nerve, does not work properly. This type of hearing loss is considered permanent. We will focus on this type of hearing loss here.





*“My husband wanted to know all the little details about our son’s hearing, but I really just wanted the big picture. Different parents want different levels of information, I think, so it’s important to work with professionals who can meet you where you are at.”*

*“When my baby was identified with ANSD, I first needed to understand how the ear usually works and how we hear to really understand what was different for my baby.”*





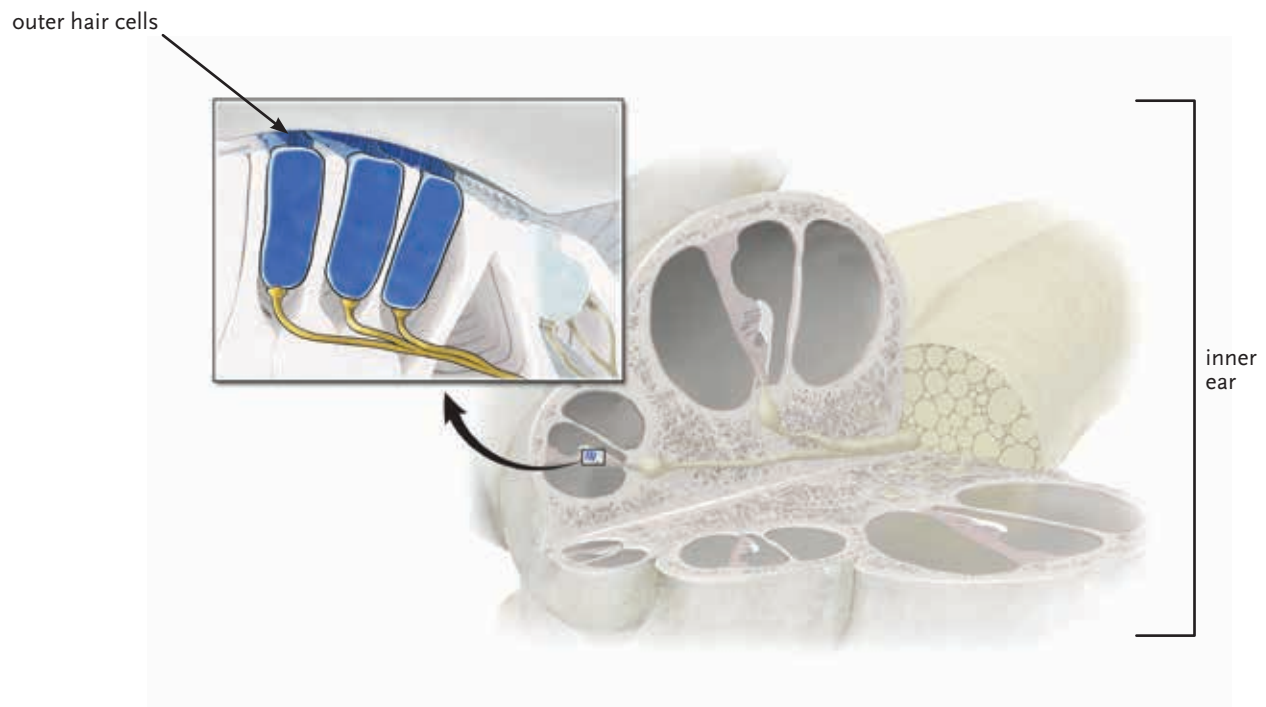
## Sensorineural Hearing Loss

The most common form of sensorineural hearing loss occurs when the outer hair cells in the cochlea are not working as they should.

The cochlea is located in the inner ear and contains thousands of sensory cells called outer hair cells and inner hair cells. These sensory cells have different roles. When functioning properly, the outer hair cells make soft sounds louder inside the cochlea and the inner hair cells change movement of the fluid in the cochlea into electrical signals. The electrical signals are then passed on to the hearing nerve, which sends this information to the brain. The brain interprets this as sound.

When outer hair cells are damaged or missing, sound is not transmitted properly to the hearing nerve. This results in hearing loss.

The diagram shows the cochlea and the outer hair cells, which are affected in the most common form of sensorineural hearing loss.



### Auditory Neuropathy Spectrum Disorder (ANSD)

A less common form of sensorineural hearing loss is ANSD. With ANSD, the outer hair cells in the cochlea may work well but a problem occurs after this point in the hearing pathway. The timing or pattern of sound transmission is disrupted somewhere along the path to the brain. As a result, sounds are not transmitted to the brain as they should be.

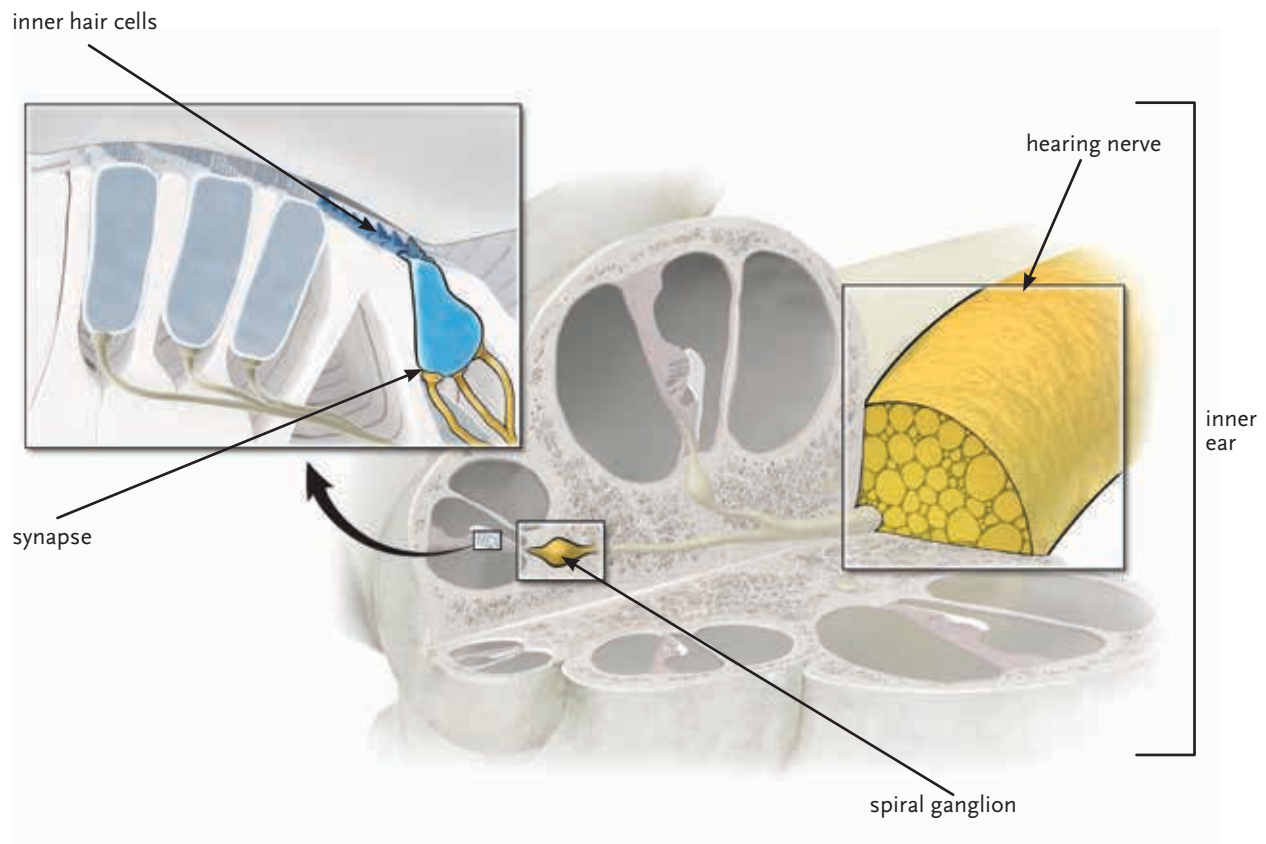
The specific locations that can cause ANSD likely differ from person to person and cannot always be determined from clinical tests. Parts of the hearing pathway that might be affected include the inner hair cells in the cochlea, the connection between the inner hair cells and the hearing nerve (synapse), the hearing nerve itself or the brainstem (part of the nerve pathway between the ear and the brain). There can also be a combination of areas affected.

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*The most common form of sensorineural hearing loss occurs when the outer hair cells in the cochlea are not working as they should. With ANSD, there is a problem with how sounds are transmitted after the outer hair cells.*



The diagram below shows the cochlea and the parts of the hearing pathway that may contribute to ANSD.



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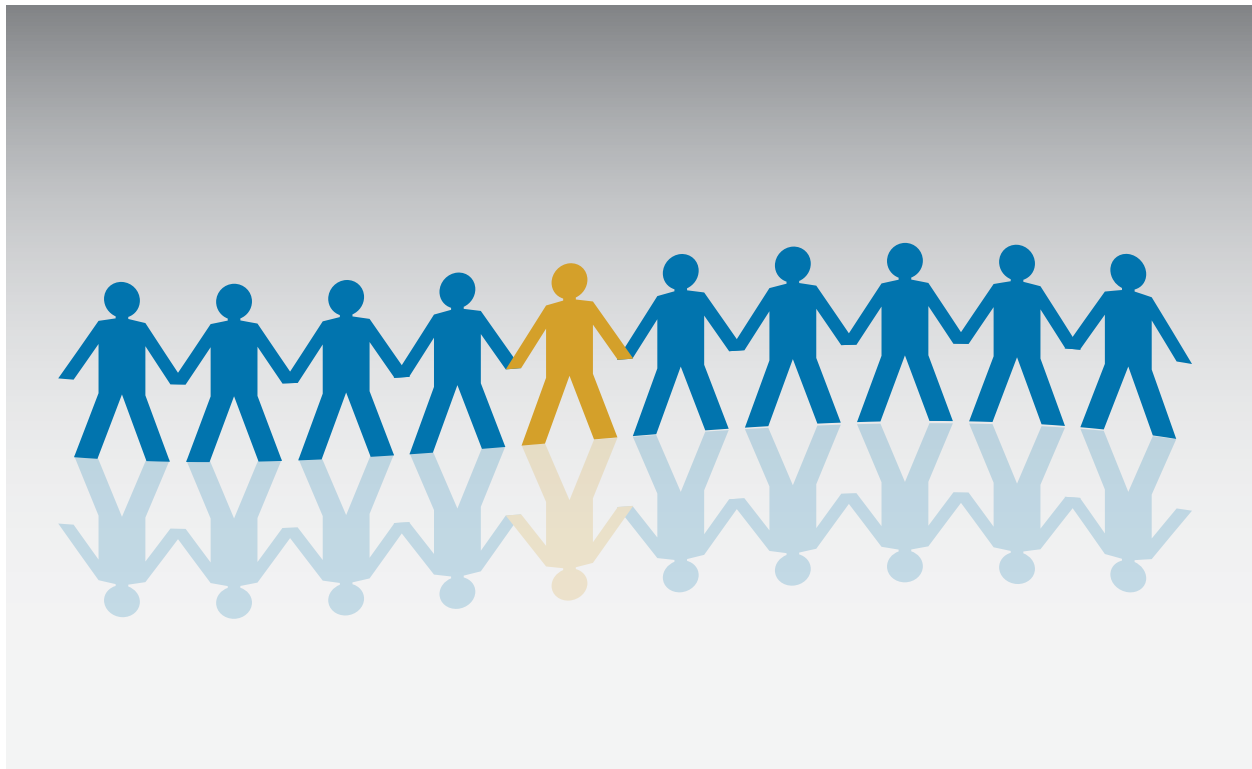
***Auditory Neuropathy Spectrum Disorder (ANSD) is different from other more common types of hearing loss because sound information reaching the brain is not always transmitted in a way the brain can understand. This affects the way in which a child is able to understand sounds, including speech.***





## How common is Auditory Neuropathy Spectrum Disorder?

Two out of 1000 babies are born with sensorineural hearing loss each year. ANSD is a form of sensorineural hearing loss. **For every 10 children with sensorineural hearing loss, approximately 1 will have ANSD.**



*In BC, approximately 44,000 babies are born each year. Of these, 100 will be deaf or hard of hearing; 10 of them will have ANSD.*

## What causes Auditory Neuropathy Spectrum Disorder?

The reasons why children have ANSD are often not known. We do know that ANSD is more common among children with complex additional needs or babies who have a history of serious health problems during or shortly after their birth. Many of these children spent time in the newborn intensive care unit (NICU), and may have had other conditions when they were born such as:

- ▶ prematurity
- ▶ very low birth weight
- ▶ lack of oxygen at birth
- ▶ severe jaundice (hyperbilirubinemia)

Causes of ANSD before birth may be genetic reasons (inherited), even if there is no history of hearing loss being passed down in the family. ANSD may be associated with certain infections during pregnancy, such as cytomegalovirus (CMV), mumps and meningitis. Sometimes ANSD can involve an absent or underdeveloped hearing nerve.



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***ANSD has many possible causes and is more common among babies with complex additional needs or babies who have a history of serious health issues as a newborn.***



*“Our daughter was born at 24 weeks, weighing only 650 grams. She was considered a Micro Premie (a premature baby born younger than 26 weeks gestation) and was given a 60% chance of survival. Today, she is a healthy, happy, active and very feisty 2 year old. She loves going out, singing and dancing. She can say words such as “shoe, jacket, outside, upstairs, downstairs, Jeh Jeh for sister in Chinese, Daddy, sit, hot, wash hands, sorry”.*



3 months

*We don't know what the future will bring, with her ANSD and how it will affect her hearing and language development. We're optimistic that she will excel and shine no matter what the outcome will be.”*



2 years





## What tests identify ANSD?

If ANSD is suspected your child will participate in several types of hearing tests. Each of them tests a different part of the hearing pathway.

- ▶ **Otoacoustic Emission (OAE)** testing is a quick measure of cochlear function that tells us how the outer hair cells in the cochlea are working. For a child with ANSD, there is a problem with how sounds are transmitted after the outer hair cells. Therefore, even though the outer hair cells may be working and OAEs may be present, it is possible that the child is unable to hear sound or that the child hears sound but it is unclear.



- ▶ The **Auditory Brainstem Response (ABR)** test does a good job of predicting a child's hearing levels when the hearing nerve sends signals to the brain in a coordinated way. With ANSD, however, the ABR measurements are absent or severely abnormal because there is a problem with how information is sent along the hearing nerve to the brain – that is, the hearing signals are not sent in a coordinated way. This means that although the ABR test can *identify* ANSD, it cannot be used to predict how a child will respond to sounds. Very often, the ABR will be absent even though a child may respond to some sounds. Part of the ABR test measures the “cochlear microphonic” which is an indication of how the outer hair cells in the cochlea are working.
- ▶ The **Middle Ear Muscle Reflex** is a contraction of tiny muscles in the middle ear that typically occurs in response to a loud sound. Audiologists often measure this reflex as part of a child's hearing test. These reflexes are often absent in children with ANSD because there is a problem with how the hearing nerves send messages to and from the brain.



*“My little boy was born at 25 weeks and spent 7 months in the NICU at Children’s hospital. My early experience with his hearing loss was very frustrating because I knew that he could hear although he did not always respond to sounds. After two ABR tests, it was a relief for me to finally get answers. When he was identified with ANSD, it was one more piece of the complex puzzle. I am so glad that I persevered to get a final result.”*

The combined results from these tests can help identify ANSD. The typical pattern of hearing test results seen in children with ANSD is:

- ▶ Evidence of outer hair cell function [otoacoustic emissions (OAEs) and/or cochlear microphonic]
- ▶ Absent or severely abnormal Auditory Brainstem Response (ABR)
- ▶ Absent middle ear muscle reflexes even though ears are clear of fluid or wax

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*Unlike other types of hearing loss, the ABR results in ANSD cannot predict a child's hearing levels.*





### How will ANSD affect my child?

Infant hearing testing results can *identify* ANSD, but do not provide enough information about hearing abilities. Possible technology options such as hearing aids, therefore, are only available when children with ANSD are old enough and developmentally ready for behavioural testing. The best way to determine the ways ANSD affects your child will be through working with your early communication specialists and audiologist to observe your child's behaviour in response to sounds, and to closely monitor his or her speech and language development. Regular testing and careful observation will be very important to build a complete picture.

**Behavioural hearing testing** in a quiet room can provide information about a child's hearing levels for different pitches (frequencies) of sound, as well as his or her understanding of speech. Children who are typically-developing are usually ready to start behavioural testing when they are 6-9 months old. Children born prematurely or who have complex medical needs may be older before an accurate assessment can be made.



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*Early hearing assessment can identify whether ANSD is present, but it will take time, frequent hearing testing, and involvement with early communication specialists to learn how your child uses their hearing to understand speech. It is often not until your child is ready for behavioural hearing testing that the Audiologist will be able to determine how your child understands sound.*

For young infants or children with ANSD who are not able to provide reliable behavioural hearing test results, **cortical auditory evoked potential testing** may be completed to help provide an estimate of how well a child hears. Cortical auditory evoked potential responses may be present in a child with ANSD when the ABR is absent because this test can sometimes detect hearing messages even when the hearing nerve responses are not well coordinated.

The setup for this test is similar to the ABR except your child may be awake during the assessment.

Although this test is not yet widely used clinically, it is offered at BC Children's Hospital to any child who has been identified with ANSD. Please discuss this option with your Audiologist.

**Parent observations** about how your child responds to everyday sounds are very important. For example:

- ▶ How does your child react to different sounds?
- ▶ Does your child react differently when he or she cannot see the sound source?
- ▶ Does your child startle, jump or cry when there is an unexpected or loud sound?
- ▶ What does your child do when you speak to him/her?
- ▶ What types of sounds is your child producing?
- ▶ Does he/she make the same sound all of the time or make a variety of different sounds?
- ▶ Does he/she imitate sounds you make?

Between hearing tests, it is helpful to videotape or keep a journal of your child's responses to sounds, as well as examples of when they do not respond.



## How will I communicate with my child?

**No one approach works for every child with ANSD.** Some children will benefit from using visual communication (i.e., sign language) as their primary language, while other children may use technologies such as hearing aids and/or cochlear implants to help develop spoken language. Some children will use a combination of visual communication, listening technology and spoken language. Families may start with one communication approach and then adapt or change the approach as they learn more about their child's strengths and preferences.

*“Our consultant could see that our direction was changing, and she was open and supportive of our decision to change to another agency.”*

**It is important to begin working as soon as possible with an early communication specialist** because babies' brains are programmed to begin learning language

*“We had sessions in our home to meet the needs of our daughter's language development. We learned communication and observation techniques.”*

even before birth. It is more difficult for your child to catch up once there are language and communication delays. Involvement with qualified professionals will provide your family with the information to help monitor and develop your baby's language.



**Every child with ANSD is unique and requires their own communication approach.** Your child's early communication specialist will work closely with you and your child's audiologist to help develop an Individualized Family Service Plan (IFSP) that will outline your goals for your child, and will identify strategies to achieve them. The early communication specialist will help you observe your child's responses and determine what language strategies and hearing equipment bring about the best responses to sound and early language.

**It is through careful monitoring, guidance and teamwork that families can try a communication approach to evaluate how well it is working over a period of time.** Communication approaches can and should be modified in a timely way to ensure your child's language continues to progress. It is critical to ensure your child is progressing as expected.

*"We felt bonded with our daughter but the lack of spoken language and communication caused frustrations. We began to provide her with auditory and visual cues and sign language to give her as much information as we could. This not only helped her immensely at the time but continues to be a communication saver."*

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*It is important to begin working with an early communication specialist as soon as possible. The early communication specialist will help you evaluate and monitor whether a communication approach is successful and will help you to adapt or change approaches to ensure your child's language development continues to progress. They'll also help introduce you to other parents and role models.*





## What kind of technology might help my child?

ANSD cannot be corrected with medication or surgery but many children with ANSD may benefit from the use of hearing technology. Some children with ANSD will benefit from the use of hearing aids while others may benefit more from a cochlear implant. For these children, the use of a Frequency Modulated (FM) or Digitally Modulated (DM) system will often improve their listening in the presence of background noise, for example, during storytime in the classroom. A small number of children with ANSD will not use either type of technology, either because they have near-typical hearing, or they will not benefit from the technology (no hearing nerve).



**Hearing aids** are small electronic devices that sit in or on the ears and work by making sounds louder. For most permanent hearing losses, using hearing aids to make sounds louder will help to make speech easier to understand. Hearing aids may help some children with ANSD. However, for other children with ANSD, making sounds louder may not necessarily make speech easier to understand.

*“It was very difficult to get our son to accept the use of his hearing aids, and this remains an ongoing struggle. Knowing that there are other strategies to encourage our son’s language development was helpful.”*

*“When our daughter got hearing aids, we could tell right away she could now hear us whisper. Previously, she did not respond to soft sounds or whispers but now she does. It was a huge relief that the aids helped.”*

At this time, there is no reliable way to predict whether hearing aids will work well for your child. The only way to find out if they are a good option for your child is to try them, usually as soon as reliable behavioural hearing test results are available. A hearing aid trial will be arranged by your local audiology clinic.



A **cochlear implant** is a surgically implanted device that directly stimulates the hearing nerve. It may provide access to sound for people who have an intact hearing nerve and severe-to-profound hearing loss in both ears when they receive little or no benefit from hearing aids. Children with ANSD may benefit from a cochlear implant, but a wide range of results are observed. Ask your audiologist if you would like more information about cochlear implants.

*“When our child was first identified with ANSD she did not get much benefit from hearing aids so we looked into a cochlear implant. Our daughter received a cochlear implant and today talks and behaves as any 5-year-old would. My advice to families would be that knowledge helps you to make the best choices for your family. Always be an advocate, do your research and ask questions!”*





A **Frequency Modulated (FM) system** or **Digitally Modulated (DM) system** is a wireless device that sends information from a speaker to a listener. FM/DM systems may be used in addition to hearing aid(s) or cochlear implant(s), but can also be used on their own. An FM/DM system is made up of a microphone that is usually worn by a teacher or parent and a small radio receiver.

The radio receiver may be connected to a child's hearing aid(s) or cochlear implant(s) but it can also be worn on its own just like a hearing aid. Some classrooms have a soundfield/speaker system on the wall so that all the children in the classroom can hear the teacher talking. FM/DM systems make speech easier to understand in difficult places to listen such as in background noise, when the speaker is at a distance or in groups (e.g., classrooms, gyms, or restaurants). Talk to your audiologist about purchasing or borrowing options.

*“We are just beginning to use an FM/DM system. We have been slowly introducing the FM/DM system on walks with our daughter and in noisy situations. We hope that she'll be able to benefit from using it in noisy places such as a daycare setting.”*

*“I still remember the first day my son wore his FM/DM system to preschool. He came home singing five verses of “I am a Pumpkin.” He never picked up so much in one day of preschool!”*





## Who will help me support my child's development?

Your child and your family will be supported by a team of professionals. In addition to regular contact with your Audiologist, key members of your team will be specialists that have expertise in working with deaf and hard of hearing infants and young children for early communication development. You will work with a service provider whose professional background may be:

- ▶ Speech-Language Pathology
- ▶ Teacher of the Deaf and Hard of Hearing
- ▶ Listening and Spoken Language Specialist

If your child is under 2 years a service coordinator will contact you about the services available.



Depending on the needs of your child, you may work together with:

- ▶ Sign Language Instructor
- ▶ Infant Development Program Consultant
- ▶ Occupational Therapist
- ▶ Physiotherapist
- ▶ Developmental Psychologist
- ▶ Physicians from the fields of Otolaryngology, Pediatrics, Genetics and Neurology
- ▶ Ophthalmology and Vision Consultant
- ▶ Nurses

*“Parents play an important role in relaying information to the team. You need to make sure your health care professionals understand what else is going on with your child outside of their specialities.”*

*“Understanding our family as a whole and how to balance our daughter's special needs within the family dynamic was important.”*



Parents often tell us that the support and practical information they receive from other parents and role models is very helpful. You may benefit from connecting with another parent or a role model through the BC Early Hearing Program Hands and Voices Guide By Your Side program. Trained parent guides and role models can provide you with emotional support and unbiased information and they can share their experiences. Ask your team about a referral to this program.

*“Our guide was outstanding in every aspect. She made us aware of programs and gave us the support to pursue them.”*

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*You are the most important person on your child's team. It will be important for every member of your child's team to work together.*



## Where can I get more information?

### Are there websites with more information?

If you are interested in learning more about ANSD, we suggest you visit the following websites:

<http://www.phsa.ca/earlyhearing>

<http://www.bchandsandvoices.com/>

**“What works for your child is what makes the choice right”**

– Hands & Voices™

<http://www.handsandvoices.org/>

<http://www.babyhearing.org/hearingamplification/causes/neuropathy.asp>

<http://kidshealth.org/en/parents/ansd.html>

[http://www.ndcs.org.uk/family\\_support/how\\_ndcs\\_can\\_help/our\\_videos/videos\\_greenhill.html](http://www.ndcs.org.uk/family_support/how_ndcs_can_help/our_videos/videos_greenhill.html)

<http://www.nidcd.nih.gov/health/hearing/pages/neuropathy.aspx>





**ANSD is relatively uncommon. Parents have told us that it would be helpful to have a one-page description of what ANSD is to give to professionals on their child's team.**

**See next page.**

**Please feel free to share this with others involved with your child.**



To whom it may concern,

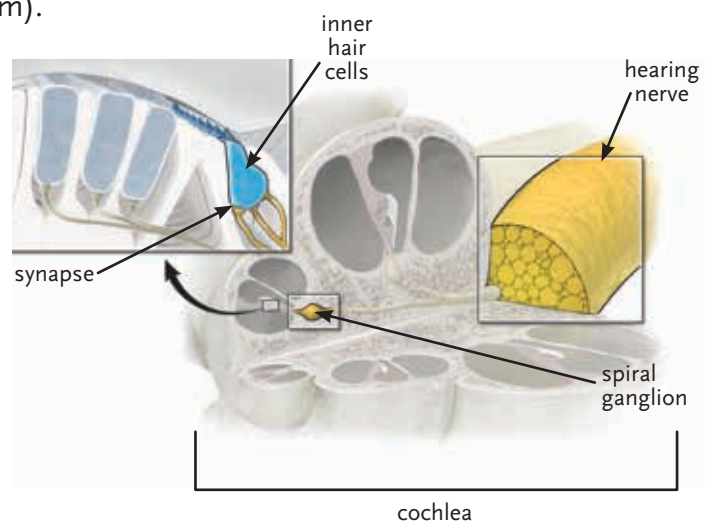
My child has Auditory Neuropathy Spectrum Disorder (ANSD). ANSD is relatively uncommon. Approximately 10 babies born each year in BC will have this disorder. The reasons why children have ANSD are not always known. We do know that ANSD is more common among children with complex additional needs or babies who have a history of serious health problems after birth. ANSD may be genetic or associated with certain infections during pregnancy. Sometimes ANSD can involve an absent or underdeveloped hearing nerve. With ANSD, the outer hair cells in the cochlea may work well, but a problem occurs beyond this point in the hearing pathway (see diagram).

It is thought that ANSD results from a disruption of the sound signals received in the inner ear along their way to the brain. This disruption of how sound typically travels to the brain will often cause a child with ANSD to have difficulty understanding complex sounds, such as speech.

An Auditory Brainstem Response (ABR) test can detect ANSD, while other tests, such as otoacoustic emissions (OAE), cannot detect ANSD in isolation. Although tests during infancy can identify ANSD, they do not provide information about hearing abilities, such as a child's hearing levels and/or speech understanding. Possible technology options such as hearing aids, therefore, are only considered when children with ANSD are old enough and developmentally ready for behavioural testing (typically not before 7-8 months corrected age).

Our observations of my child's responses to sound are important. A team of professionals is helping us closely monitor my child's listening, speech and language development. We are learning communication strategies to help prevent language delays.

For more information please visit: [www.phsa.ca/earlyhearing](http://www.phsa.ca/earlyhearing) or contact my child's Audiologist:



*The name of my child's Audiologist*

*Contact Info*

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For more information, contact your  
local public health audiology clinic, visit the  
BCEHP website at [www.phsa.ca/earlyhearing](http://www.phsa.ca/earlyhearing)  
or call 1-866-612-2347

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